Primary osteogenic sarcoma of the calvaria

Case report

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The author presents a case of skull neoplasm identified as osteogenic sarcoma. En bloc extirpation was performed. No other primary location of this rare tumor was found, and no metastasis has occurred nearly 2 years postoperatively.

Key Words • skull neoplasm • osteogenic sarcoma

According to Courville, et al., sarcomas of the cranial vault are probably the rarest of cranial tumors to be observed even in an extensive neuropathological practice. The following report is of a case of osteogenic sarcoma.

Case Report

A 19-year-old left-handed woman presented in October, 1973. She had a recent history of a walnut-sized mass in the left posterior parietal region initially noted to be firm, immobile, and nontender. There had been no associated drainage. For approximately 1 1/2 to 2 weeks prior to evaluation, the lesion had become progressively tender and soft, and in retrospect there was probably rapid enlargement in the antecedent 2 to 3 months. There was no history of injury to the area. For many years she had suffered from migraine headaches with an aura of photopsias. No other significant symptoms were elicited.

Examination. The patient was large-boned and moderately obese. In the left posterior parietal region was a firm, slightly distensible, and quite tender 8 × 8 × 3-cm enlargement. At the base laterally, there appeared to be a tender, well-defined, irregular margin of bone. There was no erythema, drainage, or cutaneous anomaly, and no bruit was audible. Neurological examination was entirely normal. A skull x-ray film (Fig. 1 left) revealed a "sun-flare" type of skull neoplasm with evidence of a significant overlying soft tissue mass. A brain scan showed abnormal accumulation of radionuclide, and transfemoral selective angiography revealed numerous local contributing vessels derived from the left superficial temporal and the occipital arteries (Fig. 1 right). There was questionable additional vascular supply via posterior branches of the left middle meningeal artery. A bone scan showed no other abnormality, and a chest film was normal. Serum alkaline phosphatase was 190 units.

Operation. Ligation of the left external carotid artery and en bloc excision were performed. There was no attachment to or involvement of the galea, nor was there any gross evidence of inner table disease (Fig. 2 upper). Primary cranioplasty was carried out.

Postoperative Course. The patient has done well with no evidence of local or distant...
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Fig. 1. Left: X-ray film, tangential view of calvaria, shows “sun-flare” osseous abnormality and soft tissue mass. Right: Selective external carotid angiography, tangential view, shows meningeal and scalp vasculature and tumor vessels.

recurrence as late as August, 1975. Initially, chemotherapy was started but side effects caused her to refuse further therapy. The radiation therapy consultant did not feel treatment was indicated.

Pathological Findings. Pathology specimens revealed clear surgical margins (Fig. 2 lower) and tumor at the inner table but microscopic examination showed no tumor cells breaching it. The specimens were reviewed by three neuropathologists independently, who concurred in the diagnosis of primary osteogenic sarcoma (Fig. 3).

Discussion

The term “osteogenic sarcoma” was originally coined by Ewing, whose classification included six types and two subgroups; this definition was impractical except for registry purposes or for specialists in neoplastic os-

Fig. 2. Upper: X-ray film of excised calvaria. Lower: Surgical specimen cut to show center of tumor.

Fig. 3. Photomicrograph of tumor shows anaplastic pattern and osseous component. H & E, × 200.
seous disease. The tumor is not derived from structures associated with bone, such as marrow or endothelial cells of blood vessels, but rather from osteoblasts. According to Vandenberg and Coley, since the bones of the cranial vault are ossified directly from primitive connective tissue, they are not likely to develop primary skull neoplasms; most other bones, including those of the cranial base, develop from connective tissue through a complex intermediary process of cartilage degeneration and absorption. Most primary bone tumors arise as a result of the transition incident to the development of cartilage or endochondrial bone. Courville, et al., have divided sarcomas into three groups depending upon their cellular origin: 1) myeloblastic sarcomas, exceedingly rare, arising from the marrow and resembling various histological types of myeloma that can occur in the petrous portion of the temporal bone as well as within the vault; 2) periosteal or fibrous sarcomas involving the temporal bone and characteristically presenting behind the auricle as sessile, firm, and at times multilobulated masses; and 3) osteogenic sarcomas, in which sarcomatous tumor osteoid is directly produced by the tumor cells. Chondroid, fibroid, and myxoid elements are often present to a variable extent; the presence of tumor osteoid, however, establishes the diagnosis even if only a small quantity is present. The unit cell of the tumor is spindle-shaped with variation in size and shape, some slender and fusiform, others larger and polyhedral.

The liability of or susceptibility to osteogenic sarcoma of bone already afflicted by a long-standing disorder is well known. In 1889, Paget noted neoplastic changes in two of 23 patients in his description of osteitis deformans, thus documenting the well-established relationship between these disorders. Even so, sarcomatous degeneration is unusual. The other common pre-existing conditions are radiation osteitis and benign chondroma; Paget’s disease, however, is the most common.

The medical literature of the 18th and 19th centuries was replete with reports of skull neoplasms and many were thought to be a consequence of focal injury. There appears to be widespread skepticism among pathologists as to the role of trauma, however, and Thompson, et al., believe that the relationship between trauma and sarcoma is unlikely, considering the relative frequency of the two conditions.

Osteogenic sarcoma usually arises in the metaphyseal ends of the shafts of long bones, particularly the distal femur, proximal tibia, and humerus. It is the most common primary malignant neoplasm of bone in man except myeloma, but it is rare in the calvaria. Geschickter in 1936 reviewed 500 bone neoplasms of which 13 (or 2%) originated in the calvaria. Of these, eight were osteogenic sarcomas but apparently not all were spontaneous. Cade reported only two cases of calvarial involvement in 133 patients with the disease. Thompson, et al., in 1970 reported three cases of “spontaneous” tumor in 14 patients with sarcoma of the calvaria. Between 1926 and 1948, Coley and Vandenberg reported 888 primary malignant tumors of bone with only seven (or 0.8%) originating in the calvaria; of the seven only four were osteogenic, but only two were found to be without underlying disease. In a 50 year period at the Mayo Clinic (1909-1958), 129 children under the age of 16 were treated for osteogenic sarcoma with no lesions of the calvaria included, and in the total of 2276 pathologically verified bone tumors at the Mayo Clinic up to 1955, 490 were osteogenic sarcomas of which only three were calvarial. Weinfeld and Dudley confirmed only one case at the Massachusetts General Hospital.

Primary osteogenic sarcoma affects children and young adults with a peak incidence in the second decade. The prognosis in general is grave because the tumor disseminates early and widely. Pulmonary metastases are common, as are lesions in other bones. Thompson, et al., have one patient who is well more than 10 years following surgery for primary osteogenic sarcoma of the calvaria; this is the first long-term survivor on record. Even one survival of this length suggests that radical surgery, as recommended by Long, et al., is the treatment of choice, provided metastases are not already evident.

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Osteogenic sarcoma of calvaria

References

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