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Giant Cell Tumor of the Skull: A Case Report and Review of the Literature

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BACKGROUND
Giant cell tumors are benign lesions that typically occur at the epiphyses of long bones that typically present with pain or swelling. Most data on giant cell tumors in the skull consist of case reports, and many large series of giant cell tumors have no examples in the skull.

METHODS
We report a case of giant cell tumor of the skull and review the literature on these lesions.

RESULTS
A 24-year-old woman presented with localized tenderness and mild swelling over the left inferior parietal and occipital bones. She was neurologically intact with a nonmobile, tender, palpable mass over the left suboccipital area. A computed tomography (CT) scan showed a radiolucent, expansile, lytic lesion involving the left occipital bone. The patient underwent a left occipital craniectomy with resection of the bone and epidural mass. Permanent histopathologic sections and immunostains revealed a giant cell tumor.

CONCLUSIONS
Giant cell tumors are generally benign, locally aggressive lesions for which surgical excision is the treatment of choice. This report contributes to the scarce literature on these tumors in the skull.

KEY WORDS
Giant cell tumor, skull tumor.

Giant cell tumors are benign lesions that typically occur at the epiphyses of long bones [11]. Clinically, they often present with pain and occasionally with swelling or decreased range of motion of an affected joint [4]. On physical examination a hard, sometimes crepitant, and painful mass is typically found [11]. The roentgenographic features are a lytic, radiolucent mass without a scelerotic border located at the ends of long bones [9]. Most data on giant cell tumors in the skull consist of case reports [1,5–7,10,12,13], and many large series of giant cell tumors have no examples in the skull. We report another case of giant cell tumor of the skull and review the literature on diagnosis, treatment, and prognosis.

Case Report

A 24-year-old right-handed woman with a history of migraines presented with localized tenderness and mild swelling over the left inferior parietal and occipital bones. Her medical history was otherwise noncontributory, and she had no prior surgeries. On physical examination, she was neurologically intact with a nonmobile, tender, palpable mass over the left suboccipital area. A computed tomography (CT) scan was obtained which showed a radiolucent, expansile, lytic lesion involving the left occipital bone, without extension into brain parenchyma (see Figure 1). The differential diagnosis included eosinophilic granuloma, plasmacytoma, metastatic lesion, fibrous dysplasia, or brown tumor of hyperparathyroidism.

The patient underwent a left occipital craniectomy with resection of the bone and epidural mass, duraplasty, and cranioplasty using a titanium plate. The dura appeared to be involved intraoperatively and was resected and sent for pathology. The intraoperative consult revealed a giant cell tumor. The patient's postoperative course was unremarkable, and she was discharged home the following day. She was to receive no adjuvant radiation therapy.

