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CASE REPORT

Giant intracranial mesenchymal chondrosarcoma with uncal herniation

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Summary Mesenchymal chondrosarcomas are very rare central nervous system (CNS) tumors consisting of undifferentiated mesenchymal cells plus islets of cartilage. We report a case of giant intracranial mesenchymal chondrosarcomas presenting with acute neurologic deterioration. A 22-year-old woman presented with right facial and hand numbness and ocular torsion for about 2 weeks. Magnetic resonance imaging (MRI) disclosed a mass in the right middle cranial fossa with a mass effect. Two days before scheduled surgery, the patient suddenly lost consciousness and was found to have uncal herniation. At emergency surgery, the tumor was totally excised. Pathologically, there was a dimorphic pattern of undifferentiated cells and islets of cartilage, consistent with mesenchymal chondrosarcoma. Although the patient survived surgery, she remained in a vegetative state. The tumor recurred 3 years after surgery. Intracranial mesenchymal chondrosarcoma is rare and should be considered when evaluating a tumor with dural involvement, particularly in younger adults. Differential diagnoses include intracranial meningioma and schwannoma. An aggressive surgical resection is highly recommended because of the high risk of recurrence.

1. Introduction

Mesenchymal chondrosarcoma is an extremely rare central nervous system (CNS) tumor. It is a variant of chondrosarcoma, itself quite rare and accounting for only 0.16% of all cranial and intracranial tumors.[1] Mesenchymal chondrosarcoma, first described by Lightenstein and Bernstein[2]

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in 1959, has distinct pathological findings that differentiate it from classical chondrosarcoma. The tumor comprises two components: undifferentiated mesenchymal cells and islets of cartilage.[3] We present a case of mesenchymal chondrosarcoma and review published reports of this entity.

2. Case report

A 22-year-old woman presented with a 2-week history of right facial and hand numbness, dizziness, headache, ocular torsion, and neck pain. She reported a transient episode of right peripheral facial palsy six months previously that had subsided spontaneously. On examination, the patient was alert. The pupils were isochoric with normal light reflexes. There was no visual field abnormality by confrontation, but there was right abducens palsy. There was right facial paresthesia, but the masseter muscles had full strength bilaterally. There was no facial paralysis, tongue deviation, or hoarseness.

An outpatient magnetic resonance imaging (MRI) of the brain showed a large lobulated tumor in the right middle cranial fossa, causing compression of the right lateral ventricle and a midline shift to the left. The signal intensity was isointense to gray matter on T1- and T2-weighted images. The lesion enhanced strongly after contrast administration (Fig. 1). The preoperative diagnosis was Schwannoma or meningioma of the right middle fossa. An operation was scheduled.

Two days before the scheduled surgery, however, the patient was brought to the emergency room with severe headache and fever. She was initially alert but rapidly deteriorated and had a Glasgow Coma Scale score of E3M4V1. The pupils were dilated bilaterally and without light reflex. Brain computed tomography (CT) revealed a lobulated tumor in the right middle cranial fossa with focal bony erosion of the base of the skull and scattered calcification within the mass. There was compression of the right lateral ventricle, right-to-left subfalcine herniation, and right uncal herniation. Perifocal edema was present (Fig. 2A).

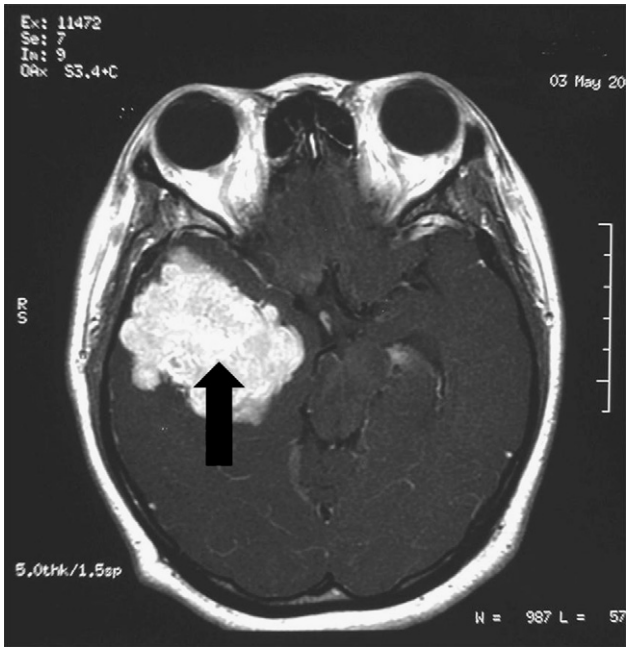

Figure 1 MRI of intracranial mesenchymal chondrosarcoma. The tumor is strongly enhanced with contrast (arrow).

An emergency decompressive craniectomy of the right fronto-temporo-parietal region was performed, and an external ventricular drain was placed. The tumor was dark-brown, lobulated, and hypervascular. The interface between brain and tumor was well defined. The tumor and its dural attachment were totally removed (Fig. 2B). Two days after the first operation, decompressive craniectomy was performed contralaterally because of persistently elevated intracranial pressure.

