CASE REPORT

*Giant Cell Tumor of the Sphenoid Bone

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Abstract—
Objective and Importance: Giant cell tumor of the sphenoid bone is extremely unusual. This tumor is usually located in long bones and is rarely located in the cranial vault. It must be differentiated from reparative granuloma and the Brown tumor of hyperparathyroidism. This case report is only the second involving a patient in the first decade.
Clinical Presentation: A 9-year-old girl presented with cephalgia and diplopia. Radiographic imaging revealed an erosive mass in the clivus.
Intervention: The patient underwent an extended transphenoidal resection of the clivus. This was followed with 57.6 CGE in 32 fractionated doses.
Conclusion: At 1 year follow-up, the tumor remains stable. Based on anecdotal evidence, attempted resection with adjuvant radiation therapy allows for long-term survival in these patients.

CASE REPORT

A 9-year-old girl presented with 1 month of progressive frontal cephalgia. She experienced 2 days of diplopia at which time she was evaluated by neurosurgery.

Physical examination revealed a Tanner 1 white female, alert and oriented. A left six nerve palsy was identified, but fundoscopic examination revealed no papillodema. Visual fields and acuity testing were normal. The remainder of the neurological exam was unremarkable.

CT scan was obtained and showed destruction of the bone surrounding the sphenoid sinus (Fig. 1). MRI further revealed an upper clival mass that enhanced with contrast. Some extension into the right cavernous sinus with effacement of the right internal carotid artery were identified (Fig. 2). Thorough endocrine testing (including PTH) was unremarkable. Serum calcium and phosphates were also normal.

The patient underwent a trans-septal transphenoidal hypophysectomy. Biopsy revealed multinucleated giant cells with surrounding short spindle cells, and small nucleoli with no significant nuclear atypia were noted. A giant cell tumor was diagnosed (Fig. 3).

The patient required a second procedure that involved resection of the clivus through an extended transphenoidal route. This was performed utilizing a MKM microscope. The patient's postoperative course was unremarkable in both procedures. Adjuvant radiation was applied through a course of 57.6 CGE, delivered in 32 fractionated doses over 44 elapsed days. At 1 year follow-up, the patient's clinical course remains stable. No progression of tumor was noted on repeat MRI (Fig. 4).

DISCUSSION

Giant cell tumors involving the sphenoid bone are rare. These tumors primarily appear in long bones and

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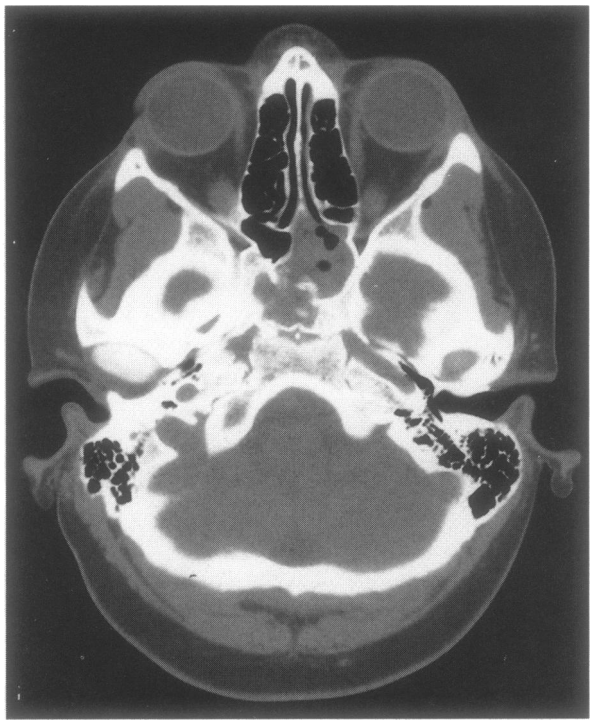


Figure 1. Computerized tomography scan without contrast reveals erosion of the right floor of the sella turcica.

