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ORIGINAL ARTICLE

Orbital hemangiopericytoma in an Asian population

Chi-Hsin Hsu [a,b], Yi-Hsuan Wei [a], Yeh Peng [a,c], Shu-Lang Liao [a,*]

[a] *Department of Ophthalmology, School of Medicine, National Taiwan University Hospital, Taipei, Taiwan*
[b] *Department of Ophthalmology, Shuang Ho Hospital, Taipei Medical University, New Taipei City, Taiwan*
[c] *Department of Pathology, National Taiwan University Hospital, Taipei, Taiwan*

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Background/Purpose: Hemangiopericytoma is a very rare orbital tumor. The purpose of this study was to report the clinical and histopathological features of six cases of orbital hemangiopericytoma in an Asian population.
Methods: Clinical and histopathological features were reviewed in six patients who were histopathologically confirmed as having primary orbital hemangiopericytoma in National Taiwan University Hospital between May 2001 and December 2010.
Results: Among the six cases who were diagnosed as having primary orbital hemangiopericytoma, all lesions were reported as vascular tumors and featured branching "staghorn appearance" vessels. All patients, including one male and five females, presented with progressive proptosis and some associated symptoms such as extraocular motility limitation with diplopia, displacement of the globe, afferent pupillary defect, congested vessels of conjunctiva, or decreased visual acuity. On computed tomography, the orbital tumors tended to manifest as circumscribed masses with homogeneous medium-to-high enhancement with contrast studies. All six patients received surgical treatments, and four of them had additional radiotherapy. Three patients had recurrence after surgeries, and one of them had multiple metastases to lung and liver. All patients were still alive after a follow-up period of 5–10 years.
Conclusion: Orbital hemangiopericytoma has malignant potential, which may lead to local recurrence and/or metastasis. Histopathological findings alone are insufficient to predict

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* Corresponding author. Department of Ophthalmology, National Taiwan University Hospital, 7, Chung Shan South Road, Taipei 100, Taiwan.
E-mail address: liaosl89@ntu.edu.tw (S.-L. Liao).

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the behavior of this tumor. Therefore, both clinical and histopathological findings are important to evaluate the treatment outcomes. Total excision accompanied with radiotherapy is suggested and long-term follow-up is required.

Introduction

Hemangiopericytoma, first described in 1942 as a distinct vascular tumor of soft tissue, is a rare mesenchymal neoplasm originating from the pericytes of blood vessels.[1] Pericytes are mesodermal cells surrounding small blood vessels; therefore, this tumor may develop wherever capillaries are present. Although the incidence of this rare tumor in the head and neck region is 16%, hemangiopericytomas originating in the orbits account for less than 5%.[2] In one of Shields's series, the incidence of this tumor accounts for only 1% of all orbital biopsies and 4% of all orbital vascular tumors.[3]

Primary orbital hemangiopericytoma is a well-defined, slowly growing mass. B-scan ultrasonography usually shows this tumor as a well-delineated round or oval lesion with low to medium internal reflectivity; the tumor is mostly irregular, with solid and cystic components.[4] On computed tomography (CT) imaging, it is seen as a well-circumscribed mass occupying either the extraconal or the intraconal space, which intensely enhances with contrast.[5,6] The definite diagnosis of hemangiopericytoma must depend on pathological examination of the surgical specimen. The largest series of this orbital neoplasm, a report of 30 cases, was conducted by Croxatto and Font in 1982.[5] However, within the past 15 years, the largest study, reported in 1997 by Karcioglu et al,[6] included only seven patients. In Asia, only a few single-case reports have ever been published.[7,8] In this study, we present our experience with six patients diagnosed with unilateral orbital hemangiopericytomas within the last 10 years.