In pathological study, the tumor was light tan to dark brown and contained foci of oval-shaped bony tissue grossly. Microscopically, there were many small vascular spaces, some with a staghorn pattern. The tumor was composed of sheets of small, undifferentiated, oval or spindle-shaped cells and islets of cartilaginous tissue with foci of central calcification and ossification (Fig. 3). Mitoses were rare. Areas of necrosis were found. The dimorphic pattern and the presence of necrosis were consistent with a diagnosis of mesenchymal chondrosarcoma. In spite of all our efforts, the patient did not regain consciousness and remained in a vegetative state postoperatively.

The patient was brought back three years after the first operation with bulging of the scalp flap at the right craniectomy site. Brain CT revealed local recurrence of the tumor in the right middle cranial fossa (Fig. 4). Because of her persistent vegetative state, the family refused further surgery and other treatment modalities.

3. Discussion

Intracranial mesenchymal chondrosarcoma is an extremely rare CNS tumor. Given the rarity of this tumor, making the diagnosis and choosing proper management are challenging. We have reviewed cases reported in the English literature and updated several earlier literature reviews.

Intracranial mesenchymal chondrosarcoma was initially thought to arise from bone. However, in nearly half of reported cases it was extraskeletal, with the meninges being the most common site of origin.[3] Hypotheses about the pathogenesis posit an origin in embryonal cartilaginous rests in the skull and dura or from meningeal fibroblasts or multipotent mesenchymal cells in the dura or arachnoid,[4] but the evidence available is inadequate to draw a firm conclusion.

As noted above, mesenchymal chondrosarcomas have two basic components. The tumors are highly cellular with undifferentiated mesenchymal cells, but they also contain islands of well-differentiated hyaline cartilage.[3] The undifferentiated cells have round to spindle-shaped nuclei and are randomly arranged in sheets surrounding the cartilage. This appearance contrasts with classical chondrosarcoma, in which cells are usually well-differentiated with few or no mesenchymal components. Instead, the cartilaginous element predominates and gradually merges with the stroma. The microscopic distinction between

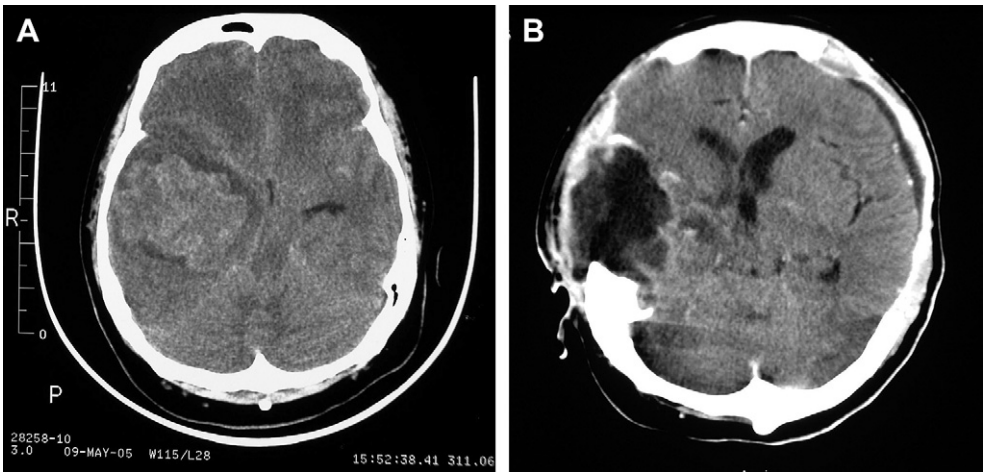

Figure 2 (A) Brain CT showing right temporal mass with perifocal edema and uncal herniation. (B) Repeat CT 2 weeks after surgery.

classical and mesenchymal chondrosarcoma is thus not difficult. Although the arrangement of undifferentiated cells of mesenchymal chondrosarcoma closely resembles that of hemangiopericytoma, the presence of cartilaginous foci is sufficient to exclude hemangiopericytoma.[5,6] If light microscopic features are equivocal, ultrastructural examination may be useful in delineating these two entities. In our case, the light microscopic findings were quite clear and sufficient to confirm the diagnosis of mesenchymal chondrosarcoma.