account for 5% of all primary bone tumors.[1] The usual locations include the femur, tibia, radius, sacrum, and vertebral bodies.[2–4] They are only rarely found in the cranium. Giant cell tumors of the cranial base are found in both the sphenoid and temporal bones. It has been postulated that these bones are affected because they are the only cranial bones that are derived from endochondral bone formation.[5]

Currently, we are aware of only 40 cases in which giant cell tumors involve the sphenoid bone. The two most extensive studies were published in 1983 by Wolfe et al.[6] and in 1970 by Gessinger et al.[7] These two studies account for 22 of the reported cases. Watkins et al.,[8] in their review of 1992, brought the total to 38 cases. Two more cases were reported recently, including one involving a 10-year-old boy.[8,9] Our research indicates that this patient and our case are the only reported cases of sphenoid giant cell tumors in the first decade of life.

Presenting Symptoms

Giant cell tumors of the sphenoid bone present clinically with either cephalgia or cranial nerve dysfunction in the majority of cases. The cranial nerve dysfunction usually involves diplopia, visual loss, and fifth and seventh nerve lesions.[7,8]

Less frequent presentations include proptosis, endocrine dysfunction (ammenorrhia, galactorrhea, and hypopituitarism), and seizure activity.[7,9–13] One case of hemiparesis has been reported.[7]

Three cases associated with pregnancy have been reported.[7,8] However, 23 previously reported cases were female and all cases were between the ages of 13 and 52. Because the cases are seldom, it is difficult to establish any type of causal relationship.

Polyostotic Paget's disease has been associated with giant cell tumors found in the axial skeleton. Goldenberg et al.,[3] however, did identify two patients with "skull giant cell tumors." This article did not identify if these were giant cell tumors that were specific to the sphenoid bone.