Patients and methods

This retrospective study included six patients histopathologically diagnosed as having orbital hemangiopericytomas in National Taiwan University Hospital between May 2001 and December 2010. All lesions were reported as vascular tumors with featured branching "staghorn appearance" vessels. There were five females and one male, aged from 30 to 62 years, with a median of 44. Three patients had right orbital involvement, while the others had left orbital involvement. The duration of symptoms prior to initial visit ranged from 5 months to 10 years, and the follow-up period ranged from 5 to 10 years. Clinical data, and radiologic and histopathological examinations were reviewed. Special attention was paid to treatment modalities and clinical outcomes after long-term follow-up. Local administrative and ethics committees approved all aspects of this study.

Results

Six patients, five females (aged 30—62 years; median 41 years) and one male (aged 53 years), were included in this study. Initial presentations varied: two patients (Cases 3 and 4) presented as having palpable masses on their eyelids, one patient (Case 1) as having eyelid swelling, one patient (Case 5) as having eye protrusion, and two patients (Cases 2 and 6) complained loss of vision with best-corrected visual acuity around counting fingers (CFs) 20 cm (Case 2) to light perception (LP) (Case 6) (Table 1).

However, under thorough ophthalmic examinations, all patients had unilateral proptosis ranging from 3 to 7 mm; five had different degrees of extraocular muscle limitation (Case 2—6) and therefore complained of double vision (Cases 2—5). Blurring or loss of vision was an associated symptom in four patients (Cases 2, 4—6), and visual acuity was severely decreased to CF or LP level for three of them (Case 2: CF 20 cm, Case 4: CF 50 cm, and Case 6: LP). Visible protruding masses with conjunctival engorged vessels were found in two patients (Cases 3 and 6) (Table 1). All patients except one who had severe displacement of globe and marked strabismus (Case 4) (Fig. 1A) underwent indirect ophthalmoscopy examination. A pigmented, mass-like lesion was noted on the retina in Case 2. Papilloedema and retinal folds on the temporal retina were evident on the fundus examination in Case 5, who had the poorest visual acuity. In addition, these two patients presented anisocoria without any evidence of pupillary light reflex on the affected eye.

The results of imaging studies are shown in Table 1. Ultrasonography was performed only on the first two patients (Cases 1 and 2): one of the tumors showed low-to-moderate echogenicity (Case 2) (Fig. 2), while the other revealed moderate-to-high echogenicity (Case 1). Initial CT for all patients depicted orbital masses, and four of them were superiorly located (Cases 1—3, 5) (Fig. 3A). Homogeneous orbital mass lesion was noted in three cases (Cases 1—3), but the other three lesions were heterogeneous (Cases 4—6) (Figs. 1B and 3A). The tumors were all well enhanced with contrast medium. Neither the central nervous system extension nor any bony change has been found in all patients.

Differential diagnoses varied regarding these six tumors after the initial clinical examinations and image studies (Table 1). The diagnoses included hemangioma, lymphoma, meningioma, pseudotumor, rhabdomyosarcoma, and glioma. Hemangiopericytoma was considered among the initial list of differential diagnoses only in one case even after CT was completed.

Histopathological and immunochemical findings are shown in Table 2. All lesions were reported as vascular tumors and featured branching "staghorn appearance" vessels (Figs. 3B and 4A). The proliferating pericytes in four of the specimens had benign-looking patterns: two (Cases 1 and 2) were described as "bland cells" and other two (Cases 3 and 5) as "with low to occasional mitotic rates." Only the specimen for Patient 4 showing occasional necrosis and the specimen for Patient 6 demonstrating frequent mitotic

Table 1 Summary of clinical and imaging findings.

