Giant cell tumors of the calvaria

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The authors report and discuss four rare cases of primary, giant cell tumor of the cranial vault, a usually benign tumor that develops in young patients and has an extremely low rate of latent malignancy. The authors believe the preferred treatment to be total surgical removal of the tumor; only in rare cases of malignancy when the tumor is impossible to remove totally should this be combined with radiotherapy. Associated minor trauma played only a tumor-revealing role and was not involved in the etiology.

KEY WORDS · giant cell tumor · myeloplaxes · trauma

In general pathology, giant cell tumors are encountered in the short or long cartilaginous bones at the epiphysial-diaphysial level; the many synonyms include “giant cell tumor” (American), “brown tumor” (German), “tumor with myeloplaxes” (French), and “osteoclastoma” (British). In neurosurgery these tumors are rarely encountered in the vertebrae; there were only 15 cases in the series of 1180 vertebral tumors reported by Arseni, et al.1 The tumors are still more rare in the skull where they preferentially involve the base; calvarial tumors are indeed exceptional. In a monograph on primary tumors of the calvaria, Jelsma9 reported no giant cell calvarial tumors, nor did Lichtenstein and Jaffe11 or Coley and Bradley3 in their studies of giant cell tumors. Pancoast, et al.,12 described six cases encountered in the literature. Rowbotham14 described an osteoclastoma of the cranium. Recently giant cell tumors of the sphenoid bone have been reported by Pitkethly and Kempe,13 Geissinger, et al.,6 and by Derome.5

In the Clinic of Neurosurgery in Bucharest, between 1936 and March 1, 1974, we encountered four cases of giant cell tumors whose primary location was the calvaria; we are describing these cases in this paper.

Clinical Material

Table 1 summarizes the data on these patients. Certain additional characteristics are worth noting. The skin covering the tumor was always normal in appearance. Palpation of the tumor produced pain only in Case 2; in all cases the tumor was firm and immovable to deep palpation. The tumor was biopsied during operation; in Case 1 the histopathological diagnosis was made preoperatively by percutaneous biopsy at another hospital.

At surgery, each tumor appeared to have developed from bone (Figs. 1 to 4). Each was of a brown-reddish hue and bled slightly at the time of removal. No tumor traversed or invaded dura grossly even when the internal table had been destroyed. The bone surround-
FIG. 1. Case 1. **Upper:** Skull films showing extensive occipital bone disintegration predominantly on the right side. **Lower Left:** Microscopic appearance of tumor shows multinucleated tumor cells scattered throughout a moderately polymorphous cellular population. H & E, X 200. **Lower Right:** Tissue adjacent to the tumor shows osteofibrous proliferation. H & E, X 100.

**TABLE 1**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, (yrs)</th>
<th>Sex</th>
<th>Cranial Trauma</th>
<th>Duration of Symptoms</th>
<th>Tumor Size (cm), Location</th>
<th>X-ray Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8</td>
<td>F</td>
<td>none</td>
<td>5 yrs</td>
<td>8 x 5, occipital</td>
<td>occipital bone lysis</td>
</tr>
<tr>
<td>2</td>
<td>13</td>
<td>M</td>
<td>minor, lt temporal</td>
<td>3 wks</td>
<td>6 x 6, lt temporal</td>
<td>lt frontotemporal bone lysis</td>
</tr>
<tr>
<td>3</td>
<td>7</td>
<td>M</td>
<td>minor, lt parietal</td>
<td>3 wks</td>
<td>2 x 2, lt parietal</td>
<td>lt retrocoronal bone lysis</td>
</tr>
<tr>
<td>4</td>
<td>25</td>
<td>M</td>
<td>minor, frontoparietal</td>
<td>3 wks</td>
<td>6 x 4, frontoparietal</td>
<td>rt frontoparietal bone lysis</td>
</tr>
</tbody>
</table>

* All four patients had normal neurological and physical examinations, and all were treated by surgery with successful results. Histological examination showed giant cell tumor in all cases; Case 4 also had atypical cells and infiltration of dura.
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Fig. 2. Case 2. Upper Left: Skull film, lateral view, shows lysis of left frontotemporal bone. Upper Right: Tangential view of the lesion shows disintegration of bone. Lower Right: Photomicrograph of the tumor showing numerous multinuclear giant cells lying in a less dense cell mass. H & E, × 400.

Discussion

Historically giant cell bone tumor has been described under a variety of designations. At first it was considered a malignant tumor, and termed “giant cell sarcoma.” Subsequently, however, when surgical extirpation produced healing, the concept of malignant tumor was dismissed and it was classified as a benign tumor under the designation “giant cell tumor.” Yet, the local recurrences in certain operated or irradiated cases, as well as the rare metastases, have led to the conclusion that the tumor does possess a certain malignant potential.

Jaffe reported the occasional transformation of giant cell tumors, classifying them as Groups I, II, and III according to...
microscopical features. Group I includes the typical giant cell tumor described in our first three cases. Groups II and III (our Case 4) show sarcomatous alterations with clusters of fusiform cells, atypical formations of the stroma, and a reduced size of the giant cells. After total extirpation, local recurrence is extremely rare, and distant metastasis exceptional; when it does occur the usual location for the latter is the lung. Since no instance of a neoformative process in bone was encountered and the giant cell of the tumor resembles an osteoclast, some authors believe that the
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**Fig. 4.** Case 4. *Upper:* Lateral skull film showing lysis of right frontoparietal bone. *Middle Left:* Photomicrograph of giant cell tumor with dense polymorphous cells. H & E, × 200. *Lower Left:* Photomicrograph showing mitoses. H & E, × 600. *Lower Right:* Photomicrograph showing section through dura mater invaded by tumor proliferation. H & E, × 100.

designation of "tumor with myelophasis" is inappropriate. The lysis of bone without reformation is typical of giant cell tumors, and is assumed to be secondary to osteoclastic activity. Since giant cell tumors are most common in long bones and occasionally in the cartilaginous bones of the cranium such as the sphenoid, it has been assumed that the origin of the tumor is in cartilaginous cells. However, the primary occurrence of this tumor in the membranous bones of the calvaria argues against this hypothesis. No specific cell has yet been identified as an origin of the giant cells.
Minor trauma may be responsible for the reactive changes of local bleeding, but there is an obvious difference between simple, local, multicellular, posttraumatic reactivity and disintegration, and new formation of bone, recurrence, and metastasis. The recent minor trauma in some of our cases only played the part of revealing a latent neoformative process; by producing a hemorrhage in the pre-existing tumor, it increased its size and made it apparent. In forensic medicine the role of trauma in giant cell tumors ought to be regarded in this context. For example, in our Case 1, the tumor had been developing since early childhood; possibly a minor trauma that passed unnoticed had increased its size during the 3 weeks preceding admission. It is also noteworthy that all of our patients were young and passing through the stage of biological development or early maturation.

Clinical and radiological differential diagnosis must consider all primary or metastatic tumors of the cranial vault that produce local swelling and bone disintegration. These include intraosseous dermal tumor, eosinophilic granuloma, Recklinghausen's osseous disease, osseous aneurysm, and osseous hydatid cysts of the calvaria. Positive diagnosis is only possible by biopsy.

The prognosis in such giant cell tumors of bone is good, even though 10% to 15% are malignant. The relapses reported always occurred within 4 years of the initial treatment. Giant cell tumors are radiosensitive; however, recurrences after radiotherapy are more malignant than those occurring after surgery. In our cases we chose surgery with complete removal of the tumor, believing that this treatment has the best immediate and late results. Combined surgery and irradiation is appropriate when the tumor's location prohibits total removal.

References
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