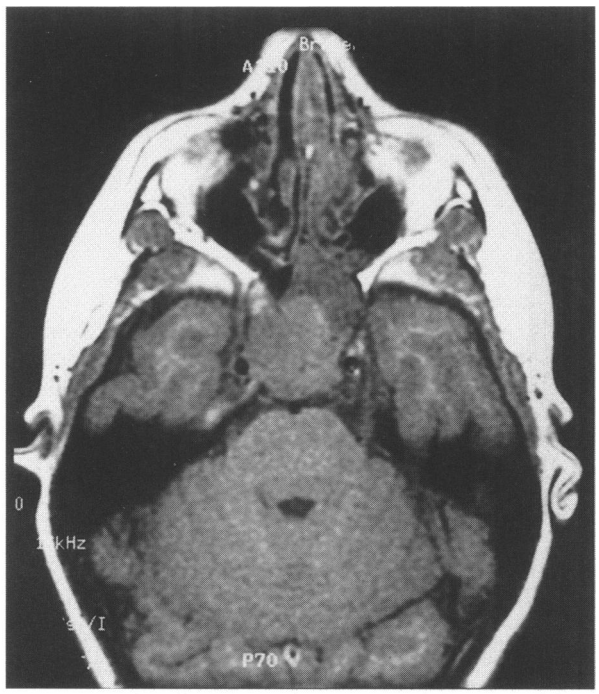
Radiological Evaluation

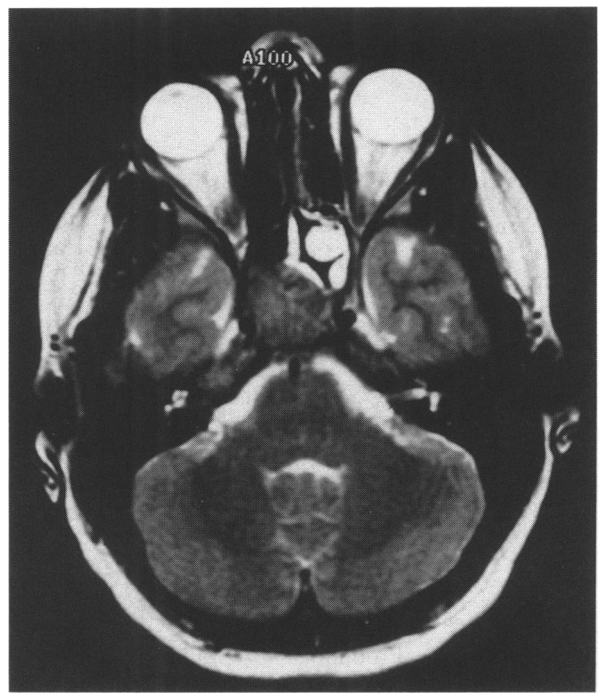
CT scanning has been used since 1983 for the evaluation of giant cell tumors.[14] This technique is helpful in demonstrating the extent of the tumor and in identifying the bony destruction seen with these lesions. However, no specific characteristics can be found that would help identify this tumor from other parasellar masses that also destroy surrounding bone. MRI may prove to be more effective, but the number of reported cases using MRI has been low, making tumor characteristics difficult to identify.[8,9,15–17] Giant cell tumors have a tendency to push away vascular structures and not encase them.[8] This was evident in our case, where the MRI showed the right carotid artery to be effaced but not narrowed or encased.

Histological Diagnosis

The diagnosis of giant cell tumors rests on the histology. These tumors reveal large numbers of multinucleated giant cells which are evenly distributed throughout the lesion. Another feature is a second population of smaller spindle-shaped cells. Mitotic activity may or may not be present.[8,9] Immunohistochemistry or evaluation with cell surface markers have not been productive means of histological diagnosis (*personal communication* with Dr. Unni of the Mayo Clinic Foundation).

Giant cell tumors must be differentiated from reparative granuloma. A giant cell reparative granuloma is a benign process that is possibly linked to trauma and intraosseous hemorrhage of the sphenoid bone. It is primarily found in the mandible and maxilla, and can occasionally be seen in the sphenoid bone.[18] The multinucleated giant cells can be found in both lesions. Giant cell tumors have a tendency to have larger giant cells and are more evenly distributed throughout the specimen. On the other hand, giant cell reparative granuloma have clumping of the giant cells around areas of hemorrhage.





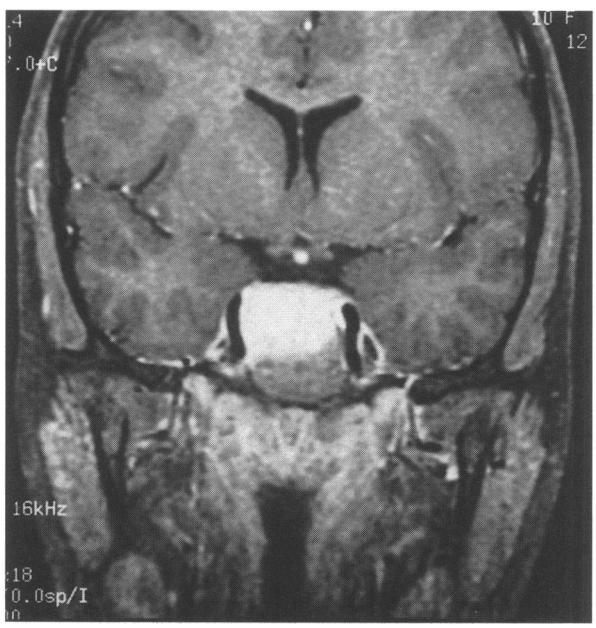


Figure 2. (A) T1 weighted axial magnetic resonance reveals a soft tumeric mass of the sella and sphenoid sinus. (B) T2 weighted axial MR again reveals hypo- and iso- intense mass of the sphenoid sinus. (C) Coronal enhancement MR with fat suppression showing tumor encroaching the cavernous sinus. Effacement of the right carotid artery is present but no narrowing is evident.