No.	Age, sex, orbit	Duration before diagnosis	Initial presentation	Initial BCVA (R, L)	Associated symptoms	Computed tomography	Initial differential diagnosis
1	30, F, R	2 y	Upper eyelid swelling	20/20, 20/20	Eye protrusion	→ Location: anterosuperior → Enlargement of lacrimal gland	Lacrimal gland tumor
2	50, F, R	10 y	Loss of vision	CF 20 cm, 20/30	Eye protrusion Double vision	→ Location: retrobulbar, posterosuperior → Round, homogeneous, well-enhanced mass; eyeball displacement	Meningioma Glioma Hemangioma Lymphoma
3	41, F, L	5 mo	Upper eyelid mass	20/30, 20/25	Eye protrusion Double vision Subconjunctival mass Conjunctival redness	→ Location: superior → Homogeneous extraconal mass, displacement of the superior rectus muscle and eyeball	Lymphoma Lymphangioma Pseudotumor
4	62, F, R	6 mo	Lower eyelid mass	CF 50 cm, 20/30	Double vision Blurred vision Eye protrusion	→ Location: retrobulbar, inferior → Heterogeneous, well-enhanced mass; eyeball displacement; optic nerve involvement	Hemangioma Meningioma Hemangiopericytoma
5	30, F, L	6 mo	Eye protrusion	20/20, 20/60	Double vision Blurred vision	→ Location: medial-superior → Intraconal well-enhanced mass; eyeball is compressed	Hemangioma
6	50, M, L	6 mo	Loss of vision	20/30, LP	Eye protrusion Conjunctival redness	→ Location: retrobulbar → Heterogeneous intraconal mass, eyeball displacement, medial rectus and optic nerve involvement	Hemangioma Pseudotumor Rhabdomyosarcoma

BCVA = best-corrected visual acuity; CF = counting finger; F = female; L = left; LP = light perception; M = male; R = right.

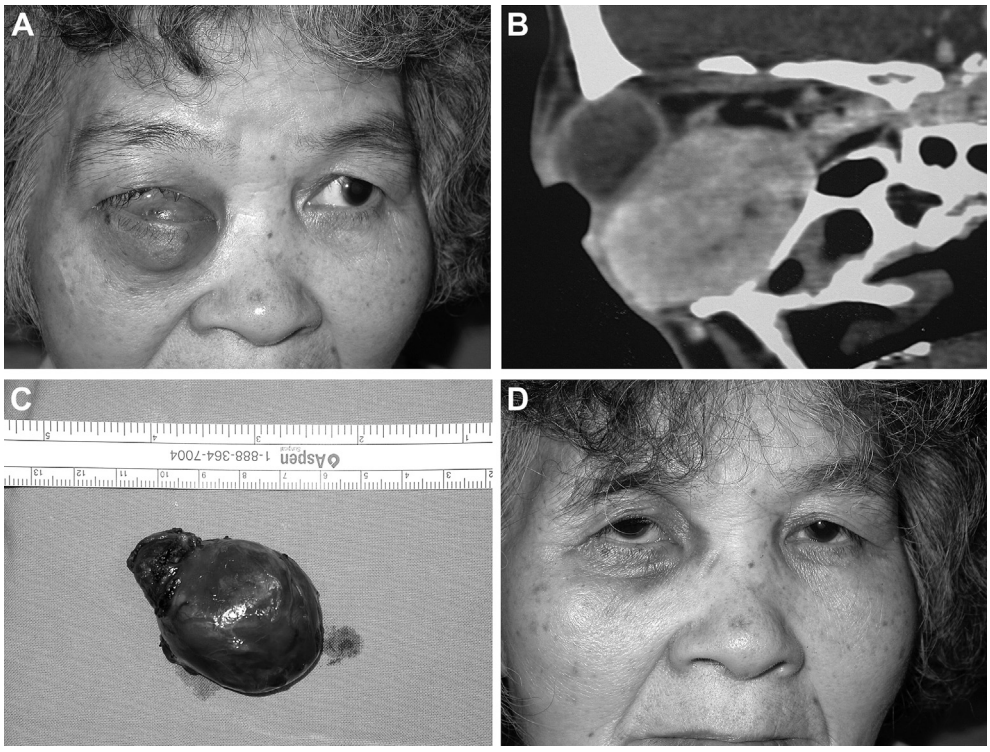

Figure 1 Case 4. (A) Facial appearance at initial examination reveals a protruding mass with severe displacement of globe and marked strabismus. (B) Sagittal computed tomographic image shows well-enhanced heterogeneous soft-tissue mass in the inferior orbit. (C) A well-circumscribed tumor measuring 5 × 3.5 × 1.5 cm^3. (D) Good prognosis after surgical treatment and adjuvant radiotherapy.

figures and apoptosis were described as having some malignant features.

