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Brief Reports

Massive Congenital Orbital Teratoma

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Abstract: **Orbital teratomas are rare embryonic tumors composed of a wide diversity of tissues derived from the three germinal layers. The presenting features include, a healthy newborn with extreme unilateral proptosis; marked stretching of the eyelids over a tense, fluctuating mass, with elongation of the palpebral fissure; enlargement of the bony orbit (two to three times normal size) with subsequent nasal and malar deformities; and transillumination of all or part of the orbital mass. Commonly the eye is normally developed but often vision is not preserved either due to exposure or secondary optic atrophy. The objective in the management of orbital teratoma is to save the eye to encourage orbitofacial development, maintain cosmesis and retain some vision. We report a case of massive congenital orbital teratoma successfully removed by an eyelid-sparing exenteration technique.**

Congenital teratomas of the orbit are rare tumors that produce massive unilateral proptosis in a newborn.[1,2] Primarily intraorbital and histopathologically benign, they are composed of a wide variety of tissue derived from all three embryonic germinal cell layers.[1] Teratomas can, however, cause secondary damage to the globe and extensive destruction of the bony orbital and surrounding facial structures.[3] We report a case of massive primary congenital orbital teratoma successfully removed by an eyelid-sparing modified exenteration technique.

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CASE REPORT

An otherwise healthy girl was born at 38 weeks gestation, by elective cesarean section, with an obvious dramatic left eye proptosis. Her 36-year-old mother was healthy with normal antenatal history and ultrasonography.

Ophthalmologic examination of the infant revealed massive left proptosis and a large tumor filling the orbital contents (Figure, A). The mass was soft to palpation, nonpulsatile, and irreducible. There were large cystic areas that transilluminated along approximately one-half of the tumor. The globe was displaced superotemporally and protruded through the palpebral fissure with conjunctival chemosis, corneal edema, flat anterior chamber, prominent iris vessels, nonreactive pupils, and no red reflex. The right ocular and orbital examination was normal.

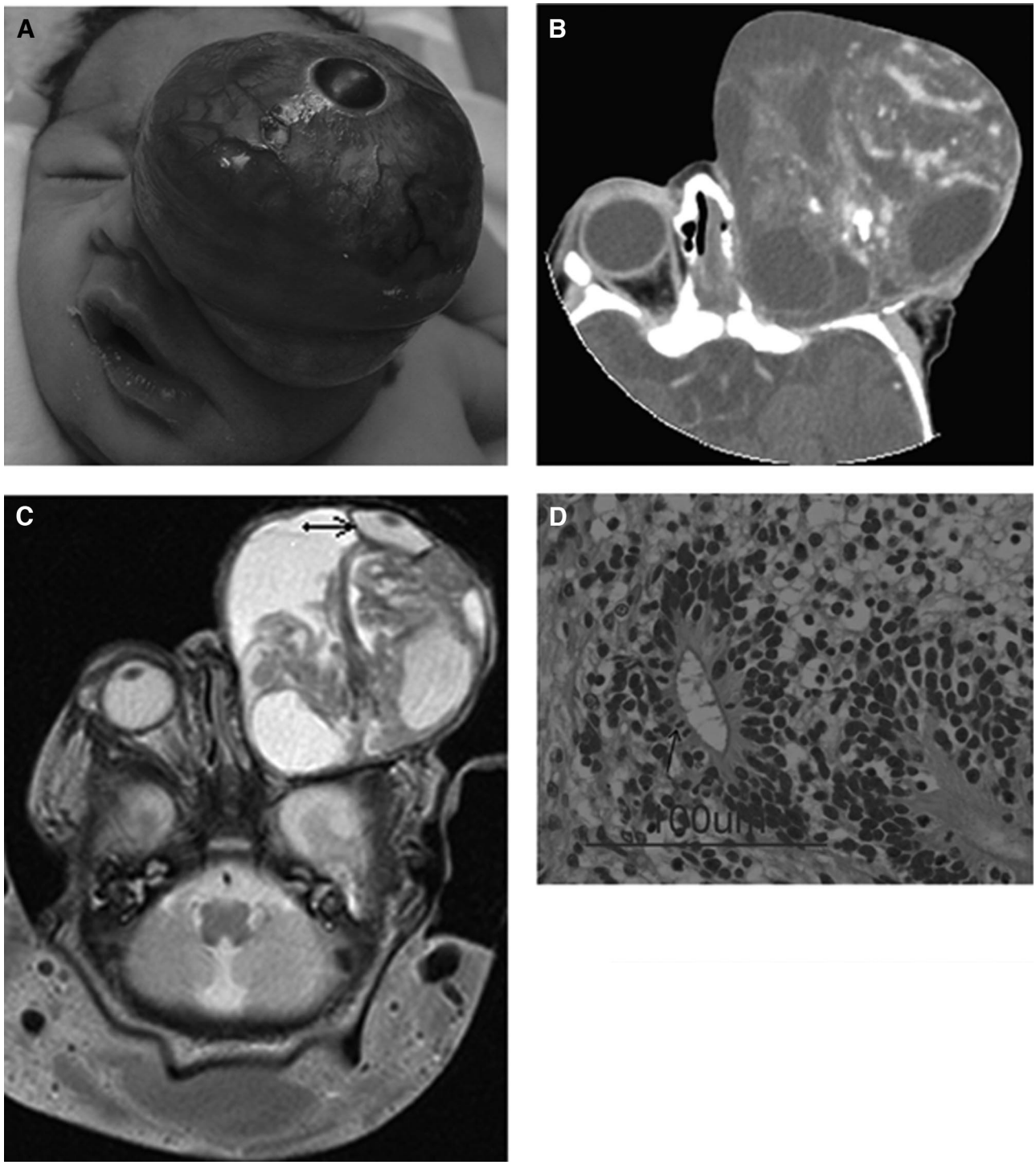
CT (Figure, B) and MRI (Figure, C) of the orbit and brain showed a large mass measuring $6.4 \times 6.5 \times 5.8$ cm that encased the entire orbit, bulging anteriorly and displacing the globe forward. It was heterogeneous with solid and cystic components and fat, coarse calcification, and large enhancing blood vessels. There was significant expansion of the left orbital structures, with no evidence of intracranial extension or erosion through the paranasal sinuses. The right orbit appeared normal.

At the age of 11 days, after informed consent, an eyelid-sparing exenteration procedure was carried out and the tumor was excised *en bloc* posteriorly. The postoperative period was uneventful.

The microscopic appearance of the tumor consisted of innumerable cystic structures lined by a variety of epithelia. Derivatives of all three germ lines were represented. Both central nervous system and peripheral nervous system were noted, with evidence of primitive neuroepithelium (Figure, D). Cartilage, bone, fat, and skeletal muscle were represented with scattered foci of immature mesenchyme with mitotic activity. The eye was distorted with the optic nerve surrounded by teratoma. The final diagnosis was Grade 1 immature teratoma. Three months postoperatively, the socket was well healed with no evidence of infection. Further reconstructive surgery will be deferred.

DISCUSSION

Teratomas are embryonic tumors composed of a wide diversity of tissues foreign to the anatomic site in which they arise. The characteristic features of orbital teratomas include an otherwise healthy newborn with extreme



A, Clinical photograph showing massive proptosis, superotemporally displaced globe protruding through the palpebral fissure with conjunctival chemosis and corneal edema. B, Axial post contrast CT section through the head and orbits shows a large soft tissue mass protruding from the left orbit. The mass is of heterogeneous density, containing fat, various soft tissue components, calcifications, and enhances markedly though not uniformly. The left medial orbital wall is bowed medially secondary to mass effect, but there is no gross intranasal extension. C, Axial T_2-weighted MRI shows the heterogeneous mass with both its fatty and cystic components having high signal characteristics. The globe is severely compressed (arrow) on the anterior edge of the orbital mass as the course of the optic nerve can be followed posteriorly in the orbit. D, A primitive neuroepithelium shows two mitotic figures (arrow) immediately adjacent to the left luminal surface (hematoxylin and eosin stain).

unilateral proptosis, marked stretching of the eyelids over a tense, fluctuating mass, elongation of the palpebral fissure, absence of demonstrable communication between the cyst and the intracranial cavity, normally developed eye that may exhibit degenerative changes secondary to the displacement by the teratoma, and transillumination of all or part of the orbital mass.[2] Other features include no family history of congenital deformities with non-consanguineous parents and normal siblings, normal pregnancy and delivery, no history of teratogenic influences to the mother, the presence of enlarged bony orbit without bony destruction, and calcification and ossification of the tumor evident on imaging.[3] Commonly the eye is normally developed but often vision is not preserved either due to optic atrophy or corneal exposure.[3] Imaging characteristically shows a heterogeneous structure with focal punctuate calcification and areas of fat density.

Histologically, teratomas are composed of tissues derived from the three germinal layers.[1,5] The predominant germ cell types observed in orbital teratoma are surface ectoderm producing squamous epithelium-lined cyst, hair follicles, and sweat glands. Neuroectodermal tissues include primitive neural tubes, choroidal plexus, and ganglia. Mesoderm is the next most common cell layer represented by the muscle, bone, cartilage, and fat. Endoderm is the least common and may produce gastrointestinal tissue cysts lined by respiratory-type psuedostratified columnar epithelium. Cystic spaces lined by glandular epithelium are responsible for the rapid enlargement of the lesions. These tumors have been previously reported as benign,[5] and are considered malignant when the tissue is embryonal or immature in nature.[3] The prognosis of the immature teratoma is related to several factors including the age, site, and grade of immaturity, and is usually good if complete excision is performed early in neonatal life.[5]

The objective in the management of orbital teratoma is to save the eye to encourage orbitofacial development, maintain cosmesis, and retain some vision. Preservation of the globe is not possible if there is no organized eye or optic nerve present, when the tumor engulfs the optic nerve, and there is extreme proptosis, exposure keratopathy, and no pupillary reflex.[6] Our patient had a massive tumor encasing the globe and filling the orbital contents. There was no visual potential; thus an eyelid-sparing, exenteration was performed and the tumor was excised *en bloc* posteriorly. Further craniofacial surgery and rehabilitation will be dependent on future midfacial growth and development.

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Orbital Perforating Branch of the Infraorbital Artery: An Important Landmark in Orbital Surgery

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Abstract: **The orbital branch of the infraorbital artery is an important surgical landmark but is frequently omitted from texts that discuss orbital anatomy and surgical technique. This report reviews the anatomy of this artery to familiarize the reader with its existence and location to reduce the risk of intraoperative hemorrhage from this vessel.**

Successful orbital surgery is predicated on an excellent understanding of the regional anatomy to maximize surgical exposure while minimizing complications. This is facilitated by the identification of key landmarks during dissection within the orbit.

Well known to the orbital surgeon are the zygomaticotemporal, zygomaticofacial, and anterior ethmoidal vessels. These not only serve as landmarks but are frequently intentionally sought to cauterize to prevent hemorrhage during surgery.

Perhaps less well known is the orbital branch of the infraorbital artery. Our lack of familiarity with this vessel resulted in potentially preventable intraoperative orbital hemorrhage and prompted us to investigate its consistency and precise anatomic location.

We reviewed oculoplastic surgery and orbital anatomy texts to determine whether the orbital branch of the infraorbital nerve was described. Additionally, we measured the location of the artery along the orbital floor in 10 consecutive floor explorations (excluding acute trauma). Finally, we reviewed the clinical course of a

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Artery location in 10 consecutive patients in whom the orbital floor was explored (excluding acute trauma)

Procedure	Location from infraorbital rim (mm)
Exenteration	15
Exenteration	14
Enophthalmos repair	14
Enophthalmos repair	14
Enophthalmos repair	17
Enophthalmos repair	13
Orbital decompression	15
Ahmed shunt to maxillary sinus	13
Ahmed shunt to maxillary sinus	13
Ahmed shunt to maxillary sinus	15

patient who had significant intraoperative hemorrhage from the orbital branch of the intraorbital vessel.

CASE REPORT

A 41-year-old woman with atraumatic enophthalmos caused by maxillary atelectasis underwent surgical exploration of the intact, depressed orbital floor for repair of the induced enophthalmos and hypoglobus. Immediately on releasing the arcus marginalis at the level of the anterior orbital rim, an avascular dissection plane was encountered, as anticipated, as the orbital floor dissection was carried out posteriorly to facilitate placement of a subperiosteal orbital floor implant. While proceeding with the subperiosteal floor dissection, a perforating artery connecting the infraorbital canal with the periorbita in the region of the inferior rectus was accidentally lysed, resulting in intraoperative orbital hemorrhage. Cautery in the region of this vessel resulted in transient hypesthesia in the V-2 distribution and a focal burn to the eyelid margin caused by the use of uninsulated bipolar forceps. This unanticipated event prompted us to more systematically review the vascular anatomy of the orbital floor described in this report.

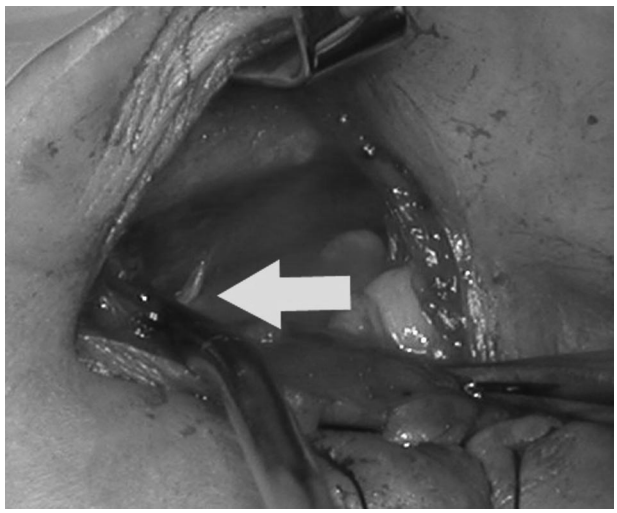

FIG. 1. Intraoperative photograph during orbital floor exposure demonstrating the location of the orbital branch of the infraorbital artery.

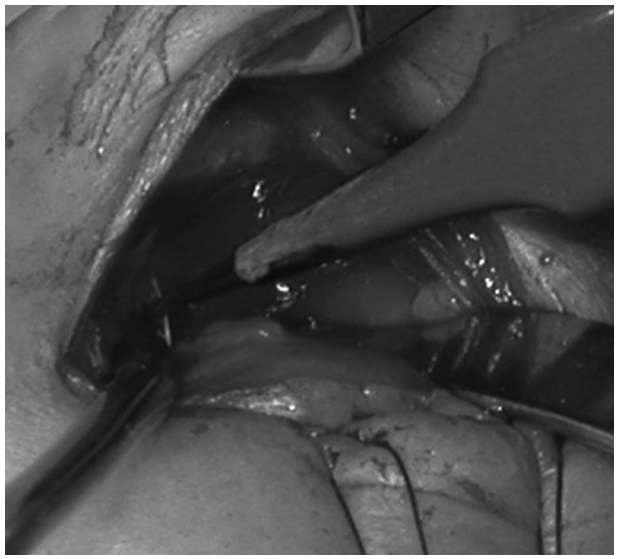

FIG. 2. Bipolar cautery applied to the artery to prevent hemorrhage.