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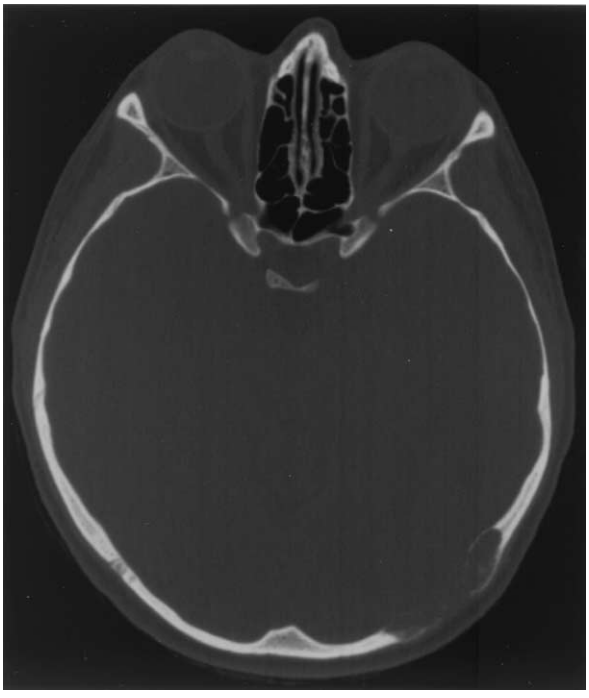
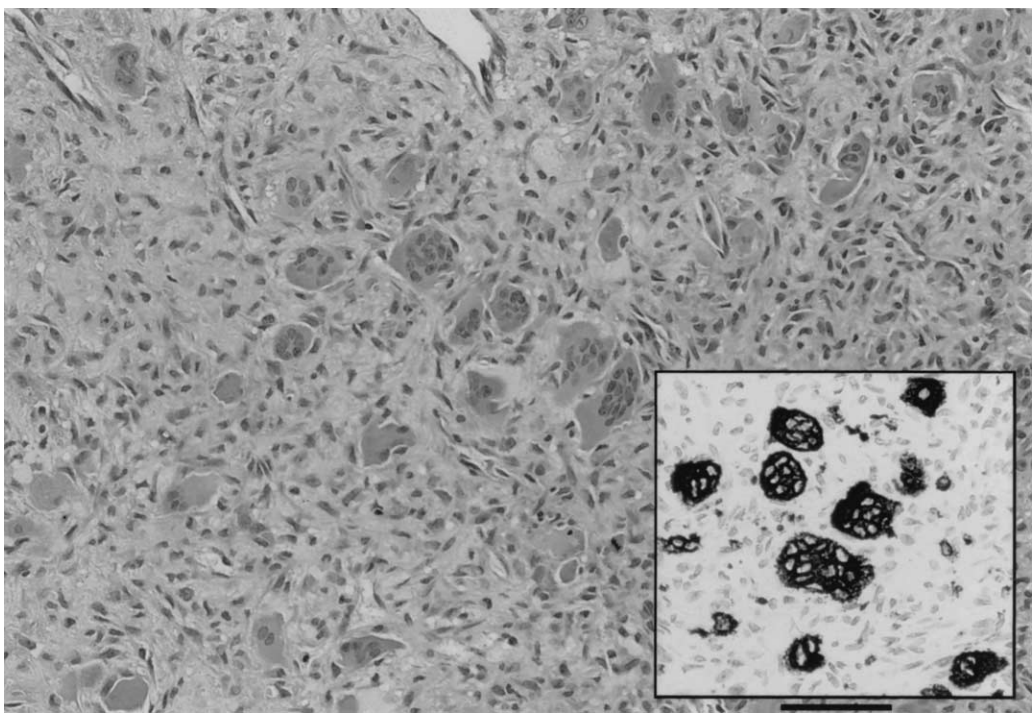


1 CT scan showing a radiolucent, expansile, lytic lesion involving the left occipital bone.

Permanent histopathologic sections demonstrated a mesenchymal tumor composed of a uniform distribution of multinucleated giant cells in a background of bland, oval to short-spindled stromal cells. Tumor involved skull, dura, and subgaleal tissue. Occasional mitotic figures were noted in the stroma. Immunostains for CD68 (macrophage marker) were positive in the giant cells; these cells also stained positively for acid phosphatase (histiocytic cell marker). A few of the stromal cells were positive for Ki67 (cell proliferation marker), but the giant cells were negative (see Figure 2).

Discussion

Giant cell tumors (also known as osteoclastomas) are thought to be derived from monocytic/histiocytic cells of the hematopoetic system [11]. They have often been confused with nonosseous fibroma, chondroblastoma, chondromyxoid fibroma, unicameral bone cyst with a cellular lining, giant cell (reparative) granuloma, aneurysmal bone cyst, brown tumor of hyperparathyroidism, and giant cell containing osteosarcoma [11]. Recent experiments have characterized these lesions as consisting of three cell types: osteoclast-like multinucleated giant cells; round mononuclear cells resembling monocytes; and a spindle-shaped, fibroblast-like stromal cell [15]. These experiments further suggested that the stromal cells secrete var-



2 Photomicrographs of the tumor demonstrate numerous giant cells dispersed in a predominantly spindled cell stroma. The larger one is stained with hematoxylin and eosin. The insert demonstrates positive reactivity of the giant cells with anti-CD68 (a macrophage marker) using immunoperoxidase staining with diaminobenzidine development. The magnification bar, 100 μm, seen below the insert indicates the magnification for both photomicrographs.

ious monocyte chemoattractants that stimulate monocyte migration to bone and their subsequent fusion into osteoclast-like, multinucleated giant cells. Thus, the stromal cell component may actually be of neoplastic origin, with the multinucleated giant cells being a reactive component [15].