Immunochemical studies in five of our six cases demonstrated CD34 reactivity (Fig. 4B), and only Case 6 showed negative CD34 but positive Vimentin and CD99 reactivity. However, cytokeratin, leukocyte common antigen (LCA), epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP), and smooth muscle actin were not detected in any patient.

Summaries of treatments and outcomes during follow-up periods are also listed in Table 2. Each patient underwent at least one operation and had regular follow-up. Only two cases (Cases 1 and 5) responded well to surgical treatment alone and have been doing well without recurrence or metastasis for at least 4 years. The other four patients had tumor excision combined with radiotherapy. Case 4 began intensity-modulated radiation therapy (IMRT) course immediately following the surgery with the goal of minimizing local recurrence. No sign of recurrence was found during 4 years of follow-up. Three patients (Cases 2, 3, and 6) received IMRT for therapeutic reasons after recurrences. In Patient 2, recurrence occurred 6 years after the surgery and the patient received IMRT after the second operation including total tumor excision with enucleation. The patient has been doing well since the last operation.

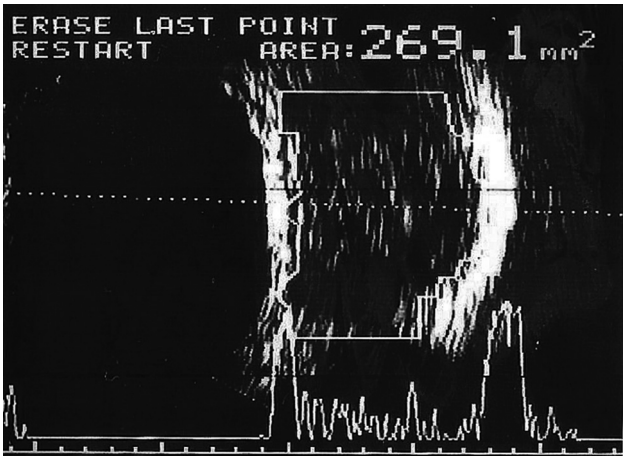

Figure 2 Ultrasonography of Case 2 showed low to moderate echogenicity.

Patient 3 had three surgeries due to two recurrences (in 2005 and 2006). The second and third operations were performed by an experienced neurosurgeon. However, subtotal excision (in 2006 and 2007) with adjuvant radiotherapy was performed after recurrences because total removal of the lesion was not possible. There has not been another recurrence for the past 4 years since the last operation.

Patient 6, who had the poorest outcome, having three orbital recurrences and multiple metastases of the lung and liver, had also received radiotherapy along with multiple surgical excisions. Two of the three orbital surgeries were performed by oculoplastic surgeon through orbitotomy with tumor excision, and the other one was performed by a neurosurgeon undergoing Simpson grade IV surgery. Two courses of IMRT were given for the second and third orbital recurrences. After the third surgery followed by