We observed the artery in every case in which we have explored a nontraumatized orbital floor. In 10 consecutive cases in which the orbital floor was explored (excluding acute trauma), we observed the artery 13 to 17 mm posterior to the rim (average = 14.4 mm; Table).

DISCUSSION

Few texts on orbital surgery describe the orbital branch of the infraorbital artery and the importance of identifying it to reduce the risk of hemorrhage. The artery is mentioned in the anatomy texts of Whitnall[1] and Dutton[2] and was addressed by Coulter et al.,[3] who described a patient with orbital hemorrhage after fracture repair. Re-exploration of the floor in that patient revealed bleeding from the orbital branch of the infraorbital artery.

The orbital branch of the infraorbital artery is an important landmark during orbital surgery, and, once sensitized to its presence, we observed it in all cases of nontraumatic floor exploration. It is consistently located approximately 14 mm posterior to the rim, in line with the infraorbital nerve. We advocate exploration and bipolar electrocauterization of this structure, which may be mistaken for avascular connective tissue band when exploring the orbital floor. Unipolar (Bovie) cautery should be avoided to minimize damage to the adjacent infraorbital nerve; bipolar cautery with insulated bayonet forceps on a low setting is preferable. This is especially pertinent in cases in which there has been no prior trauma to the orbital floor that could have induced other artifacts in the surgical field. Failure to recognize this artery may result in orbital hemorrhage (Fig. 1 and Fig. 2).

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Primary Orbital Ewing Sarcoma in a Middle-Aged Man

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Abstract: **A 54-year-old man presented with a painful left proptosis and a soft tissue mass at the inferolateral aspect of the left orbit with bone involvement. There was no clinical or investigational evidence of systemic disease. Both light microscopy and immunohistochemistry were required for the diagnosis of Ewing sarcoma. After failure of chemotherapy alone, management included extensive surgical excision and postoperative adjuvant radiotherapy and chemotherapy. Despite intervention, the patient died of widespread metastatic disease 17 months after initial presentation. Although rare, Ewing sarcoma should be included in the differential diagnosis of uniform round cell orbital tumors in adults.**

Ewing sarcoma is a highly malignant neoplasm, most commonly affecting young men or boys under the age of 20 years. It primarily arises in the long bones, ribs, and pelvis.[1,2] It is a round cell tumor and is thought to be of neuroectodermal derivation.[1,3] The incidence of Ewing sarcoma in the head and neck is approximately 4%, favoring the mandible and maxilla.[1] Most cases of orbital involvement are the result of metastasis from a distant site or extension from an adjacent compartment. Of the few reported primary orbital tumors,[4–10] to our knowledge, only two have occurred in patients beyond the third decade of life.[7–9]

CASE REPORT

A 54-year-old man had painful left orbital swelling unresponsive to antibiotics. Medical and ocular history were otherwise negative. Snellen acuity was 20/20 OU, and examination of the right eye was normal. Proptosis and a palpable mass were present on the left side; examination of the left eye was otherwise unremarkable.

Computed tomography revealed a soft tissue mass in the inferolateral aspect of the left orbit, with associated mottled bony destruction and extension in the maxillary antrum (Fig. 1, a and b). Comprehensive physical examination and systemic investigation, including bone scan, bone marrow biopsy, and full-body CT scan, failed to reveal other foci of disease. A biopsy was performed through a subciliary incision. Histopathological examination revealed fibrous tissue diffusely infiltrated by sheets of large, cytologically uniform, round tumor cells with oval nuclei (Fig. 2a), inconspicuous nucleoli, little cytoplasm, and a marginally increased mitotic rate. There was no frank pleomorphism or nuclear hyperchro-

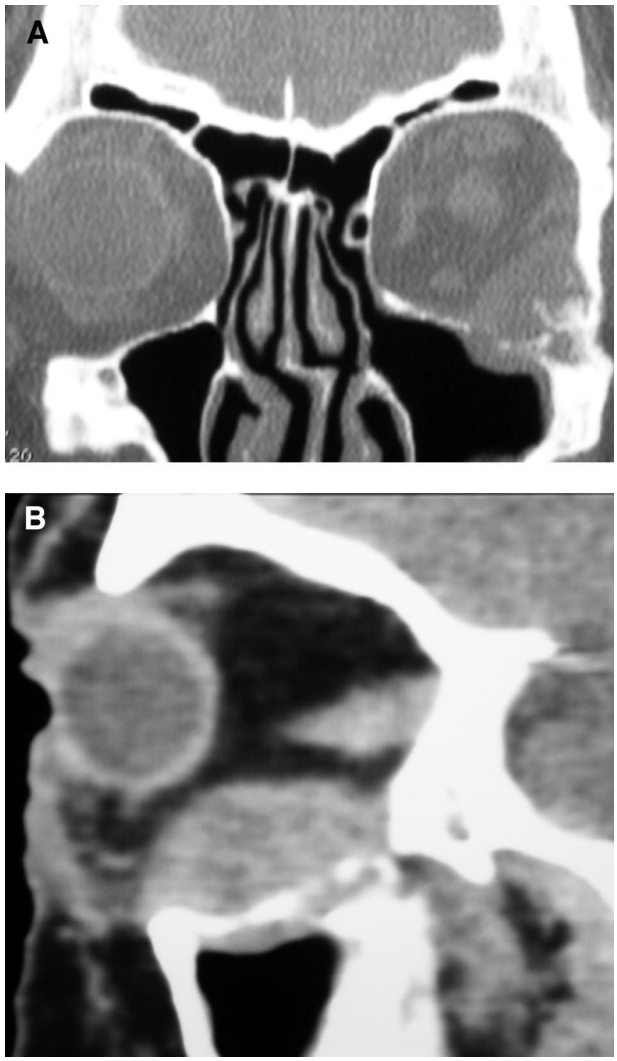

FIG. 1. *A*, Coronal CT showing mass in the inferolateral left orbit with bone destruction. *B*, Sagittal oblique soft tissue CT showing mass extending in the posterior orbit and maxillary sinus.

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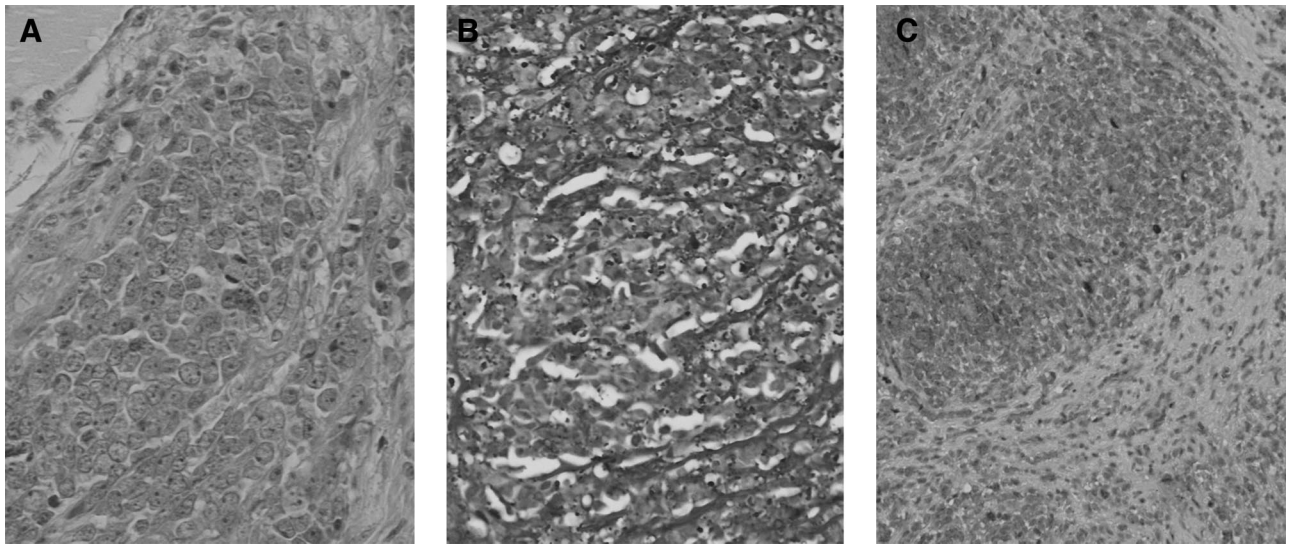

FIG. 2. *A*, Fibrous tissue is diffusely infiltrated by large, round tumor cells, with a marginally increased mitotic rate but without frank pleomorphism or nuclear hyperchromatism (hematoxylin and eosin stain, original magnification ×400). *B*, The cytoplasm of some cells contained glycogen granules (periodic acid-Schiff stain, original magnification ×400). *C*, Tumor cells positive for neuron-specific enolase (NSE) by immunohistochemistry (magnification ×100).

matism. The cytoplasm of some cells contained periodic acid-Schiff–positive glycogen (Fig. 2b). No rosettes were present. On immunohistological examination, the tumor cells reacted positively for neuron-specific enolase, vimentin, and MIC2 (CD-99) (Fig. 2c) but negatively for keratin, S-100, epithelial membrane antigen, and leukocyte common antigen. These features were consistent with a diagnosis of Ewing sarcoma.

Three cycles of induction multiagent chemotherapy (ifosfamide, doxorubicin, and vincristine) did not reduce the size of the tumor. Surgical excision included radical orbitectomy, anterior craniotomy, partial upper maxillectomy, and excision of the left zygoma and lateral wall of the nose. Histologic examination showed clear surgical margins except at the posterolateral angle of the left orbit. There was no evidence of vascular or perineural invasion, and a preauricular lymph node was free of tumor. Postoperative adjuvant radiotherapy (54 Gy in 30 fractions) and chemotherapy (three cycles of carboplatin and etoposide) were instituted. The tumor enlarged despite multitherapy, and, 6 months after surgery, CT showed diffuse skeletal metastases. The patient died of widespread metastatic disease 17 months after initial presentation.

DISCUSSION

Recent studies indicate that Ewing sarcoma and primitive neuroectodermal tumors (PNET) are both of neuroectodermal derivation and are closely related.[3] The term Ewing sarcoma is reserved for poorly differentiated tumors that lack light and electron microscopic and immunohistochemical evidence of neuroectodermal differentiation. R3-IOP200038 5 Ewing sarcoma/PNET accounts for 6% to 8% of primary malignant bone tumors,[3] shows a predilection for male subjects, and has a peak incidence in the second decade of life. The tumor preferentially arises at the diaphysis and metaphyseal-diaphyseal portion of long bones.[1–3] Involvement of the orbit is rare, and most reported cases have occurred in young children.[1,4–6,10,11]

Immunohistochemistry and light microscopy are required to differentiate Ewing sarcoma from other round cell tumors.[5] Immunohistochemical stains for neuron-specific enolase, myoglobin, and leukocyte common antigen are positive in neuroblastoma, rhabdomyosarcoma, and lymphoma, respectively. All of these stains are usually negative in Ewing sarcoma, although neuron-specific enolase may be positive,[3] as in the current case. S-100 is found in all cases of neuroblastoma but is absent in Ewing sarcoma. MIC2 (CD-99) is expressed in a characteristic membranous pattern in Ewing sarcoma/PNET.[3] Ewing sarcoma expresses a characteristic chromosomal translocation [t(11;22)(q24;12)] in 85% of cases,[3,12] which can confirm the diagnosis in uncertain cases, although this was not tested in the current case. The histology of Ewing sarcoma is variable, with most lesions composed of uniform small, round cells with nuclei containing fine chromatin and scanty cytoplasm; others have larger tumor cells with prominent nucleoli, as seen the current case.[3] Specific microscopic features of Ewing sarcoma include presence of glycogen and sparse cell junctions on electron microscopy.[3,5]

The atypical site and older age of our patient were unusual. To our knowledge, only two cases of primary orbital osseous Ewing sarcoma in a patient over 40 years of age have been reported.[7–9] In addition, two cases of

extraosseous orbital Ewing sarcoma have been reported in adults over 50 years of age.[1,13]

Our patient presented with periorbital swelling of relatively short duration, and the only clinically significant findings were mild proptosis and a palpable mass. Other reported clinical manifestations of orbital Ewing sarcoma have included pain, headache, ophthalmoplegia, papilledema, and loss of vision.[5,11] The characteristic radiologic appearance of Ewing sarcoma is an infiltrative lesion featuring moth-eaten or mottled bone destruction associated with an "onion skin–like" periosteal reaction.[1,3] Frequently, it is associated with an ill-defined soft tissue mass.[3] The associated mottling of the bone was seen in our case, but the characteristic "onion skin" periosteal reaction was lacking, consistent with similar reports in other orbital cases.[1,7] Orbital involvement by Ewing sarcoma is usually metastatic[5] or from direct extension, hence a vigilant search to exclude systemic disease is necessary. In the current case, whole-body CT and bone scans confirmed that the tumor was a primary neoplasm arising in the orbit.

A multidisciplinary approach for management of Ewing sarcoma is required. Surgery may be more effective in the initial local management, then radiotherapy for resectable lesions due to improved local control and the risk of postirradiation malignancies.[1] The prognostic factors include the stage, site, and size of the tumor. Metastatic tumors, or those that arise in the pelvis and are large, have a poor prognosis.[3] In recent years, the prognosis for Ewing sarcoma has dramatically improved with multiagent chemotherapy in conjunction with local resection and/or radiotherapy, with a 5-year survival rate of 45% to 75%.[1]

Although exceptionally rare, Ewing sarcoma should be considered in the differential diagnosis of uniform round cell orbital tumors in adults.

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Primary Orbital Leiomyosarcoma

I-Chan Lin, M.D., Chen-Tu Wu, M.D.†, Shu-Lang Liao, M.D.*, and Luke L.-K. Lin, M.D., Ph.D.**

Abstract: **An 84-year-old woman presented with a right upper eyelid nodule, first noted 1 month earlier. Incisional biopsy revealed leiomyosarcoma, which was subsequently treated with wide excision and adjunctive radiotherapy. There has been no evidence of tumor recurrence in 3 years of follow-up.**

Leiomyosarcoma is a malignant tumor that rarely develops in the orbit. It typically occurs in older women and has a high incidence of local recurrence and distant metastasis. Herein we report a case of orbital leiomyosarcoma and review the literature to distinguish characteristics of primary orbital leiomyosarcoma.