In addition, giant cell reparative granuloma are found more frequently in the younger population; most patients are less than 20 years old. The giant cell tumor is usually found in the 25 to 40 age group.[18]

The Brown tumor of hyperparathyroidism can also mimic giant cell tumors. Giant cell tumor cannot be distinguished from the Brown tumor of hyperparathyroidism by histological criteria alone.[19] It is usually differentiated through serum and urinary calcium as well as phosphate levels. PTH level may also be helpful.[6] Brown tumors of hyperparathyroidism are also multifocal and are found in other bones.[20]

As mentioned previously, Paget's disease of the bone has been associated with giant cell tumors.[3] However, we are unaware of any cases of sphenoid bone giant cell tumor that have been found in patients with Paget's disease.

Treatment

Treatment usually involves an attempt to completely resect the tumor. This, however, is extremely difficult due to the extensive involvement of the sphe-

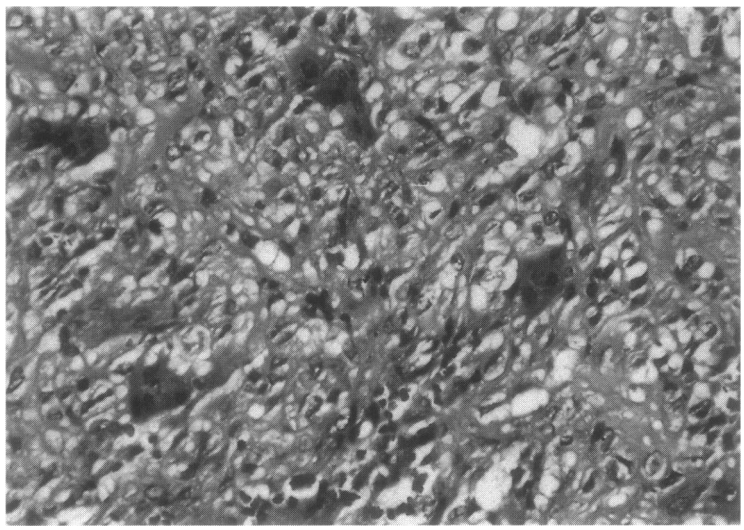
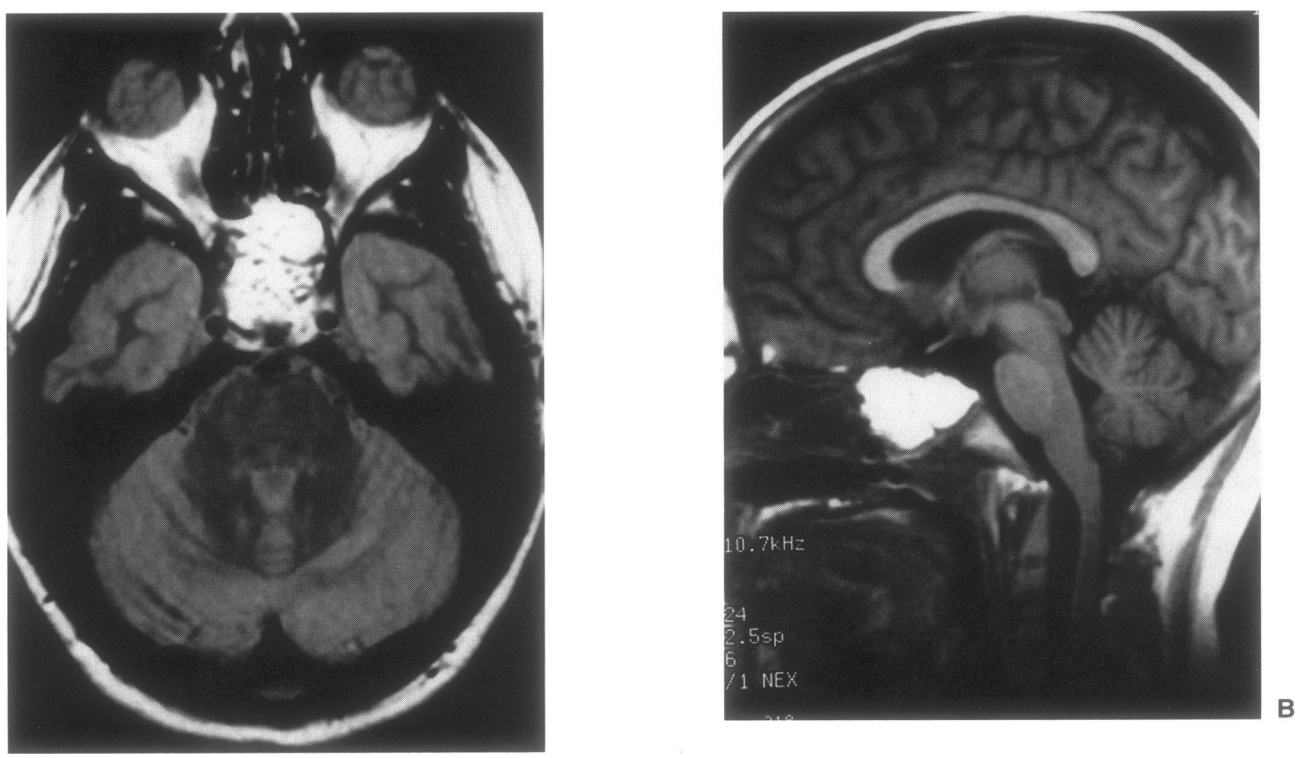