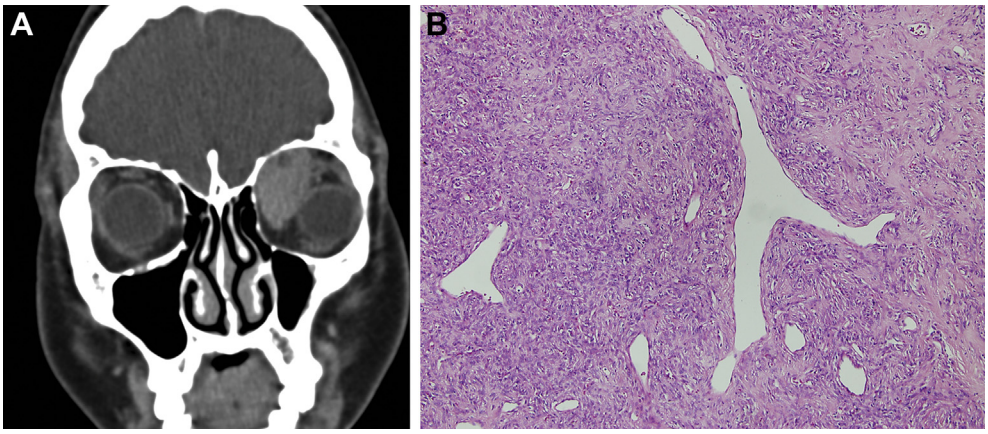

Figure 3 Case 5. (A) Coronal computed tomographic image shows heterogeneous mass in the superior orbit. (B) Histopathological appearance shows staghorn-configured, thin-walled branching vessels (hematoxylin—eosin; 200×).

radiotherapy, the orbital tumor was latent for 3 years. However, in 2009, another orbital recurrence was discovered via imaging studies, but only IMRT was performed. The residual lesion has been stable since. Regarding the metastases, pulmonary lesions were found 5 years after primary tumor was diagnosed and therefore three video-assisted thoracoscopic surgeries were performed in 2008, 2009, and 2010. Liver metastasis occurred in 2010, and an atypical hepatectomy was performed as the surgical treatment. The patient is still under intensive follow-up for the control of this malignant disease.

Discussion

Hemangiopericytoma, a rare vascular tumor arising from pericytes, which are of mesenchymal origin, was first described about 65 years ago.[1] Pelvic retroperitoneal spaces and soft tissues of the neck, extremities, trunk, and paraspinal areas are the most common sites of tumor growth.[9] Orbital involvement is rare.[2] It primarily affects adults with the median age ranging from 42 to 45 years, as found in two large series.[5,6] Our study was similar to previous studies; ages ranged from 30 to 62 years, with a median of 44 years. Most of the orbital tumors occur unilaterally without apparent predisposition for race or sex,[2] although in males twice incidence has been reported by Croxatto and Font.[5] However, in this series, five of the six cases were females. Because of the limiting case number, it is difficult to explain the influence of sex preference on disease incidence compared to previous studies. It may be the racial difference between the Caucasian and Asian people. Further case collections and studies are needed to explain the finding.

Orbital hemangiopericytoma almost always manifests with slowly increasing proptosis with or without associated pain and impaired visual acuity. Croxatto and Font[5] reported that 21% and 17% of their patients had pain and visual impairment, respectively, at initial examination. Among this six cases, only one patient (16.7%) had pain at initial presentation and four patients (66.7%) complained blurring or loss of vision. Among the four patients who suffered from severely decreased visual acuity, there was no obvious association with sex, duration prior to diagnosis, or size of the tumors. CT images showed compression of the optic nerve in three cases (50%). The cause of the impaired vision seems to be the location of the lesion. As presented in this study, other manifestations of a space-occupying mass within the orbit may include extraocular motility abnormalities with diplopia, displacement of the globe, afferent pupillary defect, and congested vessels. The majority of orbital hemangiopericytomas occur superiorly in the orbit, as reported by Sullivan and associates,[10] and therefore downward displacement is common. A similar result was also found in our series: four of the tumors (66.7%) were at least partially located superiorly, one (16.7%) had an inferiorly localized lesion (Fig. 1B), and the other one (16.7%) had a medially placed tumor. In addition, symptoms of eyelid swelling with blue or red discoloration of the adnexa oculi were occasionally noted, but congestion and edema of the conjunctivae or other soft tissue might not be as severe as those seen in patients with inflammatory pseudotumor.