CASE REPORT

An 84-year-old woman was referred to our clinic for evaluation of a right upper eyelid nodule, which had been noted for 1 month. Ophthalmic examination showed a best corrected visual acuity of 40/50 OU, no relative afferent pupillary defect, and intact extraocular motility. Mild cataracts were present OU. A nontender, smooth, firm nodule superior to the medial canthal tendon was present. Orbital CT confirmed a right medial extraconal mass without bone involvement (Fig. 1). The patient underwent an incisional biopsy. The initial intraoperative frozen section analysis suggested a spindle cell tumor, probably of myogenic origin. On the permanent hematoxylin and eosin sections, the hypervascular tumor was composed of spindle cells arranged in fascicles or po-

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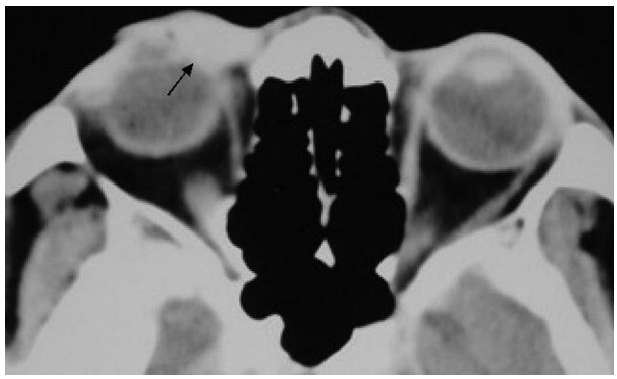

FIG. 1. Orbital CT shows focal area of a homogenous soft tissue prominence adjacent to the right medial rectus muscle without bone destruction.

lygonal cells in sheets with 7 mitotic figures per 50 high-power fields. The atypical nuclei were large and cigar-shaped. Immunohistochemical stains for the tumor cells were positive for smooth muscle actin (Fig. 2) but negative for S100 and CD 34. The features are diagnostic for a leiomyosarcoma. CT of the head, chest, and abdomen and a bone scan were negative. Three weeks later, the patient underwent anterior orbitotomy with a wide resection margin and semicircular flap reconstruction. After surgery, she received radiation therapy (5,000 cGY/25 fractions). She has done well, without evidence of tumor recurrence after 3 years of follow-up.

DISCUSSION

Leiomyosarcoma, a neoplasm of smooth muscle origin, mostly occurs in the uterus, gastrointestinal tract, and vascular tissue. Rarely does this tumor develop primarily in the orbit. Only a few cases have been reported in the English literature[1–4] (cases 1 through 4; Table, available online at www.op-rs.com). It typically develops in older women, and the average reported age of patients is 63.8 years (range, 36 to 84 years). Only one male patient has been described in the literature, and he was also the youngest patient with the disease to date.[2] This is in contrast to a leiomyoma, the benign counterpart of leiomyosarcoma, which characteristically occurs in younger individuals. The most common symptom for primary orbital leiomyosarcoma is painless proptosis, but decreased vision, diplopia, and palpable mass may also be present. The average duration of symptoms is 7.7 months, with a range of 1 to 18 months.

Orbital leiomyosarcoma has been reported as a complication of orbital radiation for retinoblastoma. Font et al.[5] reported a 31-year-old woman with bilateral retinoblastoma. The right eye was enucleated and the left eye irradiated. Leiomyosarcoma then developed in left periorbital area 23 years after. Folberg et al.[6] described two cases of orbital leiomyosarcoma secondary to radiation. Both patients were young men with bilateral retinoblastoma. This is in contrast to a primary orbital leiomyosarcoma, which predominantly develops in older women.[1,3,4]

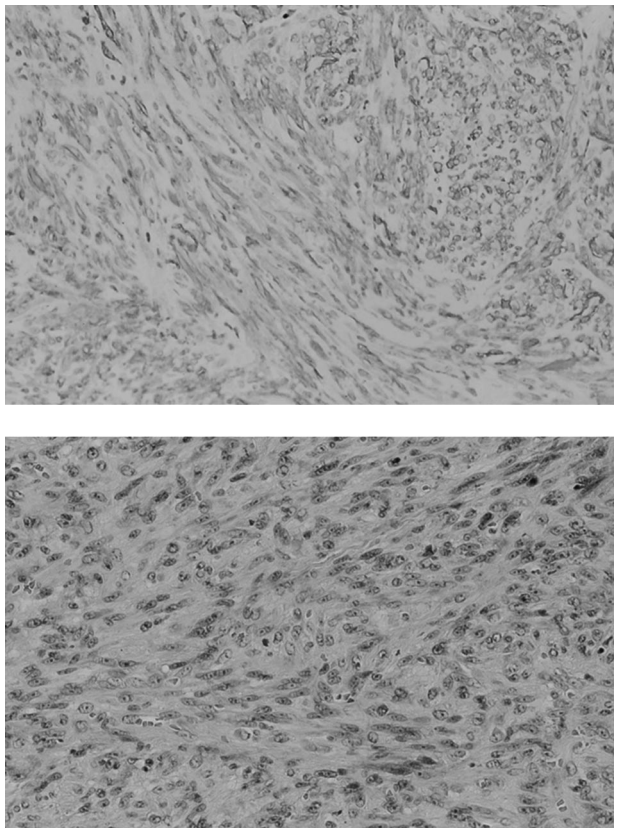

FIG. 2. Upper, Tumor cells are diffusely reactive to smooth muscle actin (ABC method, magnification ×66). Lower, Eosinophilic, hypercellular spindle tumor cells are in a vague fascicular pattern. The nuclei are large and cigar-shaped, with frequent nucleoli. Mitotic figures are seen frequently (hematoxylin and eosin stain, magnification ×66).

The origin of primary myogenic tumor in the orbit is suspected to be vascular smooth muscle. Three of the reported tumors developed in the posterior orbit, where vascular smooth muscle is found.[1,3,4] In the remaining case,[2] tumors were located in the anterior orbit where Müller sympathetic muscle origin was suspected. In our case, however, the tumor was located in the anterior orbit, but the vascular walls showed tumor cell proliferation and a vascular origin was suspected. The underlying cause of smooth muscle malignancies is controversial. De novo genesis appears to be rather more likely than malignant degeneration of primary benign lesions.

Histopathologic examination is necessary to make the diagnosis of leiomyosarcoma. The tumor is composed of spindle-shaped cells that have cigar-shaped, often bizarre, nuclei. Multinucleated cells are not uncommon. Nuclear pleomorphism, hyperchromatism, giant cells, and mitotic figures distinguish leiomyosarcoma from leiomyoma.[1] The grade of malignancy derives from the

number of mitoses, degree of cellular atypia, and the extent of tumor necrosis. Sometimes there are difficulties in differentiating smooth muscle tumors from peripheral nerve sheath tumors. Immunohistochemistry may provide useful clues to the diagnosis: Desmin and smooth muscle actin immunoreactivities are most frequently detectable in smooth muscle tumors. S-100 protein negativity, as in our case, rules out the diagnosis of neurogenic tumor.

The tumor could also be confirmed by electromicroscopic findings of well-developed basal lamina and characteristic dense bodies formed by contracted elongated bundles of smooth muscle myofilaments.[3]

Although CT and ultrasound are commonly used to evaluate cases of orbital leiomyosarcoma, Hou et al.[4] suggested that MRI can provide more important information regarding tumor location and extent. Hypointensity relative to the cerebral cortex on T_2-weighted MRI, moderate peripheral enhancement with contrast, and an extraconal location represent an unusual constellation of findings that may be useful for the diagnosis of a primary orbital leiomyosarcoma.

The initial treatment of all cases is surgical wide local resection. In a reported case with recurrent tumor, the patient underwent secondary local resection and eventual exenteration,[1] which failed to prevent distant metastasis and tumor-related death.

Orbital leiomyosarcoma may be friable or unencapsulated, which can prevent complete surgical resection and result in a high recurrence rate.[1] Orbital exenteration has been suggested as the treatment of choice.[1] We agree that early orbital exenteration offers the best chance for cure. Once there is local recurrence, orbital exenteration may not be of any long-term benefit, as evidenced by the cases reported by Jakobiec et al.[1] For tumor arising in the anterior orbit, as in our case, wide local resection with histologic frozen section control may be adequate, and adjunctive radiotherapy should be considered. In reviewing the literature, two patients[3,4] received postoperative radiotherapy; one of them[3] did not develop early tumor recurrence after radiotherapy. (Postradiotherapy follow-up data were not available for Hou's patient.[4]) Likewise, our patient received postoperative radiotherapy and did not have early tumor recurrence thereafter. Only Hou's patient received postoperative adjunctive chemotherapy (doxorubin and isofamide), resulting in shrinkage of the tumor.[4] Whether adjunctive chemotherapy will alter the long-term prognosis remains to be determined.

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Imaging of Metastatic Orbital Leiomyosarcoma

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Abstract: **A 74-year-old man with a previous history of lower leg soft tissues leiomyosarcoma and multiple metastasis presented with a progressive painless proptosis of the left eye. Orbital ultrasound, CT, and MRI revealed a large mass in the left medial orbit. The mass was surgically removed and pathologic examination confirmed the diagnosis of a metastatic orbital leiomyosarcoma. The imaging characteristics of this rare tumor are comprehensively detailed, using complementary ultrasound, CT, and MRI, the combination of which allowed planning of total excision of the lesion.**

A 74-year-old man presented with diplopia and painless left proptosis that had developed over 3 months. Twenty years previously he had surgical excision of a leiomyosarcoma (Trojani grade 1) from the soft tissues of his left lower leg. Later, he had had development of bilateral lung and multiple muscle metastases for which he declined treatment.

His left visual acuity was 20/20, he had normal color vision and visual field and no relative afferent pupillary defect. The left globe was displaced inferotemporally, and Hertel exophthalmometry revealed 3 mm of nonaxial

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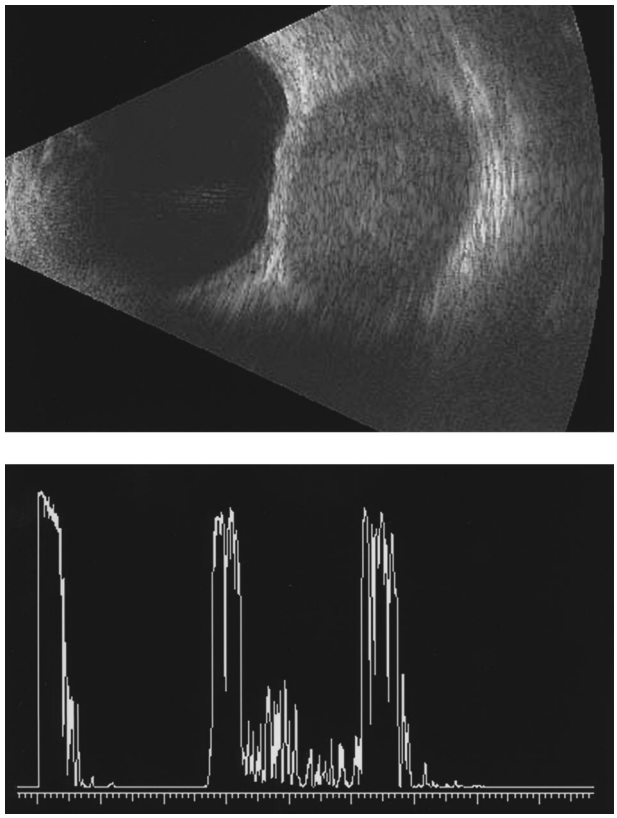

FIG. 1. Orbital ultrasonography. Upper, Large, well-defined, hyporeflective mass in the left medial orbit, which indents the medial globe with overlying choroidal folds. Lower, Standardized A-scan shows internal reflectivity with some irregularity in amplitude but fairly regular interval.

proptosis. Significant conjunctival hyperemia was observed medially. Fundoscopy revealed choroidal folds nasally.

B-mode ultrasonography showed a well-defined, hyporeflective mass in the left medial orbit measuring 25 × 21 × 16 mm, with highly reflective anterior border possibly suggesting a capsule. It was causing indentation of the medial globe and choroidal folds (Fig. 1A), its posterior edge was displacing laterally the optic nerve, and it could not be separately identified from the medial rectus muscle. Standardized A-mode ultrasonography showed medium to low internal reflectivity with some irregularity in amplitude but fairly regular internal structure (Fig. 1B).

Orbital CT showed a well-defined isodense soft tissue mass within the medial aspect of the left orbit, not clearly separable from the medial rectus muscle. MRI confirmed the presence of a well-defined intraconal soft tissue mass separate from the medial rectus and abutting the optic nerve. The mass demonstrated predominant signal intensity that was isointense to gray matter on T_1 and T_2 imaging and displayed prominent enhancement after contrast administration, particularly centrally (Fig. 2).

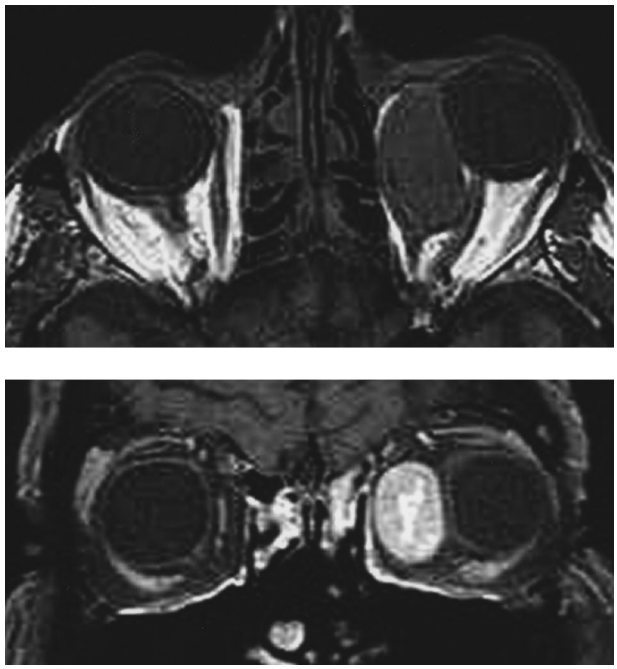

FIG. 2. Orbital MRI. Upper, Axial precontrast T_1 image of the orbits confirms the presence of a well-defined intraconal soft tissue mass, predominantly isointense to cerebral gray matter; it is separate from the medial rectus and abuts the optic nerve, with maintained fat planes. Lower, Coronal postgadopentetate image shows prominent diffuse enhancement of the mass, particularly in the central component.

Left lateral orbitotomy was performed, and the mass was approached through a medial transconjunctival incision. A solid well-demarcated mass was removed (Fig. 3A). Separation of the tumor from the medial rectus muscle and the optic nerve was readily achieved by dissection. Gross pathological examination revealed a well-circumscribed white firm mass measuring 27 × 21 × 16 mm. Microscopic examination revealed a spindle cell tumor, with a vaguely fascicular architectural pattern. The cells showed marked nuclear pleomorphism and hyperchromasia. There were more than 20 mitoses per 10 high-power fields. Immunohistochemical studies were strongly diffusely positive for smooth muscle actin but negative for desmin and S100 (Fig. 3, B and C). Based on these findings, the diagnosis of a Trojani grade 3 poorly differentiated leiomyosarcoma was made.