Figure 3. (A) Microscopic evaluation with H & E staining. Multinucleated giant cells with oval sphenoid nuclei are evenly dispersed throughout the specimen. No significant nuclei atypia or mitotic figures are identified. A second population of short spindle cells are also noted.

noid bone and surrounding structures. Obviously, the route of surgery is dictated by the location of the tumor, and our patient was treated through two transphenoidal routes. However, more extensive involvement of the clivus may require a LeFort I Maxillotomy.[8] In cases where the orbit or lateral sphenoid wing are involved, frontal-orbital or pterional craniotomy may be more appropriate.[9]

Adjuvant radiotherapy remains controversial. There has been no conclusive evidence that radiation benefits patients with sphenoid giant cell tumors. However, there has been some experience in treating giant cell tumors in other locations. Bell et al.[21] showed that there was no local recurrence or malignant transformation in their evaluation of 15 patients at 12 year follow-up.



A B

Figure 4. (A) T1 weighted axial magnetic resonance image reveals tumor to be stable at 1 year. Fat graft has been placed in the sphenoid sinus. (B) T1 weighted sagittal magnetic resonance image shows extent of clival resection.

There has been only one case report of a malignant transformation of a sphenoid bone giant cell tumor. A 20-year-old underwent 5000 rads of cobalt-60 irradiation over a 40 day period. Two years later the tumor recurred. Autopsy revealed the tumor to be a fibrosarcoma.

Further experience with giant cell tumors of the sphenoid bone will be necessary to determine the potential benefits of radiotherapy.

Prognosis

Giant cell tumors are typically histologically benign. Mitotic figures can be seen in the specimens of giant cell tumors, but not usually to a significant degree. However, malignant giant cell tumors of the sphenoid wing have been reported in the literature. In this patient, malignancy was based on histologically appearing "bizarre tumor cells." The patient received radiation therapy and was tumor-free at 8 months.[22] No long-term follow-up was noted. Prognosis remains variable. Survivability has been reported between 1 and 0.5 months up to 14 years after diagnosis.[6,22] Only seven cases were followed up for more than five years.[6,7] Of these cases, all three methods of treatment (biopsy and radiation, excision-only, and excision and radiation) were performed. Because of the rarity of this tumor and lack of long-term follow-up, no conclusions can be drawn. However, most cases reported with long-term follow-up had both attempted resection and adjuvant radiotherapy performed (five cases).

CONCLUSIONS

Giant cell tumors of sphenoid bone are amongst the most rare of cranial-based tumors. It is extremely important to differentiate this tumor from other pathologies that are located in the parasellar region. This is founded primarily on the histological criteria, because imaging studies are inconclusive. The Brown tumor of hyperparathyroid must be excluded, based on laboratory studies. Most cases have been treated with attempted excision combined with adjuvant radiotherapy. Based on anecdotal evidence, this regimen appears to allow for long-term survivability with this benign neoplasm.

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