Another clinical finding found in Patient 2 has seldom been reported. A tumor-like lesion with pigmentation was located around the macular area on the retina, which caused shallow retinal detachment. The patient had not received any examination until 10 years after first experiencing blurred vision. The tumor, located retrobulbarly, displaced the eyeball anteroinferiorly and compressed the globe. No sign of tumor invasion to the globe was found on image studies or in histopathological examinations. The change in the retina may be only due to the compressive effects of the lesion.

In the Croxatto and Font series,[5] the symptoms were present for less than 1 year in the majority of their patients. In this study, the symptoms were noted for not more than 6 months in four of six patients (66.7%), which are similar to those in previous studies.[5] We think that double vision or decreased visual acuity urged these four patients to undergo further ophthalmic examinations. Only Cases 1 and 2 presented symptoms 2 and 10 years prior to diagnosis, respectively. No visual disturbance and only cosmetic eye protrusion make Patient 1 unaware of

Table 2 Summary of pathological findings, treatment, and outcome.

No.	Histologic features (cells)	Histologic features (tumor type)	Histologic features (vessel appearance)	Immunohistochemistry	Treatments	Outcome
1	Spindle, bland appearing	Vascular tumor	Staghorn vessels	CD 34 (+) Cytokeratin (−) LCA (−)	→ Tumor excision (2001)	Doing well 9.5 y postoperatively
2	Bland with vesicular chromatin	Vascular tumor	Staghorn vessels	CD34 (+) EMA (−) GFAP (−)	→ Tumor excision (2001) → Total tumor excision with enucleation (2008) → IMRT (2008)	Recurrence (2008) Doing well 2 y post radiotherapy
3	Ovoid nuclei, inconspicuous nucleoli, and ill-defined cytoplasm, occasional mitosis	Vascular tumor	Staghorn vessels	CD34 (+) Cytokeratin (−) LCA (−)	→ Tumor excision (2003) → Total tumor excision (2005) → Subtotal excision (2006) → IMRT (2006)	Recurrence (2005, 2006) Doing well 4 y post radiotherapy
4	Monotonous spindle, arranged in fascicular pattern Occasional necrosis	Hypercellular tumor	Staghorn vessels	CD34 (+)	→ Tumor excision (2006) → IMRT (2006)	Doing well 4 y postoperatively
5	Oval to spindle; vesicular nuclei, indistinct nucleoli, indistinct cell borders, and low mitotic rate	Vascular tumor	Staghorn vessels	CD34 (+) GFAP (−)	→ Tumor excision (2006)	Doing well 4 y postoperatively
6	Vesicular nuclei, indistinct nucleoli; frequent mitotic figures and apoptosis	Moderate vascularity tumor	Staghorn vessels	Vimentin (+) CD99 (+) CD34 (−) EMA (−) LCA (−) Cytokeratin (−)	→ Tumor excision (2003, 2005 and 2006) → IMRT (2006) → Evisceration (2007) → VATS: wedge resection (2008, 2009) → IMRT (2009) → VATS: Lobectomy (2010) → Hepatectomy (2010)	Recurrence (2005, 2006, 2009) Metastasis Lung (2008) Liver (2010) Still under intensive follow-up

EMA = epithelial membrane antigen; GFAP = glial fibrillary acidic protein; IMRT = intensity-modulated radiation therapy; LCA = leukocyte common antigen; VATS = video-assisted thoracoscopic surgery.