Six months after orbitotomy, diplopia had resolved and the left globe position and motility was normal. The left eye maintained 20/20 vision and normal color vision and visual field. Recent CT showed no local recurrence of the orbital tumor but did reveal rapid progression of his disease, with numerous new metastases involving multiple organs.

DISCUSSION

Metastatic orbital leiomyosarcoma has been published very rarely,[1–6] and to our knowledge detailed documen-

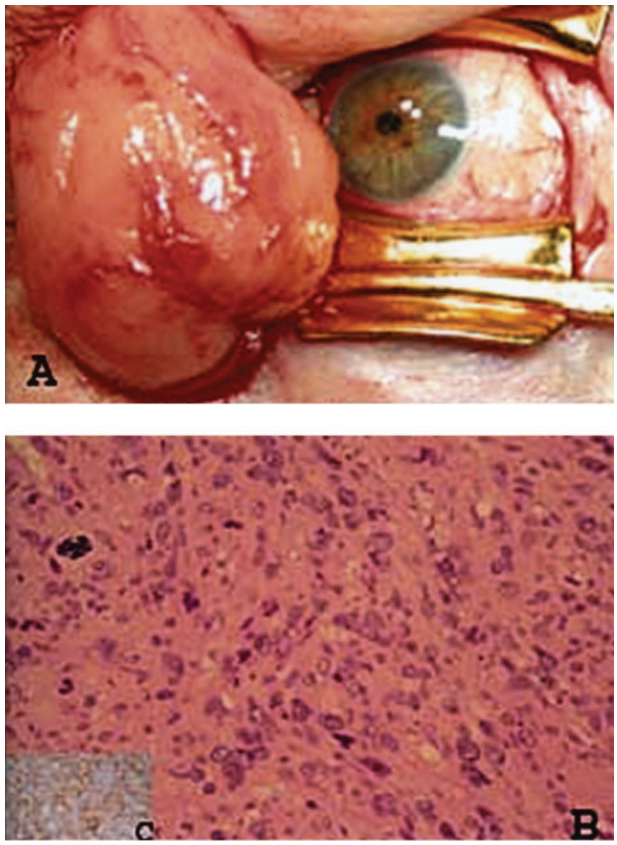

FIG. 3. A, Intraoperative photograph showing the tumor on excision. B, Photomicrograph shows a spindle-cell tumor; marked nuclear pleomorphism and high mitotic rate are noted (hematoxylin and eosin stain, original magnification ×200). C (inset), Tumor cells were immunohistochemically strongly positive for smooth muscle actin but negative for desmin and S100 (magnification ×400).

tation of the MRI appearance of this tumor is lacking. Hou et al.[7] published the MR findings of a primary orbital leiomyosarcoma and showed that the lesion was isointense relative to cerebral cortex on T_1-weighted images, hypointense relative to cerebral cortex on T_2-weighted images, and demonstrated a peripheral rim of moderate contrast enhancement after gadopentetate administration. Our report shows a differing pattern of appearances on MRI in metastatic orbital leiomyosarcoma, with higher T_2 signal intensity and more diffuse contrast enhancement than in primary disease.

B-mode ultrasonography demonstrated the presence of a capsule-like structure surrounding the tumor but could not differentiate the tumor from the medial rectus muscle. An A-scan showed a fairly homogenous internal structure.

Metastatic orbital leiomyosarcoma can be either friable and nonencapsulated[5] or well demarcated.[6] In our patient, the tumor was firm, solid, well-demarcated, and surrounded by a clearly identifiable capsule-like structure that allowed for complete removal of the tumor and restoration of normal ocular motility.

The differential diagnosis of a solid, well-circumscribed intraconal mass includes both benign (peripheral nerve sheath tumors, cavernous hemangiomas, fibrous histiocytomas, hemangiopericytomas, and so forth) and malignant lesions (i.e., metastatic tumors, and lymphomas, although the latter usually do not present as well-circumscribed masses).[8] Although with modern imaging techniques, gross differentiation of these entities is often possible, accurate diagnosis usually requires pathology confirmation.

In conclusion, we have comprehensively detailed the imaging characteristics of metastatic orbital leiomyosarcoma, a rare condition, using complementary ultrasound, CT, and MRI, the combination of which allowed planning for total surgical excision of the lesion.

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Rhabdomyosarcoma Masquerading as Acute Dacryocystitis

Cat Nguyen Burkat, M.D., and Mark J. Lucarelli, M.D.

Abstract: **An 11-year-old boy presented with right medial canthal swelling. He was initially diagnosed with dacryocystitis and treated with oral antibiotics, followed by incision and drainage of a presumed lacrimal sac abscess. Rapid recurrence of the swelling led to further clinical evaluation, including a maxillofacial CT, which revealed an extensive nasal and orbital mass that was consistent with embryonal rhabdomyosarcoma on histopathologic analysis. This represents an unusual case of rhabdomyosarcoma manifesting as acute dacryocystitis. Rhabdomyosarcoma should be considered in the differential diagnosis for acquired nasolacrimal obstruction and dacryocystitis in the pediatric population.**

Rhabdomyosarcoma is the most common soft tissue sarcoma of childhood, accounting for 5% of all pediatric cancers.[1,2] Of the 45% that occur in the head and neck,[3] approximately 25% to 30% arise in the orbit, making orbital rhabdomyosarcoma 10% to 20% of all rhabdomyosarcomas.[2] The average age of onset is 8 to 10 years,[1,4,5] with a male:female ratio of 1.3:1.[3]

The most common manifestation of orbital rhabdomyosarcoma is rapidly progressive proptosis (80% to 100%) over several weeks.[2,4] Downward and outward globe displacement (80%) occurs as a result of the mass located most often in the supranasal orbit.[2,5] Other findings include impaired extraocular motility (42%), ptosis (30% to 50%), eyelid swelling or chemosis (60% to 65%), or a palpable mass (25%).[2,5] Secondary orbital rhabdomyosarcoma occurs by extension from an adjacent sinus or nasopharyngeal space, and patients present with sinusitis, nasal congestion, or epistaxis.

The referring diagnoses of patients with orbital rhabdomyosarcoma have included orbital or preseptal cellulitis, idiopathic orbital inflammation, conjunctivitis, capillary hemangioma, and lymphangioma.[5] We report an atypical case of extensive rhabdomyosarcoma presenting initially as dacryocystitis.

CASE REPORT

An 11-year-old white boy initially presented to another facility with swelling in the right medial canthus. The swelling reportedly began after he was struck in the face by a football. The patient was referred to an ophthalmologist when the swelling did not resolve. He was diagnosed with dacryocystitis and started on amoxicillin and underwent incision and drainage of a nasolacrimal sac abscess the following week.

Within 1 week, the swelling recurred (Fig. 1). Maxillofacial CT revealed an extensive 5.0 × 3.7 × 5.4 cm (AP, transverse, craniocaudal) right nasopharyngeal mass extending to the right ethmoid cavity (Fig. 2, A and B). The mass transgressed the inferomedial orbital wall and displaced the medial and inferior rectilaterally and compressed the globe. Mass effect on the nasolacrimal duct resulted in a dacryocystocele. A biopsy specimen was taken from the nasal mass, with a preliminary diagnosis of lymphoma, and the patient was transferred for further treatment.

At this point, the patient described decreased vision in the right eye. He report no epiphora, photophobia, diplopia, pain on eye movement, dyspnea, or epistaxis. The medical history was significant only for sinusitis.

Visual acuity was 20/80 OD, 20/20 OS. Retinoscopy demonstrated 2.5 diopters of irregular astigmatism on the right, and refraction corrected the vision to 20/30. Pupillary function, extraocular motility, intraocular pressure, funduscopic examination, and visual fields were normal. Right upper eyelid ptosis was present. Exophthalmometry showed 2 mm of right proptosis. A large soft tissue mass, indurated and nontender, was present over the frontal process of the maxilla and nasal dorsum. No purulent drainage was expressed with lacrimal sac compression. Anterior rhinoscopy revealed a large mass in the anterior nasal vestibule, compressing against the nasal septum and nasal floor. There were no ulcerations

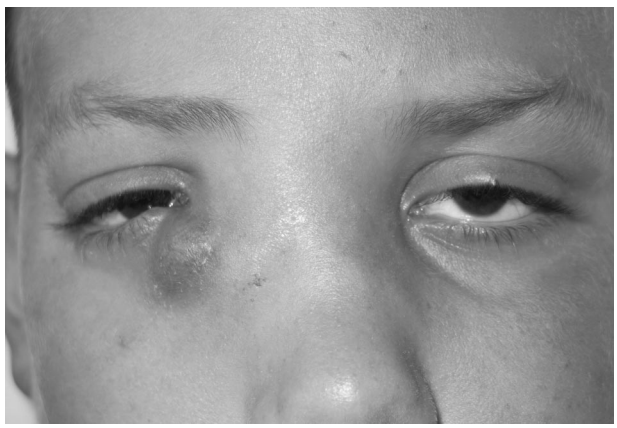

FIG. 1. Recurrence after incision and drainage. Swelling over the nasolacrimal sac in the medial canthus was clinically consistent with dacryocystitis.

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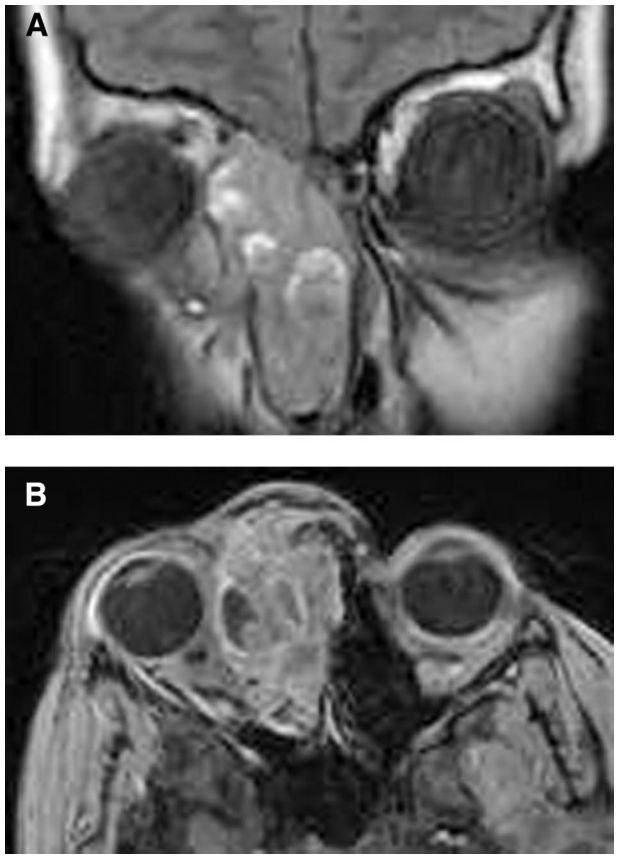

FIG. 2. Maxillofacial CT. **A**, Coronal view: Extensive right-sided nasopharyngeal mass extends into the medial orbit and inferiorly to the floor of the nasal cavity. **B**, Axial view: The globe and medial rectus are displaced laterally by mass effect from the large tumor.

or eschars over the mass. Two enlarged lymph nodes were palpated in the right cervical posterior triangle.

The patient underwent repeat intranasal biopsy of the mass and partial debulking. On histopathology, a highly cellular tumor, comprising small, round cells with hyperchromatic nuclei and dense nucleoli, was consistent with rhabdomyosarcoma, embryonal type (Fig. 3). By immunoperoxidase, the neoplastic cells were positive for desmin and myogenin, which helped identify the myoblastic nature of the tumor.

Systemic evaluation, including a bone marrow biopsy, liver function tests, and chest radiography, was normal. PET demonstrated increased metabolic activity within the tumor mass and cervical lymph nodes. The case was classified as stage III, clinical group III, embryonal rhabdomyosarcoma. Treatment consisting of 42 weeks of chemotherapy and external beam radiotherapy was initiated.

Two weeks after presentation, the patient was readmitted for worsening dacryocystitis and underwent drainage of the nasolacrimal sac abscess and a modified dacryocystorhinostomy. Five weeks later, he returned with recurrent swelling. Endonasal examination revealed complete closure of the dacryocystorhinostomy ostium, and balloon dacryoplasty was performed to reestablish the opening. Over the next 5 months, outpatient visits demonstrated intermittent epiphora and persistence of a quiescent dacryocystocele.

DISCUSSION

Approximately half of orbital rhabdomyosarcomas are located in the retrobulbar space, whereas a smaller number extend in the orbit from the paranasal sinuses, nasal cavity, or pterygopalatine fossa. In those originating in the paranasal cavities, diagnosis may be delayed until ophthalmic signs occur from orbital extension. Acquired nasolacrimal duct obstruction and dacryocystitis may occasionally result from tumors of the nasolacrimal sac,

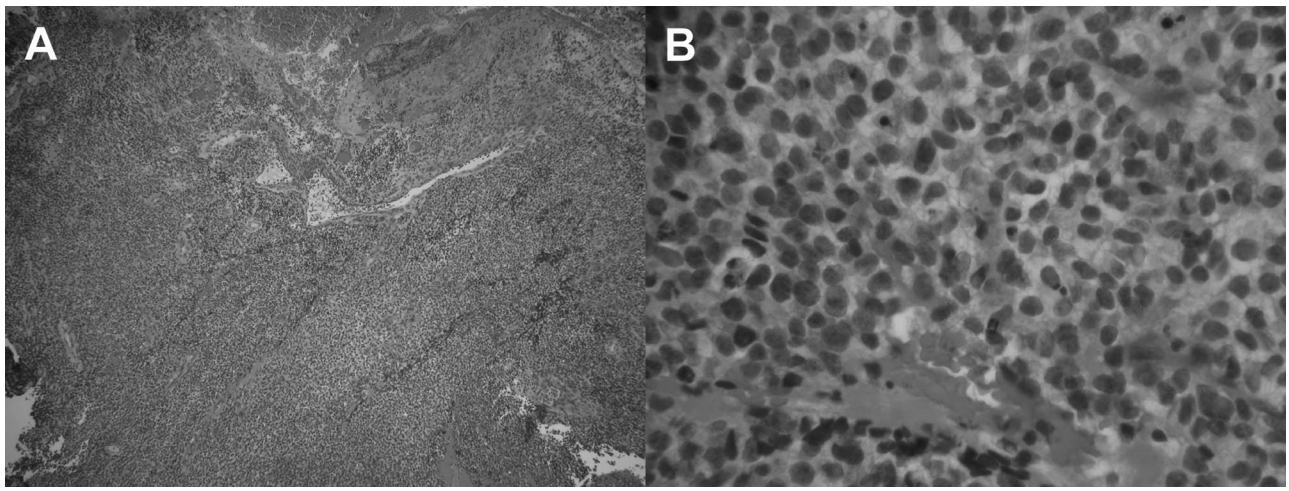

FIG. 3. Biopsy of nasal cavity mass. **A**, Highly cellular tumor composed of sheets of round or spindle cells at various stages of differentiation. Hematoxylin and eosin stain, magnification ×40. **B**, Cells have highly eosinophilic or fibrillary cytoplasm and hyperchromatic nuclei. Hematoxylin and eosin stain, magnification ×400.

inflammatory diseases such as Wegener granulomatosis and sarcoidosis, and mechanical causes[6] such as compression of the excretory system from external tumors.