Giant cell tumors comprise approximately 5% of all skeletal tumors and 21% of benign tumors [11]. Two series reported a slight female predominance [3,11], while another showed no gender predilection [15]. Peak incidence occurs during the second through fourth decades [11,15]. Pain at the affected site is the most common clinical presentation, with swelling and occasional pathologic fractures also seen [4,11,15].

Giant cell tumors usually arise at the ends of long bones and occasionally in the bones of the hands and feet and the sacrum. They rarely occur in the vertebrae above the sacrum. Very few series exists reviewing giant cell tumors of the skull. When giant cell tumors do occur in the skull, they tend to occur in the sphenoid bone. One series reported 5 of 429 tumors in the skull with one in the temporal bone, one in the mastoid, and three in the sphenoid [11]. One series, reporting on 15 cases of giant cell tumors of the skull of over 2,000 cases, found that 11 of these arose from the sphenoid bone [2]. Another series reported on 10 cases in the sphenoid bone [14]. The female preponderance appears to be even more exaggerated in cases involving the skull, and the median age may be slightly higher [2]. Two other series found none involving the skull in 189 [15] and 327 [3]. Several older case reports exist of giant cell tumors involving the sphenoid [5,10,12], temporal [10], frontal [1], and occipital bones [1]. Local bony destruction causing neurologic deficit is more common when these tumors arise in the medial sphenoid bone near the cavernous sinus because of proximity to vital structures. One case of acute visual loss has been reported because of extension of a lesion centered on the anterior clinoid process [8]. In general, however, these skull neoplasms rarely demonstrate intracranial extension causing neurologic deficits [7,13].

Osteoclastomas may be found in the presence or absence of Paget's disease [6]. Radiographically, the lesions are radiolucent with a nonsclerotic border [9]. Metastases to the lung may be seen with an incidence ranging from 1 to 2% [3,11].

Treatment options and prognosis are mainly derived from the literature on tumors in long bones. Surgical excision is generally the treatment of choice [11], with recurrence rates correlated to extent of surgical resection [3]. Prognosis is mainly related to extent of surgical excision, with little contribution from radiographic and histologic grading systems [3,11]. Chemical or physical agents such as phenol, liquid nitrogen, or methylmethacrylate have been used to augment the curettage or excision [4]. There is little evidence for a role for chemotherapy [4].

The treatment role of radiation therapy is controversial. While radiation has been used when complete surgical resection is impossible, there is evidence that irradiation predisposes the tumor to subsequent sarcomatous degeneration [4,11]. In one study, 8 of 10 patients with sarcomatous degeneration had previous irradation with greater than 4,000 rads. In the same study, 8 of the 27 patients who had received greater than 4,000 rads developed sarcomatous degeneration [3]. Despite these findings, some authors have recommended routine use of adjuvant radiation. Eleven of 15 patients in one series [2] and 9 of 10 patients in another [14] received adjuvant radiation therapy; none of these patients showed sarcomatous degeneration.

Conclusion

Giant cell tumors are generally benign, locally aggressive lesions with the potential to metastasize. This report contributes to the scarce literature on these tumors in the skull. These lesions generally present with pain and swelling. Radiographically, they appear as radiolucent lesions without sclerotic borders, which often appear in the sphenoid bone. There are three histologic cell types; the stromal cell component may stimulate monocyte immigration and fusion into multinucleated giant cells. There is a higher female predominance and a slightly higher age at presentation in giant cell tumors of the skull compared to other sites. Because of the tendency to appear in the sphenoid bone, these lesions can present with cranial nerve deficits related to cavernous sinus involvement. Surgical excision is the treatment of choice, and irradiation may predispose to sarcomatous degeneration. Radiographic and histologic grading systems do not predict clinical outcome, and extent of surgical resection has been shown to be predictive of prognosis.

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The traditionalists were younger than the World War II generation but admired them. They were caretakers of institutions.

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