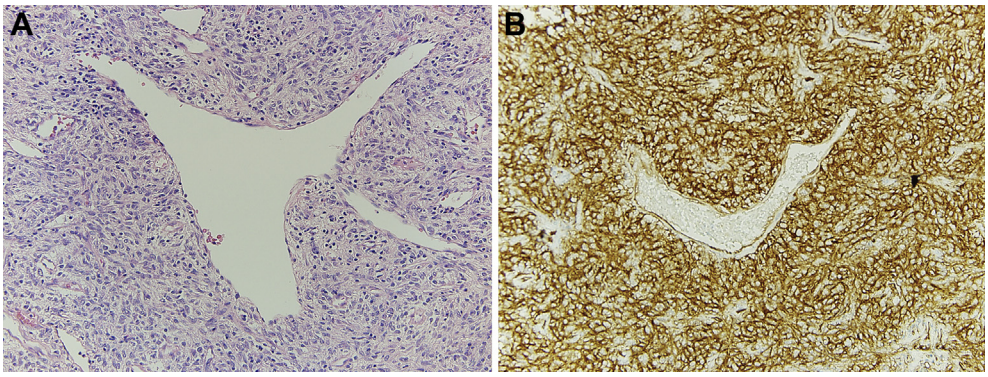

Figure 4 Case 4. (A) Histopathological appearance shows tumor cells arranged in fascicular pattern, interrupted by slit-like staghorn vessels (hematoxylin—eosin; 200×). (B) Immunohistochemical preparation reveals spindle cells with diffuse immunoreactivity to CD34 (200×).

searching for medical help. However, in Case 2, the patient came for help only after loss of vision due to personal reason. In Croxatto and Font's report, most of the tumors are circumscribed and range from 1.5 to 5.5 cm in diameter. Similar tumor sizes were also found in our series (ranging from 1.2 to 6 cm). However, neuro-ophthalmologic findings, such as abnormal pupil light reflex, do not seem to show a direct correlation to the size of orbital mass.

In previous study, ultrasonography usually showed a well-delineated round or oval lesion within the orbit with low to medium internal reflectivity. Mostly, the structure of the tumor is irregular, with solid and cystic components.[4] As Karcioglu et al concluded, although echography is helpful in delineating the masses, it does not offer any specific information for the diagnosis of hemangiopericytoma. On the other hand, cystic changes may even be misleading when formulating a differential diagnosis.[6] Therefore, ultrasonography has not been part of our routine image examinations for evaluating orbital masses within the past 10 years. This is why only two of our patients received B-scan echography prior to treatment (Fig. 2), but all six cases received CT. CT images of the orbital hemangiopericytoma tend to manifest as circumscribed, homogeneous masses, which enhanced with contrast medium.[5,6] In this study, the tumors enhanced with contrast in patients who had contrast CT study (Fig. 1B). CT appears to be a better diagnostic image examination for the tumors. However, in our series of six cases, hemangiopericytoma was considered among the initial list of differential diagnoses only in one case after CT was completed. Therefore, we concluded that image studies (including CT imaging and ultrasonography) alone may not be enough for the diagnosis of hemangiopericytoma.

The definite diagnosis of hemangiopericytoma usually depends on histopathological examination. Hemangiopericytoma typically appears as a mixture of spindle-shaped tumor cells with oval nuclei and scanty cytoplasms mixed with a network of thin-walled blood vessels or sinusoid-like spaces. This pattern is described as staghorn appearance.[9] In this study, all cases were reported as vascular tumors and featured with branching "staghorn appearance" vessels (Figs. 3B and 4A). In congruence with the previous observations, tumor cells portrayed spindle-shaped and oval nuclei in three of our pathological reports. Recently, some pathologists stated that orbital hemangiopericytoma and some cases previously designated as fibrous histiocytoma, giant cell angiofibroma of orbit, and solitary fibrous tumor have overlapping morphologic and immunohistochemical features and should be designated as solitary fibrous tumor.[11] In addition, Gengler and Guillou[12] also suggested orbital hemangiopericytoma and fibrous histiocytoma, giant cell angiofibroma of orbit, and solitary fibrous tumor designated as a solitary fibrous tumor spectrum from histopathological point of view. However, in publications listed in Pubmed, the terms "hemangiopericytoma" or "cellular variant of solitary fibrous tumor" have frequently been used as the same disease entity in recent publications. In this study, we still use "orbital hemangiopericytoma" as the diagnostic term.