A Medline search identified one previous report of rhabdomyosarcoma presenting with acquired nasolacrimal duct obstruction.[6] In the report by Baron,[6] the patient presented with epiphora only, and the young age of the patient (5 months) could be suggestive of congenital nasolacrimal duct obstruction. In all patients, it is important to determine if the cause of the nasolacrimal obstruction is congenital or acquired. The older age of the patient presented here warrants a higher suspicion for malignancy rather than congenital obstruction. This patient's age and male sex are also atypical characteristics for acquired nasolacrimal duct obstruction and should therefore necessitate further imaging and nasal endoscopy to facilitate earlier diagnosis of a tumor process. In addition, given the antecedent history of trauma and rapid growth of an isolated tender mass, nodular fasciitis should be considered in the differential diagnosis. To our knowledge, this is the first report of rhabdomyosarcoma manifesting initially as dacryocystitis.

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Orbital Lymphoma With Concomitant Sarcoid-Like Granulomas

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Abstract: **A 41-year-old man presented with chronic eyelid swelling, conjunctival injection, and decreased ocular motility in all gaze directions. MRI showed bilateral enlarged extraocular muscles, including the tendons. Laboratory tests revealed elevated levels of angiotensin-converting enzyme. An orbital biopsy showed collections of monotonous small lymphocytes, and granulomatous inflammation that included multinucleated giant cells, predominantly Langhans type. Flow cytometric analysis of tissue demonstrated a light chain-restricted clonal population of B cells, a finding that confirmed the morphologic impression of lymphoma. This case demonstrates that elevated angiotensin-converting enzyme and granulomatous inflammation can occur in lymphoma. Careful histopathologic examination and flow cytometric analysis are essential to avoid an erroneous diagnosis that could lead to inappropriate management.**

Orbital lymphoma usually presents as painless, insidious proptosis and may be accompanied by eyelid swelling, ptosis, diplopia, and optic neuropathy. Most orbital lymphomas are non-Hodgkin B-cell lymphomas. About 5% of patients with systemic lymphoma will have development of orbital or ocular adnexal involvement. Systemic inflammatory diseases, such as sarcoidosis and vasculitis and other tumors, can present with ocular involvement clinically indistinguishable from orbital lymphoma. Therefore, a definitive diagnosis requires biopsy confirmation that typically involves not only histomorphology but also ancillary studies to demonstrate a monoclonal population of lymphocytes. We present a case of orbital lymphoma that had laboratory and histopathologic features of sarcoidosis. Orbital biopsy showed neoplastic infiltrates accompanied by nonnecrotizing granulomatous inflammation.

CASE REPORT

A 41-year-old man complained of "swollen, red eyes" for 14 months. He had been treated with topical corticosteroids and tetracycline for "ocular rosacea," with no improvement. Two months before presentation, he noticed constant binocular vertical diplopia. His medical history was unremarkable for fever, weight loss, cough, chest pain, or difficulty breathing. He was a nonsmoker, reported no medication use, and his alcohol intake was limited to 2 glasses of wine per day.

Visual acuity was 20/20 OU, and there was no relative afferent pupillary defect. Color vision and red Amsler grid testing were normal. Exophthalmometry was 13 mm OU with a base of 95 mm, and there was marked resistance to retropulsion OU. Significant findings included bilateral ptosis with poor levator function and bilaterally limited gaze in all directions, with a comitant

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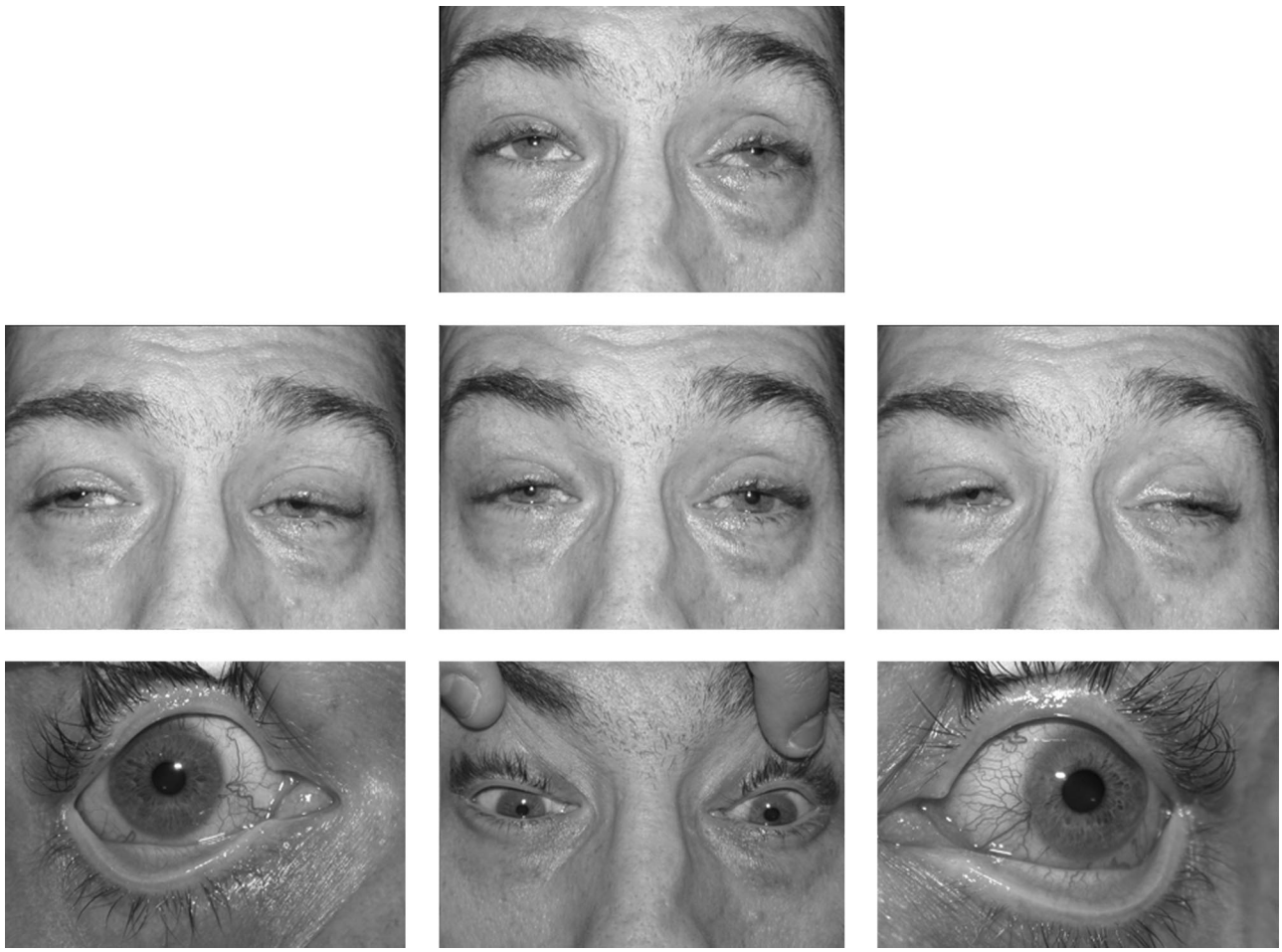

FIG. 1. Bilateral diffuse eyelid edema and limited ocular motility. The conjunctival vessels are engorged, but no subconjunctival lesions are apparent.

right hypertropia of 2 prism diopters (Fig. 1). Confrontation visual fields and fundus examination were normal.

MRI showed bilateral enlargement and enhancement of the medial and lateral rectus muscles, which included the tendinous insertions, with involvement of the Tenon capsule (Fig. 2). Laboratory testing showed mild neutropenia, a low platelet count, a normal thyroid-stimulating hormone level, and no evidence of anti-DS DNA antibody or c-antineutrophil cytoplasmic antibodies. Angiotensin-converting enzyme (ACE) level was elevated at 84 μg/L (normal $<$40).

A left orbital biopsy specimen comprised fibroadipose tissue with dense perivascular collections of monotonous small lymphocytes. In foci adjacent to the lymphocytic infiltrates, poorly formed granulomatous inflammation was present. Multinucleated giant cells, predominantly Langhans type, were seen (Fig. 3). No necrosis was identified. No acid fast bacilli or fungi were seen with special stains. Flow cytometric analysis of fresh tissue showed that 77% of lymphocytes were B cells that were positive for CD19 and CD 20 and were lambda light chain-restricted. There was no significant expression of CD10. These findings were consistent with the morphologic impression of a B-cell lymphoma, probably the extranodal marginal type.

Positron emission tomography with F-18 fluorodeoxyglucose (FDG) demonstrated high uptake in the left orbit and marked splenomegaly and cervical adenopathy without high FDG uptake. A cervical lymph node biopsy also showed a lambda light chain–restricted B-cell neoplasm. The patient was treated with radiotherapy and rituximab. Ten weeks after the orbital biopsy, vision was 20/20 OD and 20/25 OS; ocular motility improved and diplopia lessened.

DISCUSSION

Our patient with orbital lymphoma also had an elevated ACE level and histologic evidence of nonnecrotizing granulomas, which initially led to a clinical suspicion of sarcoidosis. Granulomatous inflammation can accompany both Hodgkin and non-

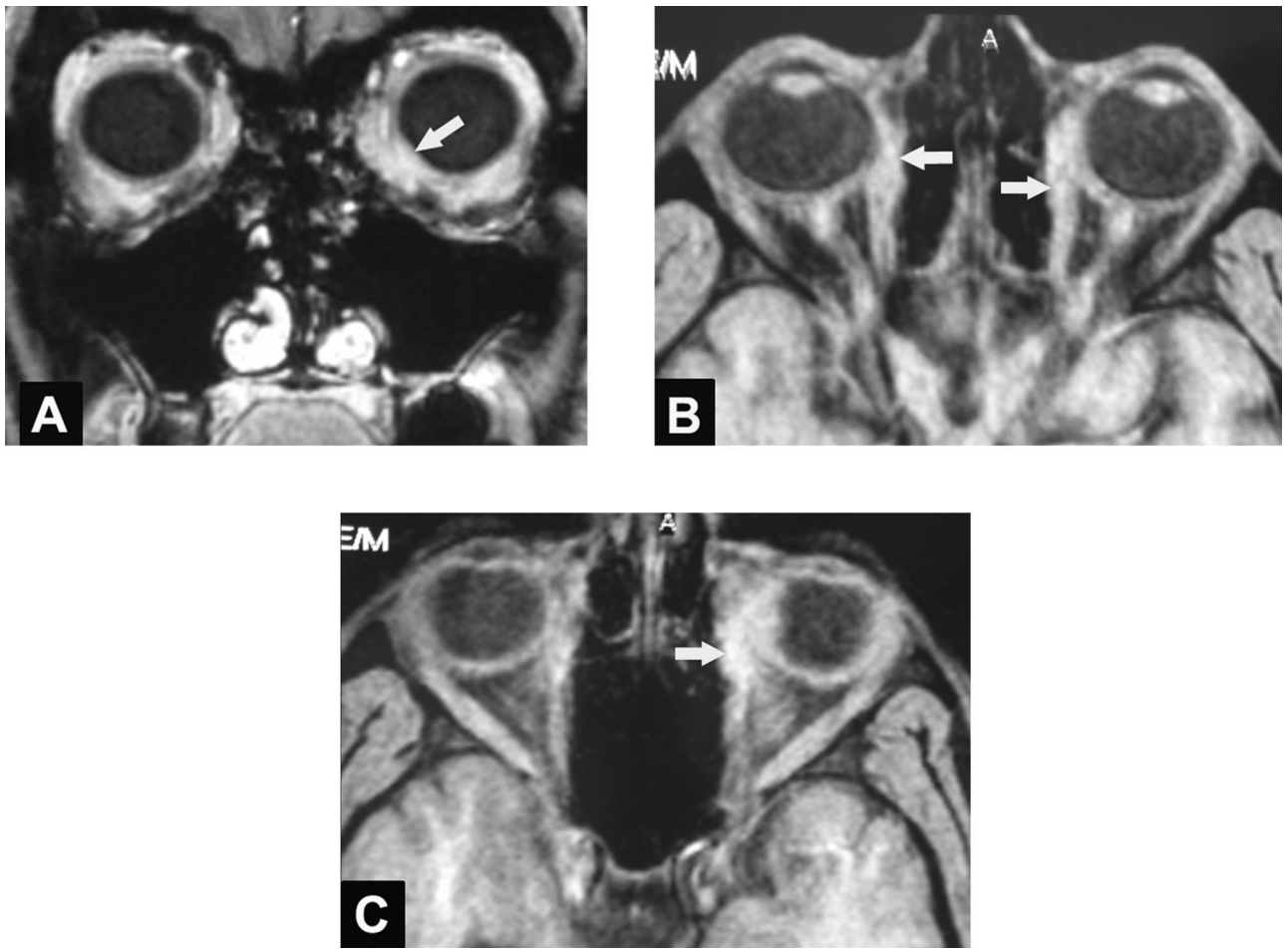

FIG. 2. MRI T_1-weighted images with gadolinium and fat suppression. A, Coronal image shows a bilateral infiltrative process, with mild distortion of the left inferomedial sclera (arrow). B, Axial image shows irregular enlargement of the medial rectus muscles (arrows); Tenon capsule is diffusely thickened. C, Infiltrating process in the anterior orbit is more pronounced on the left side (arrow). The lateral rectus muscles are mildly enlarged.

Hodgkin lymphoma (NHL),[1] with the latter including T- and B-cell neoplasms. The granulomatous reaction may mimic that of sarcoidosis or involve less well-demarcated collections of macrophages (as in the current case). Langhans and the foreign body–type multinucleated giant cells are common and may occur singly or in large numbers.

The pathogenesis of these sarcoid-like reactions may involve a tumor cell–derived, soluble antigenic factor, causing immunologic hypersensitivity and granulomatous inflammation.[2] Some investigators have explored a causal relation between sarcoidosis and lymphoproliferative disorders. In one report, orbital lymphoma developed 15 years after the onset of systemic sarcoidosis.[3] However, biopsy in that report did not show granulomatous inflammation. Another report described the simultaneous presence of conjunctival NHL and pulmonary sarcoidosis.[4] Brincker described a lymphoma-sarcoid syndrome after reviewing 131 cases of sarcoidosis associated with malignancy.[5] He found a high incidence of lymphoproliferative disorders and lung cancer in patients with sarcoidosis.

Similar results were obtained from a retrospective cohort study that compared the cancer incidence between 474 patients with sarcoidosis and 8,541 patients identified in the Swedish Inpatient Register.[6] The investigators concluded that the relative risk for lung cancer and NHL is double that of the general population during the first decade after the diagnosis of sarcoidosis.