A few years after mesenchymal chondrosarcoma of non-osseous origin was first described, Dahlin and Henderson[7] reported the first case of one occurring intracranially. Since then, 39 cases have been reported, the largest single series being that of Rushing et al in 1996 with eight intracranial tumors. Including our case, the 40 patients' ages have ranged from 6 months to 68 years (mean 21.2 years), including 19 who were 15 years old or less. The male-to-female ratio was 9:11, and the peak incidence was in the second and third decades. This differs from classical chondrosarcoma, with a male-to-female ratio of 5:3 and a peak incidence in the sixth decade.[8] The tumor was attached to the dura in 31 patients, including ours, and was intraparenchymal in the other nine patients. The high proportion of cases with dural involvement lends support to the theory that the tumor originates in multipotent mesenchymal cells in the dura or arachnoid.[6,7] The outcome varied considerably, with 17 patients dying 3 days to 9 years (mean 39.7 months) after surgery. The longest reported survival was 10 years, and that patient died of a myocardial infarction. Recurrences occurred in 18 patients, ranging from 3 weeks to 3 years after resection. One patient developed leptomeningeal dissemination, and 3 had distant metastases to the vertebrae,[9] ribs,[5] or heart and intra-abdominal organs.[10] The wide range in reported outcome seems to indicate considerable variability in the natural history of the disease. Of all the reported cases, ours is the only one with uncal herniation occurring before surgery.

Recommended treatment for intracranial mesenchymal chondrosarcoma is radical surgical excision if possible.[3,11,12] Preoperative embolization has been suggested because the tumor is highly vascular.[5,13] Recurrence is common after resection, however. Some authors therefore recommend adjuvant radiation therapy to prevent recurrence, but it is difficult to tell if this mode of adjuvant therapy is effective

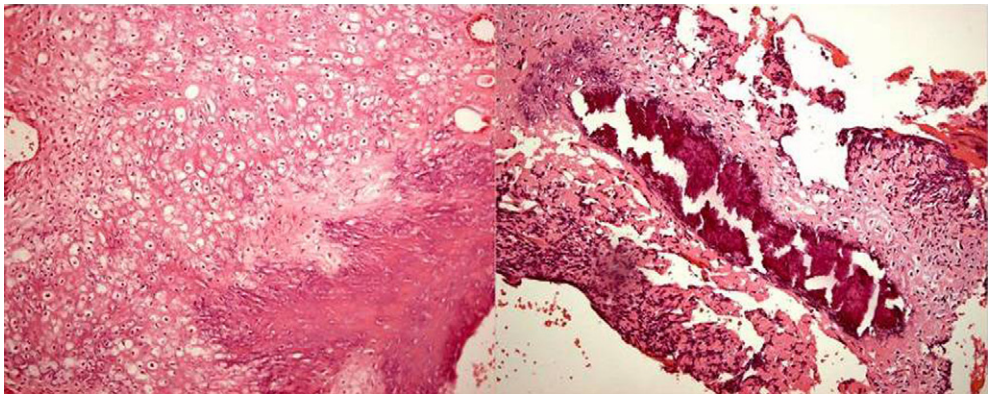

Figure 3 Tissue sample of resected tumor showing a dimorphic pattern of sheets of undifferentiated spindle-shaped small cells surrounding islets of cartilaginous tissue. Foci of calcification are present in cartilage. (H&E. × 100).

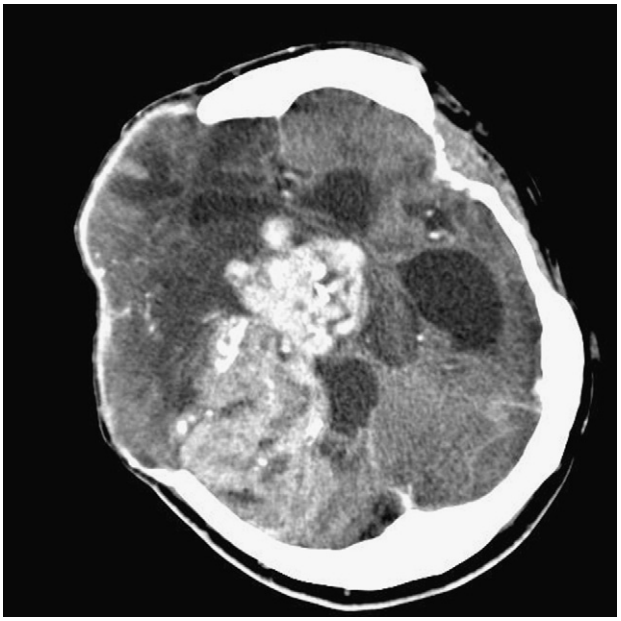

Figure 4 Brain CT with contrast demonstrating local recurrence of the tumor 3 years after surgery.

for the purpose.[3,8,14] Because of the few cases with metastases, adjuvant chemotherapy has also been recommended regardless of treatment for the primary tumor.[12,14,15] Again, the small number of cases prohibits drawing any conclusions about such treatment.

4. Conclusions

Intracranial mesenchymal chondrosarcoma should be considered when evaluating tumors with dural involvement, particularly in young adults. Aggressive surgical resection is recommended. The value of adjuvant radiation or chemotherapy is unknown, but given the rarity of the disease, it is unlikely that these modalities can be rigorously investigated, leaving it to physicians to decide in any particular case. Regardless of therapy, patients should be closely monitored after surgery because of the high risk of recurrence.

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