Hemangiopericytomas may present with a variety of malignant-appearing forms, such as cellular pleomorphism, abnormal mitotic figures, zones of necrosis, and hemorrhage.[5] However, the malignant potential of the tumor cannot be determined on the basis of histopathological findings only. As reported in previous studies, a markedly pleomorphic cellular appearance can be associated with a benign course, whereas in other studies, benign-appearing spindle cells may result in metastasis.[5] In our series, two (Cases 1 and 2) of the six pathological reports described the cells as bland in appearance and another two (Case 3 and 5) with a low mitotic rate. Only two reports (Cases 4 and 6) revealed malignant features. In Patient 4, occasional necrosis was described, but the patient was given a good prognosis after surgical excision. However, among the three patients with recurrence (Cases 2, 3, and 6), Case 6 had been reported to have frequent mitotic figures and apoptosis. Nevertheless, Patients 2 and 3 had only "bland cells with low mitotic rate." In other words, among the four cases with histopathological benign-appearing spindle cells, 50% had poor prognosis of recurrence. However, half of the two patients whose histopathological reports revealed malignant featured cells still presented a benign course. It shows that histopathological findings are necessary to diagnose this disease, although clinical findings are most important to predict the prognosis. As mentioned above, it is difficult to predict the malignant potential of hemangiopericytoma by histopathological findings alone. Many patients who develop

recurrence and metastases exhibit variable histopathological features including malignant as well as benign features at the time of their initial tissue diagnosis.[13] Croxatto and Font[5] reported 30% and 15% local recurrence and metastasis rates, respectively. In our series, half of the cases (50%) had recurrence including one (16.7%) with multiple metastasis. The other half have been doing well thus far.

Hemangiopericytomas have a specific immunohistochemical profile, which aids in their diagnosis. The tumor cells react against vimentin and, in most cases, against CD34, but they lack immunoreactivity for EMA. They also lack immunoreactivity for cytokeratins (markers for epithelial malignancies), LCA (indicator of the hematolymphoid nature of a tumor), GFAP (markers for cells of central nervous system), and carcinoembryonic antigen.[13,14] In this study, five cases were CD 34 positive (Fig. 4B), and one case was vimentin and CD 99 positive. Cytokeratin, LCA, EMA, and GFAP were negative in all tumor specimens (Table 2).

The best treatment for hemangiopericytoma is total excision of the tumor, as it is usually well circumscribed, and complete excision of initial tumor appears to have the greatest impact on subsequent tumor behavior. Incomplete excision is associated with a higher risk of local recurrence and distant metastasis. In general, radiotherapy has been used only when the lesion was incompletely excised.[15] In addition, proton beam therapy had been shown to be effective in controlling recurrent orbital hemangiopericytoma in Gear et al's study.[16] In our series, four of the six patients received radiotherapy. In Case 4, IMRT was performed as an adjuvant therapy immediately after the first surgical treatment, and the patient has been doing well without recurrence or metastasis since the surgery (Fig. 1D). The other three patients received radiotherapy as a therapeutic treatment for local recurrence, with good response. In a previous study, one case of orbital hemangiopericytoma has been reported to recur as long as 33 years later,[17] indicating that an extended follow-up is required.

In summary, our study of orbital hemangiopericytoma coincides with the conclusions from previous studies that both clinical and histopathological findings are important for diagnosing and evaluating the treatment outcomes. Immunohistochemistry is a useful adjunct to histopathological diagnosis, and histopathological features alone cannot predict the biologic behavior regarding the malignant potential of the tumor. Ultrasonography and CT both are helpful in delineating these orbital masses and for follow-up, but they do not offer any insights for the diagnosis of hemangiopericytomas. Surgical excision remains the treatment of choice for this neoplastic disease, and radiotherapy can be an effective adjuvant treatment for recurrence.

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