These findings were challenged by Seersholm et al.,[7] who followed a cohort of 254 patients with sarcoidosis for 25 years. They found the incidence of malignancy was similar to that of the general population. These conflicting results may be due in part to different criteria used to diagnose sarcoidosis. However, a causal link between sarcoidosis and lymphoma has yet to be confirmed.

This case demonstrates that neither elevated ACE levels nor the presence of granulomas is specific for sarcoidosis. Confirmatory biopsy should be performed in all cases of clinically suspected sarcoidosis. Other con-

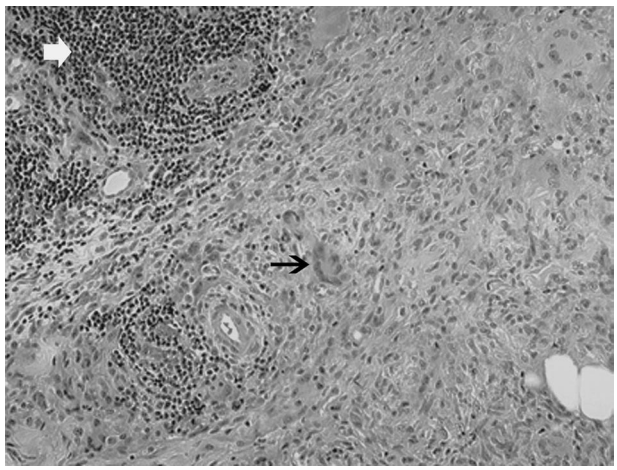

FIG. 3. Neoplastic infiltrate of small atypical lymphocytes surrounds a blood vessel (white arrowhead). The remainder of the field is occupied by a granulomatous inflammatory infiltrate with scattered multinucleated giant cells (black arrow).

ditions such as lymphoma should be ruled out with flow cytometry and immunohistochemical staining.

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Multiple Cavernous Hemangiomas Presenting as Orbital Apex Syndrome

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Abstract: **A 41-year-old man presented with a 7-year history of gradually increasing right proptosis and a 2-year history of decreased vision and limited eye movements. Right proptosis (11 mm), limitation of movement in all gazes, anisocoria, and visual impairment implied orbital apex compression. Computed tomography showed several distinct lesions with mild delayed enhancement extending to the orbital apex. Through a lateral orbitotomy, 10 discrete hemangiomas were removed uneventfully. Histopathologic findings were consistent with cavernous hemangioma in all tumors removed. Proptosis resolved after surgery at 3 to 4 weeks. Visual acuity was unchanged (20/25), and eye movement significantly improved at last follow-up, 15 months after surgery.**

Cavernous hemangioma is the most common benign orbital neoplasm in adults.[1–6] It typically occurs as a slow-growing, well-circumscribed lesion in the lateral and middle intraconal orbit.[2] Multiple unilateral and bilateral cavernous hemangiomas have been reported previously.[3–6] We describe a case of a 41-year-old man with multiple unilateral intraconal orbital cavernous hemangiomas who presented with an orbital apex syndrome. We are unaware of previous reports of this number of intraconal cavernous hemangiomas and presentation of orbital cavernous hemangioma as an orbital apex syndrome and could find no reference to it in a computerized search of Medline.

CASE REPORT

A 41-year-old man was referred to the oculoplastic service complaining of gradually progressive right proptosis for 7 years and decreased vision 2 years before presentation. He also had diplopia and retrobulbar pain for a few weeks. The patient had received multiple courses of oral steroids with the presumptive diagnosis of thyroid eye disease or orbital inflammatory syndrome by other physicians before his referral. Ophthalmic examination revealed right proptosis (11 mm) (Fig. 1), best corrected visual acuity of 20/25 OD and 20/20 OS, and a mild right relative afferent papillary defect. He correctly identified 5 of 12 Ishihara color plates with the right eye. Ocular motility examination showed moderate limitation of motion in all gazes that was associated with diplopia. There was also moderate resistance to retropulsion. Anisocoria (2 mm) was noted and attributed to the oculomotor nerve involvement. Corneal sensation was intact. Intraocular pressure was 26 mm Hg OD and 14 mm Hg OS. Slit-lamp and fundus examinations were otherwise unremarkable. These findings were consistent with an orbital apex syndrome. General physical examinations, routine blood tests, and chest radiography were also within normal limits. Visual field perimetry (central

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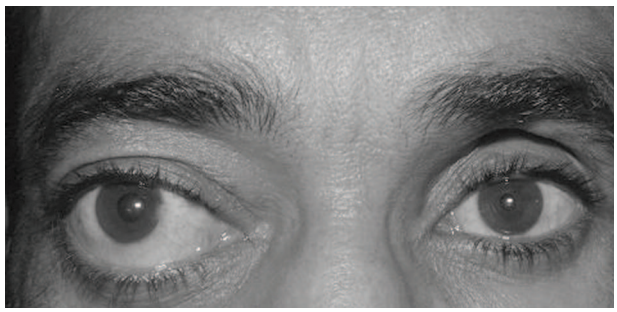

FIG. 1. Right proptosis and anisocoria in a 41-year-old man.

30–2 threshold test, SITA FAST) demonstrated a generalized depression associated with inferior localized defect (Fig. 2, left).

Contrast-enhanced axial CT revealed right proptosis and multiple irregular, discrete intraconal masses with slight enhancement, occupying the right orbit, including the orbital apex (Fig. 2, right). Clinical diagnosis was multiple cavernous hemangioma or schwannoma.

A standard lateral orbitotomy procedure was performed by means of a modified Sattler-Wright incision and bone flap. Ten discrete, encapsulated, red-brown, mulberry-like masses with sizes ranging from 40×30×20 mm to 6×4×3 mm were removed uneventfully from the muscle cone (Fig. 3, left). Three of them were bilobulated (Fig. 3, left; numbers 5, 6, and 9).

Histopathologic examination revealed variable-sized vascular channels filled with red blood cells, which were lined with flat endothelial cells and separated by fibrous connective tissue septae, consistent with cavernous hemangioma (Fig. 3, right). A complete systemic examination performed after surgery was within normal limits.

Visual acuity was unchanged at last follow-up, 15 months after surgery. Color vision (9 of 12 plates) and the relative afferent papillary defect, however, improved. There was less anisocoria (approximately 1 mm) and no proptosis. The patient did not have diplopia within a 30 degree radius of primary gaze. There was not recurrence of tumor at last follow-up.

DISCUSSION

Cavernous hemangioma of the orbit is an encapsulated low-flow hamartoma of the orbit, which has been described as the most common benign orbital neoplasm.[1–6] It typically manifests in the fourth and fifth decades, and 60% to 70% of patients are female.[1–3] The tumor is most often unilateral, but there are few reports of bilateral orbital cavernous hemangiomas.[4,5]

Multiple cavernous hemangiomas of the orbit have been described in 3 previous series of patients with orbital cavernous hemangioma: 1 of 13 cases,[3] 2 of 66 cases,[1] and 5 of 85 cases.[2] A maximum number of 8 discrete multiple orbital cavernous hemangiomas have previously been removed.[5] Our patient had 10 discrete masses removed uneventfully from his left orbital muscle cone. Harris and Jakobiec[1] performed a detailed histologic evaluation of cavernous hemangiomas and showed that this tumor is not a transitional form of capillary hemangioma but an acquired

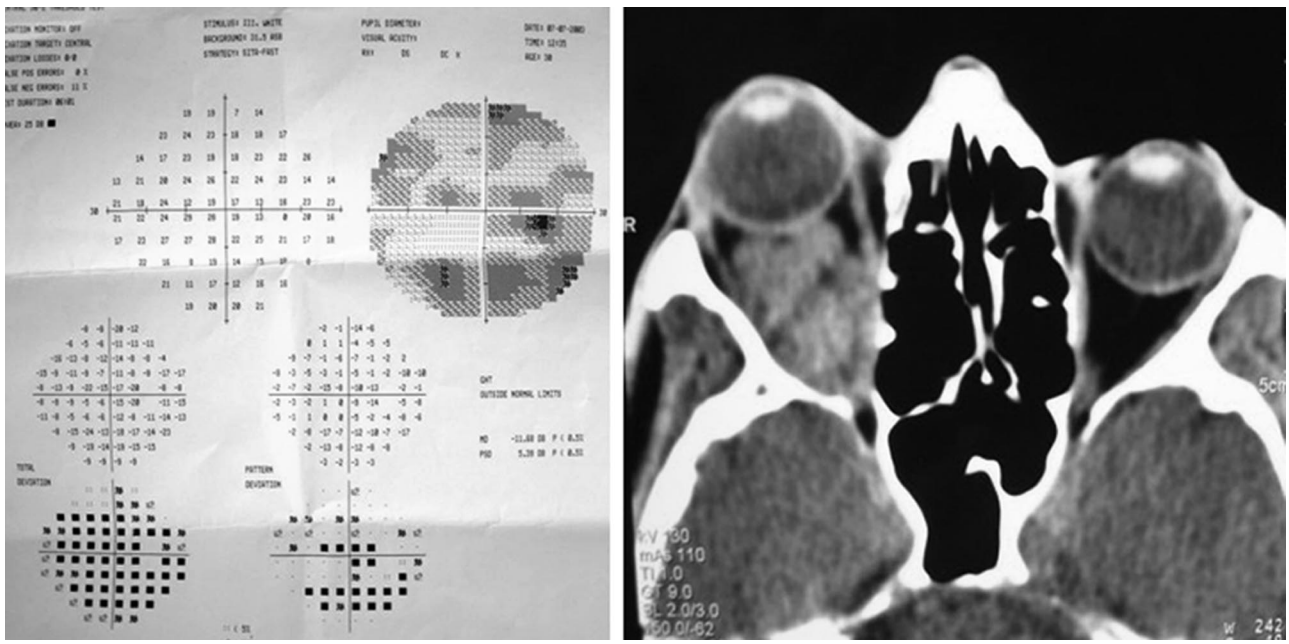

FIG. 2. Left, Visual field (central 30-2 threshold, SITA FAST) shows generalized depression associated with inferior field defect. Right, Contrast-enhanced axial CT of the orbit shows right proptosis and multiple intraconal masses with mild enhancement.

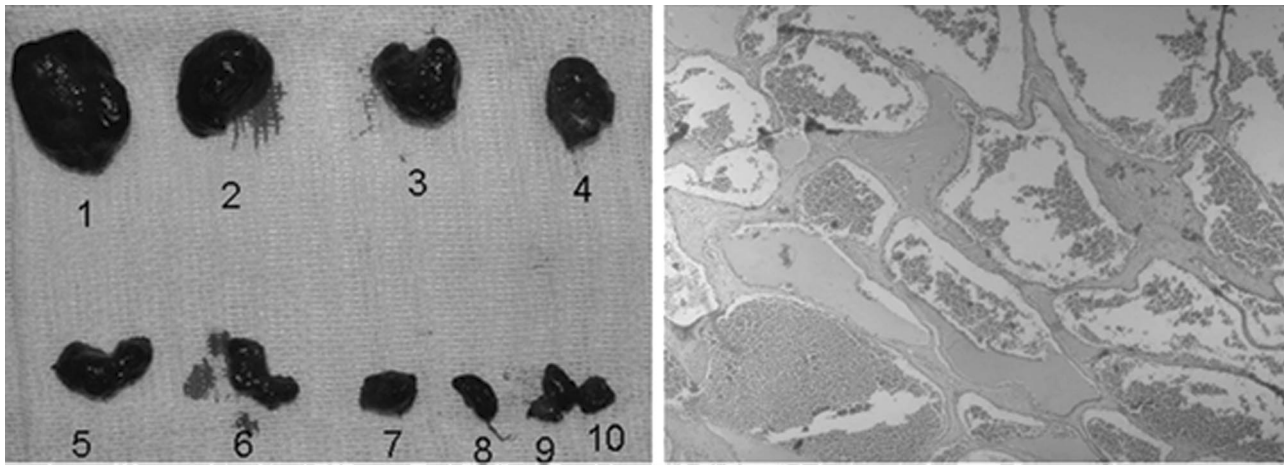

FIG. 3. Left, Ten encapsulated hemangioma-like tumors with variable sizes removed from the right orbit after a lateral orbitotomy. Right, Histopathologic examination shows variable-size vascular channels filled with red blood cells lined by flat endothelial cells and separated by fibrous connective septae, consistent with cavernous hemangioma (hematoxylin and eosin stain, magnification ×10).

lesion. Regional hemodynamic disturbance may be involved in its pathogenesis.[1]

Multiple orbital cavernous hemangiomas may be localized to one orbital cavity.[1–3] They may also involve independent orbital, ocular adnexal, facial, and intracranial structures.[5,7] Bilateral and multiple orbital cavernous hemangiomas have been reported in association with systemic hemangiomas in blue rubber bleb nevus syndrome and Maffucci syndrome.[5,8] No specific pattern of multifocality has necessarily been a definite sign of an underlying gross systemic syndrome. Although neither our patient nor others with multifocal cavernous hemangioma described previously[4,6] had any systemic syndrome, the presence of multiple cavernous hemangiomas should prompt a thorough workup for the possibility of systemic syndromes such as blue rubber bleb nevus syndrome and Maffucci syndrome.[5]

Presenting symptoms could include proptosis, impaired visual function, local pain, diplopia, ptosis, headache, or vertigo.[9] Our patient presented with a combination of severe visual dysfunction, deep orbital pain, ocular motility disturbances, and anisocoria, symptoms suggestive of an orbital apex syndrome. Although restricted extraocular motility and proptosis usually disappear after tumor removal, visual dysfunction is a result of compression of the optic nerve or its vascular supply and therefore may persist after a successful intervention.[9] Fifteen months after successful surgery, our patient had no proptosis, mild extreme upgaze and lateral gaze restriction of ocular motility, less anisocoria, and almost the same visual impairment. Unilateral proptosis must be investigated accurately, and orbital cavernous hemangiomas should be considered in the differential diagnosis of an orbital apex syndrome.

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Giant Cell Reparative Granuloma of the Orbit

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Brendan Lacey, F.R.C.Ophth.‡

Abstract: **This report describes a case of an orbital giant cell reparative granuloma in an 85-year-old woman. Giant cell reparative granulomas are fibro-osseous benign proliferations typically found in the jawbones and rarely in the orbital bones. All previously described cases in the orbit have been in younger patients, classically in the third to fourth decades.**

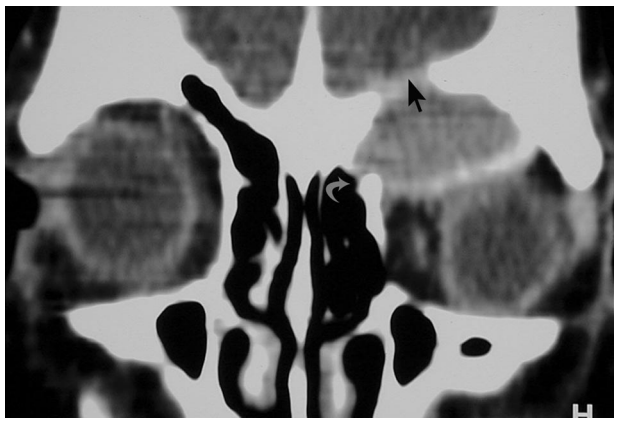

FIG. 1. Coronal CT shows an enhancing soft tissue mass arising in the orbital roof, which has displaced the globe downward. It is associated with bony erosion of the orbital roof superiorly and the medial wall of the orbit (black arrow points to the defect in the upper table of the frontal bone; gray arrow points to the defect in the medial wall).

A giant cell granuloma is a benign proliferative lesion thought to occur as a reparative response to intraosseous hemorrhage. We found 9 published case reports of giant cell granuloma presenting as an orbital mass. All of these cases occurred in patients younger than 40 years of age. To our knowledge, this is the first report of reparative giant cell granuloma of the orbit in an elderly patient.

CASE REPORT

An 85-year-old woman presented with increasing protrusion of her left eye that had developed over a 3-month period without diplopia or reduced vision. She was otherwise healthy, with an unremarkable ocular history. On ophthalmic examination, her unaided Snellen visual acuity was 6/24 OD and 6/18 OS. Fullness of the left upper eyelid associated with an anteroinferior displacement of the left globe was noted. A firm nodular mass was palpated in the supero-medial aspect of the left orbit that did not increase in size with the Valsalva maneuver. No bruit was heard over the mass on auscultation. Hertel exophthalmometry confirmed 7 mm of left proptosis. There was limitation of elevation of the left eye. Anterior segment examination revealed left pseudophakia. Fundus examination was unremarkable. A complete physical examination revealed no abnormality.

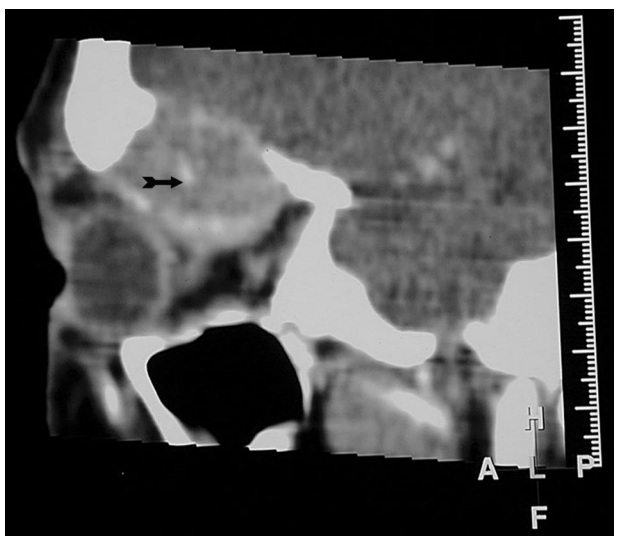

FIG. 2. Sagittal CT shows calcified material around the margins and within the lesion (arrow). This is likely to represent displaced and eroded bone.

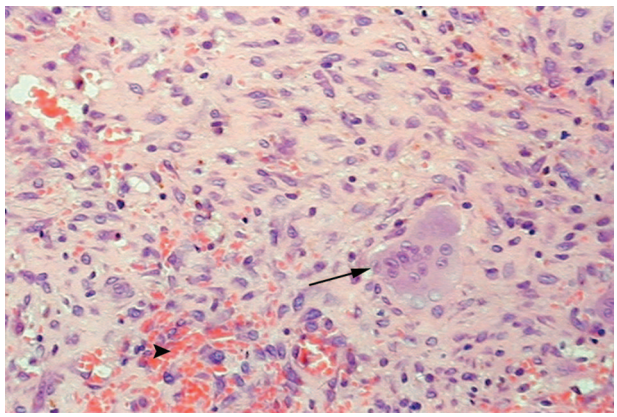

FIG. 3. Histology of the reparative granuloma (hematoxylin and eosin stain, magnification x250). Multinucleated osteoclastic giant cells (arrow) are seen dispersed in a moderately cellular fibrous stroma with areas of interstitial hemorrhage (arrowhead).

Routine hemogram, erythrocyte sedimentation rate, urea and electrolytes, serum-free calcium, and alkaline phosphatase were within normal limits. Her chest radiograph was normal. Orbital CT revealed a mass in the superior orbit separate from the optic nerve that had destroyed the sphenoid, leaving a small fragment near the center (Figs. 1 and 2). T_1- and T_2-weighted MRI confirmed the soft tissue mass to be centered in the region of the left frontal sinus and extending inferiorly in the orbit, superiorly in the floor of the middle cranial fossa; infero-medially it had eroded the medial wall of the orbit. After intravenous contrast, there was marked homogenous enhancement of the mass.

The patient underwent exploration of the left orbit and total tumor excision. The specimen was sent for histology. This revealed it to be a giant cell granuloma (Fig. 3). Eight weeks after surgery, the patient had no complaints, and the proptosis had almost completely resolved. One year after surgery, there was no recurrence.

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DISCUSSION

The term giant cell reparative granuloma was first used by Jaffe in 1953 to describe a benign lesion found in the jawbone that was thought to represent a reparative response to intraosseous hemorrhage.[1] Typically, these lesions develop in the jawbone and have been most commonly described in the mandible. However, there are case reports describing other locations such as maxilla, sphenoid, and the ethmoid bones.[2–4] Giant cell reparative granuloma in the orbital bones is rare. The first reported case was by Sood et al.[4] in 1967. All reported cases of orbital giant cell reparative granuloma have involved patients younger than 40 years of age.

Although a giant cell granuloma is thought to be a granulomatous reparative response to intraosseous hemorrhage that may be caused by trauma and chronic inflammation in the paranasal sinuses, a history of trauma or chronic sinusitis has never been consistently elucidated in patients with this lesion.[2,5] These lesions, though benign, can be locally aggressive and result in significant bone destruction.

On gross inspection, the tumor appears to consist of red or reddish-blue friable vascular tissue.[6] On histological examination, the stroma appears as a moderately cellular fibrous stroma. There is evidence of interstitial hemorrhage, and the osteoclastic multinucleated giant cells are seen dispersed in the stroma. Foci of osteoid and new bone formation also may be seen.

Other lesions must be considered in the differential diagnosis of reparative giant cell granuloma, including brown tumor of hyperparathyroidism, giant cell tumor, and aneurysmal bone cyst. The brown tumor of hyperparathyroidism is histologically almost identical to the reparative giant cell granuloma.[7] Features that differentiate the two entities are raised free calcium and reduced phosphorus levels in patients with brown tumor.

The treatment of these lesions is surgical.[6] Recurrence in 10% to 15% of cases may follow incomplete excision.[5,7] Radiation therapy has been used for inoperable cases. However, there is a risk of carcinomatous or sarcomatous change after this modality of treatment.[5]

Giant cell reparative granuloma, although rare in older patients, should be considered in the differential diagnosis of fibro-osseous lesions occurring in the orbit or other skull bones.

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Severe Subcutaneous Emphysema Following Orbital Blowout Fracture

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Abstract: **A 19-year-old man sustained a blowout fracture to the left orbital floor. Subcutaneous emphysema developed after a protracted sneezing episode. Some signs of respiratory distress ensued, and the patient was emergently intubated. He underwent surgical repair of the fracture and then had further extension of the subcutaneous emphysema. The left nasal cavity was packed and the emphysema slowly resolved. Clinically significant subcutaneous emphysema is an uncommon complication of orbital blowout fracture. Informing the patient to avoid excessive nose-blowing and to avoid occluding the nose while sneezing may prevent this complication.**

CASE REPORT

A 19-year-old incarcerated man with an extensive psychiatric history was involved in an altercation. He was hit with the blunt end of a broomstick to the left eye. He was evaluated several hours after the incident and was found to have 2 mm of enophthalmos, left infraorbital hypesthesia, and mild subconjunctival hemorrhage. Moderate swelling and ecchymoses were noted in the left malar area. The patient was orthophoric in primary and downward gaze. He was diplopic in upgaze.

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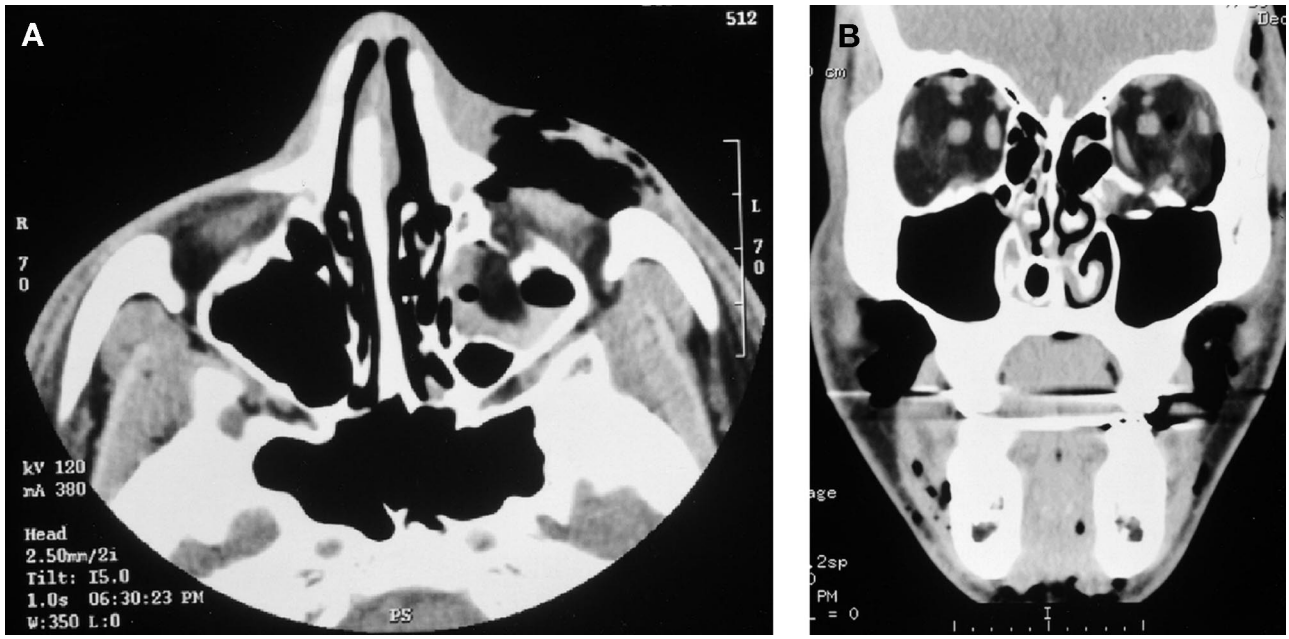

FIG. 1. A, Orbital CT shows extensive air within the left malar soft tissues and a fracture involving the roof of the maxillary sinus. B, Coronal view of left orbital floor blowout fracture. A small collection of air is visible within the inferolateral left orbit.

A CT scan performed to confirm the clinical suspicion of an orbital fracture showed an isolated blowout fracture of the orbital floor with prolapse of orbital fat into the left maxillary sinus (Fig. 1, A and B). Marked swelling and emphysema of the preseptal periorbital soft tissue was also present. The patient was treated with prophylactic oral antibiotics and oxymetazoline nasal spray and was told to avoid nose-blowing and strenuous activity. Elective surgical repair of the orbital fracture within 10 days was recommended. He was discharged back to the corrections facility in stable condition.

The following day, the patient returned, complaining of facial, neck, and chest swelling that made breathing difficult. He stated that his breathing difficulty began shortly after his discharge from the hospital the previous evening. Further questioning revealed that he had several sneezing episodes. When he had attempted to avert the sneeze, he pinched his nostrils to avoid blowing out any nasal contents. He reported no other trauma since the broomstick injury.

On examination, the patient was alert, oriented, and appeared anxious. Heart rate was 75 beats per minute, blood pressure was 122/75, respiratory rate was 28 breaths per minute, and pulse oximetry was 90% or above during the examination. Exophthalmometry readings were unchanged from the previous measurement, 20 mm on the right and 18 mm on the left. Soft tissue swelling with crepitus was noted from the mid face and neck to his anterior chest bilaterally. The furthest extent was measured approximately to the level of his nipples bilaterally. Auscultation of his chest revealed no wheezes, rales, or rhonchi.

An emergent chest radiograph showed normally inflated lungs, no pneumothorax, and no parenchymal disease. There was a significant amount of air in the subcutaneous tissues of the anterior neck and chest region. The emphysema noted on radiography corresponded to the crepitus palpated in his neck and anterior chest wall. The patient underwent emergent otolaryngology consultation, including evaluation under anesthesia to determine airway status. Fiberoptic evaluation of the upper airway revealed no obvious upper airway compromise. The otolaryngologist elected to intubate the patient to prevent airway obstruction from further extension of the emphysema (Fig. 2). Chest CT revealed emphysema within the

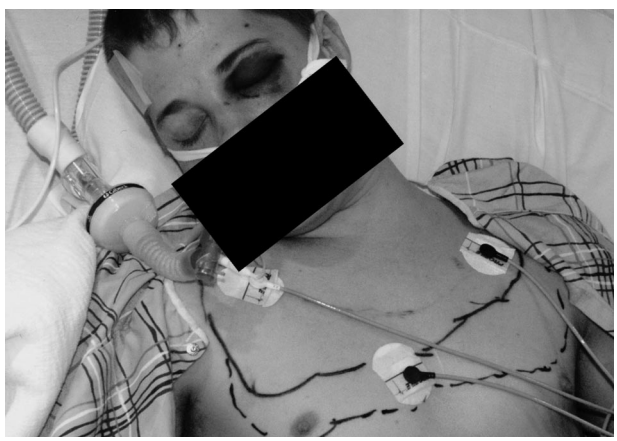

FIG. 2. Patient with left orbital blowout fracture is intubated owing to respiratory distress. Crepitus from subcutaneous air was present in the areas shown marked on the patient's chest.

superficial muscle planes and subcutaneous tissue of the neck, shoulders, and anterior chest. There was no emphysema noted within any of the deep tissue. Mediastinal enlargement was not apparent. The patient was admitted to the intensive care unit for further observation.

On hospital day 2, the patient underwent uncomplicated repair of the floor fracture. The left orbital floor was approached through a transconjunctival approach. The fracture was confined to the floor and measured 7 mm horizontally and 20 mm anteroposteriorly. The inferior orbital rim was intact. The herniated orbital contents were reposited before positioning of a bioresorbable polymer implant (Lactasorb, Lorenz Surgical, Jacksonville, FL) implant over the entire extent of the fracture. The lower eyelid retractors and conjunctiva were closed without difficulty. The patient remained intubated after the repair and was transferred back to the ICU for further observation and management.

Twenty-four hours after the fracture repair, the subcutaneous emphysema regressed and the patient's respiratory status remained stable. He was then extubated. That evening he had another sneezing episode, which resulted in recurrent extension of the subcutaneous emphysema. The patient was anxious but showed no signs of respiratory distress. His left middle meatus was packed with ribbon gauze. The subcutaneous emphysema completely cleared within 1 week of nasal packing.

DISCUSSION

Localized orbital emphysema associated with orbital fracture is seen routinely with CT. Patients rarely have complications from extravasation of modest amounts of air in surrounding tissues. Most occurrences are self-limited and resolve without treatment.[1–4] Severe orbital emphysema may cause acute proptosis, high intraocular pressure, and central retinal artery occlusion. One known risk factor for orbital emphysema is nose-blowing, and thus patients are advised to avoid it.

In this case, a disruption in the periosteum of the maxillary sinus allowed air entry in the orbital and subcutaneous soft tissues. Presumably, a breach in the orbital septum allowed passage of orbital emphysema into the subcutaneous space. The facial subcutaneous plane was expanded by the positive pressure from the maxillary sinus. This facial plane is contiguous with the subcutaneous plane of the neck and chest region. Delayed periorbital emphysema in the setting of an isolated medial wall fracture has been reported after sneezing and after nose-blowing.[2] Unusual cases of cervical emphysema, mediastinum, and pneumothorax after self-induced sublingual injuries and Valsalva maneuver further emphasize the extensive potential communications for air extravasation in the setting of facial injuries.[5] A case of extensive periorbital emphysema after laparoscopic nephrectomy[6] demonstrated communication between the subcutaneous tissues of the periorbita and face with more distant sites of the body.

Physicians should inform patients with orbital fractures adjacent to sinuses that if sneezing occurs, occluding the nose should be avoided because this could force air from the sinuses into the orbital or subcutaneous space.

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Cranio-Orbital Injury With Internal Carotid Artery Laceration and a Missing Eyelid

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Abstract: **A 37-year-old man presented to the emergency department with a visor impacted in the left orbit after a motorbike accident. His initial injury appeared to be limited to the orbit. Because the bleeding appeared disproportionate to the orbital injury, an intracranial injury was suspected. Subsequently, on hemicraniotomy, the tip of the visor was found at the bifurcation of the internal carotid artery. The avulsed upper eyelid was found under the frontal lobe. The patient underwent the clipping of internal carotid artery and middle and anterior cerebral artery with replantation of the upper eyelid. Neurologic damage was limited to loss of vision, right foot drop, and bowel and bladder incontinence. This case of penetrating orbital trauma demonstrates that the absence of focal neurologic deficit and radiologic signs suggestive of intracranial injury may not rule out life-threatening intracranial injury.**

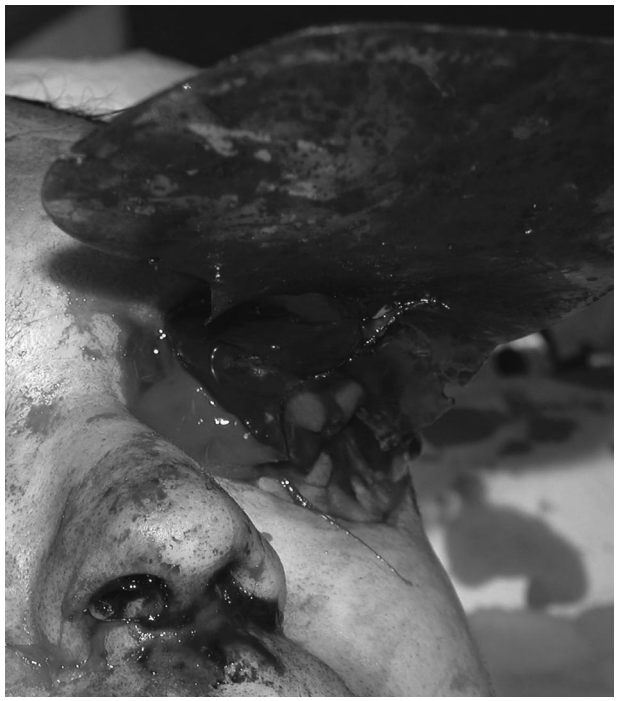

FIG. 1. Left penetrating orbital trauma with impacted helmet visor.

Penetrating orbital foreign bodies may be associated with extensive injury to the brain. Clinical examination and CT may sometimes fail to reveal the full extent of injury. We report a case of penetrating orbital trauma in which the damage appeared to be limited to the orbit but was subsequently found to be a significant life-threatening intracranial injury.

CASE REPORT

A 37-year-old man was airlifted to the emergency department after a motorbike accident. He presented with an impacted visor in the left orbit (Fig. 1). The left upper eyelid was missing. Profuse bleeding from the orbit hindered assessment of the full extent of the injury. Emergency CT of the orbits and brain was arranged. The patient was continuing to lose a significant amount of blood, and the anesthetic team struggled to steady his hemodynamic status and keep him relatively still for the scan. The CT scan was of poor quality as the result of patient motion and provided limited information regarding the extent of visor penetration and the status of the orbital roof. The neurosurgical opinion was that the injury was limited to the orbit, as the patient did not have any apparent focal neurologic deficit.

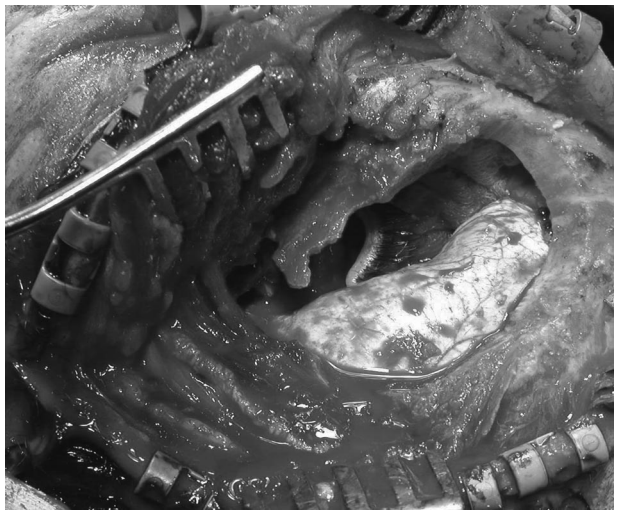

FIG. 2. Surgeon's view of the craniotomy site showing avulsed upper eyelid under the frontal lobe.

The ophthalmic surgeon, on exploration of the left orbit, noticed yellowish-white tissue prolapsing from the superior and medial part of the orbit, which resembled orbital fat. Further exploration revealed that the orbital roof was breached, with the visor firmly impacted in it. The tissue initially thought to be fat was in fact brain tissue. Disproportionate bleeding and a finding of brain tissue in the orbit raised suspicion of damage to a major intracranial vessel such as the internal carotid artery (ICA). The neurosurgeons initially tried to arrest the bleeding by ligating the left common carotid artery but were unsuccessful. The patient then underwent a left hemicraniotomy. The tip of the visor was lodged at the bifurcation of the ICA. The missing eyelid was in the anterior cranial fossa under the frontal lobe (Fig. 2). Clipping the ICA and anterior and middle cerebral arteries arrested the bleeding. The visor was removed from the orbital side. The avulsed eyelid was replanted. Despite the severe nature of the injury, the globe was intact. As expected, there was no vision in the left eye because of ICA ligation. After surgery, there was persistent cerebrospinal fluid leak from the damaged roof. Neurosurgical opinion was against the use of foreign or autologous material for roof repair because of concerns of infection. Because the left eye had no vision, an eyelid-sparing anterior orbital exenteration was recommended. It was hoped that the skin flap lining the socket would reduce the risk of infection of the exteriorized brain tissue.

Despite clipping the major vessels on the left side of the brain, the neurologic damage was limited to right foot drop and bowel and bladder incontinence.

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DISCUSSION

It is not always possible to assess the full extent of a cranio-orbital injury at presentation. The presence of any orbital foreign body in the orbit must be viewed with suspicion, and appropriate steps must be taken to rule out intracranial extension. Some materials may not appear on conventional imaging, and appropriate neuro-imaging is extremely difficult with multiple cranio-facial injuries.[1] Because the ophthalmologist is often the first clinician to evaluate a patient with a penetrating orbital injury, it is important for us to have a high index of suspicion of intracranial involvement.[2] The bleeding is rarely severe enough to cause concern in an injury limited to the orbit. Presence of focal neurologic deficit is a helpful clinical sign suggestive of intracranial trauma. However, in the presence of good collateral circulation, it may not always be present. In our case, the missing eyelid was retrieved from the undersurface of the frontal lobe of the brain. The possibility of an ectopic location of the eyelid must be considered in traumatic eyelid avulsion. The surgical reattachment of the eyelid often may be successful even if delayed.[3] A multidisciplinary approach to the management may be required because facial and cerebral injuries are often associated with penetrating orbital injury.[4]

Absence of focal neurologic deficit or radiologic signs suggestive of intracranial injury may not necessarily rule out life-threatening intracranial injury.

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Use of an Electromagnet to Retrieve a Broken Fascia Needle During Frontalis Sling Surgery

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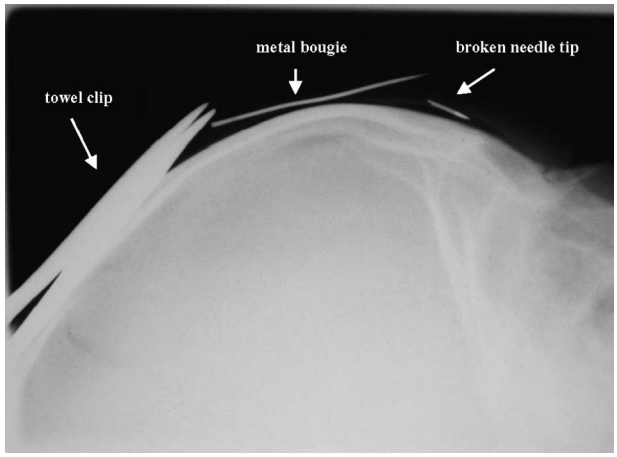

FIG. 1. Radiograph of broken needle tip in situ. Metal bougie was used as a marker.

Abstract: **Frontalis sling surgery was performed on a 2-year-old girl with congenital ptosis. During surgery, the tip of the Wright fascia needle broke off as it was being passed between incisions within the eyebrow. The tip was difficult to locate and retrieve through the original incision and was finally located with an electromagnet and removed through an enlarged incision.**

Congenital ptosis may be associated with the development of amblyopia. Surgery in children who are at risk is usually indicated when the ptotic eyelid obscures the pupillary axis.[1] Frontalis suspension is often the procedure of choice and, depending on the age of the child, either synthetic material or autogenous fascia lata may be used as a sling. Complications of frontalis sling surgery are well documented,[2] yet we encountered an unusual complication that has been reported once before in the literature.[3] During surgery on a young girl, the tip of the Wright fascia needle broke off as it was being

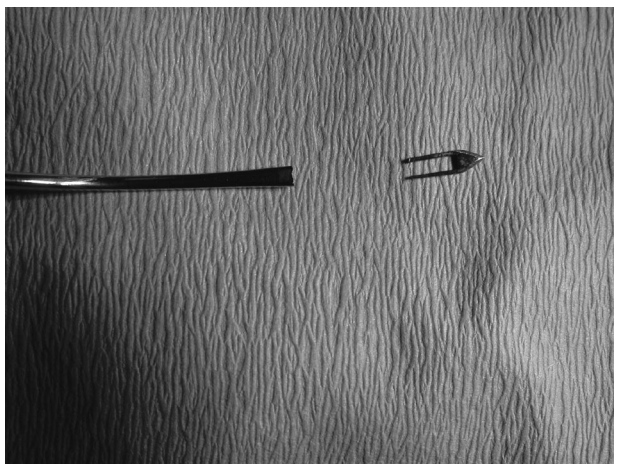

FIG. 2. Broken Wright fascia needle and retrieved needle tip.

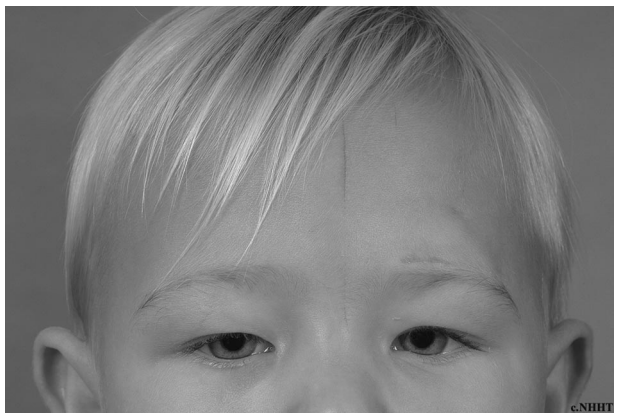

FIG. 3. Patient 2 months after surgery.

passed between incisions in the brow. We describe the difficulties encountered when trying to locate and retrieve the broken tip and how they were overcome by using an electromagnet.

CASE REPORT

A 9-month-old girl was referred with a left congenital ptosis. When she was 18 months old, it was clear that the ptotic eyelid covered the pupil much of the time and she was therefore at risk of developing stimulus deprivation amblyopia. Three months later, she underwent unilateral frontalis suspension through the use of the Fox pentagon technique but had infection of the silicone sling after surgery. The infection failed to resolve with antibiotic treatment and the sling was removed. Although there are reports of sling removal without recurrence of ptosis,[4] this patient's ptosis recurred immediately. Four months after the first operation, she underwent repeat silicone frontalis suspension, using the original Fox pentagon incisions. During the procedure, as the Wright fascia needle was passed from the apical incision to the medial eyebrow incision, the tip of the needle broke off. Attempts to locate and retrieve the tip with forceps were unsuccessful. A radiograph, using anteroposterior and lateral views, was taken at surgery to pinpoint the position of the tip (Fig. 1). Location of the metallic fragment continued to be difficult. An electromagnet, usually used for the removal of intraocular foreign bodies, was then used. By holding the electromagnet just above the skin and using intermittent power, the needle tip was easily located by observing the movement of the overlying skin. The original incision had to be enlarged only slightly, and the tip was successfully retrieved (Fig. 2). The operation was then completed with another Wright fascia needle. Postoperative recovery was uneventful, and a good functional and cosmetic result was achieved (Fig. 3).

DISCUSSION

This unusual complication of frontalis sling surgery has been reported only once before in the literature.[3] Whereas this case was successfully managed by extending the skin incision and using blunt dissection to explore under the orbicularis muscle, our case required the novel use of an electromagnet to locate the broken needle tip. Magnetic retrieval of intraocular foreign bodies is a widely recognized technique, yet the use of magnets to locate and remove broken surgical instruments has only previously been reported in other specialty journals.[5,6]

The Wright fascia needle in this case report had been used on five previous occasions, and it was thought that either metal fatigue or a manufacturing fault were responsible for the breakage. A second Wright fascia needle was readily available, and surgery was completed successfully once the broken needle tip had been removed.

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