Primary osteogenic sarcoma of the sellar region

Case report

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A case of primary osteogenic sarcoma of the sellar and parasellar region is described. A biopsy performed through the transnasal transsphenoidal approach appears to be the best method of establishing the diagnosis in lesions in this location. A previous case, documented in the German literature, is discussed together with the case presented here.

KEY WORDS  osteogenic sarcoma  transnasal approach  sella turcica  parasellar lesion

Primary osteogenic sarcoma is known to be one of the rarest tumors found in the skull. Although several cases of primary osteogenic sarcoma of the cranial vault have been reported in the literature, only a single case of primary osteogenic sarcoma of the sellar region has been reported previously. We are reporting a second such case.

Case Report

This 22-year-old man was admitted to our department on May 5, 1979, complaining of a left frontal headache of 4 weeks' duration, and diplopia which commenced 5 days prior to his admission. The patient stated that he had previously been in excellent health.

Examination. On admission, the patient was oriented and alert, and his general physical examination was within normal limits. Neurological examination revealed bilateral abducens nerve palsies, more severe on the left side. No other neurological deficit was apparent.

Laboratory data were within normal limits, except for an elevated serum alkaline phosphatase of 125 U/liter (normal values: 30–91 U/liter at 37°C incubation). Westergren erythrocyte sedimentation rate (ESR) was 10 mm in the 1st hour. Skull x-ray films, including tomographic studies of the sellar region (Fig. 1), revealed a lesion affecting the floor and the anterior clinoid process of the sella turcica. Both anterior clinoid processes were thickened, as was the tuberculum sellae. An area of new bone formation was seen to project posteriorly and superiorly from the latter structure. The anterior portion of the sphenoid sinus and the posterior cells of the ethmoid sinuses appeared opaque. The sella turcica was enlarged, and its floor irregular. The walls of the sella, particularly the anterior wall, were abnormally porotic. A bone scan revealed a pathological accumulation of radionuclide in the region of the sella turcica. A computerized tomographic (CT) scan showed a mass lesion in the region of the tuberculum sellae, encroaching on the anterior part of the sphenoid sinus. A significant and homogeneous enhancement of this mass occurred after infusion of the contrast medium (Fig. 2). Right brachial arteriography was normal.

Operation. The sphenoid sinus was approached transnasally and was found to contain a small amount of mucopus. The right lateral wall of the sphenoid sinus was covered with material resembling granulation tissue, and a biopsy was taken from this area. A tentative diagnosis of sphenoid sinusitis was made, and a polyethylene tube was left in place for irrigation.
of the sinus with antibiotics. The patient's immediate postoperative course was uneventful. Histological examination of the biopsy material revealed a fibrosarcomatous background containing osteoid tissue and spicules of newly formed bone surrounded by atypical osteoblasts, many of which showed pleomorphism and mitotic figures. The appearance was consistent with a diagnosis of osteogenic sarcoma (Fig. 3). The patient was advised to undergo x-ray and chemotherapy, but declined this treatment.

Postoperative Course. The patient returned to the hospital on July 23, 1979, with a rapidly progressive loss of vision in both eyes. His visual acuity in the right eye was limited to counting fingers at 20 cm, and he had no light perception in the left eye. The optic discs were still of normal color. At this stage, he agreed to receive radiotherapy combined with chemotherapy.

From August 2, 1979, over a period of 5½ weeks, the patient received cobalt-60 radiotherapy, with a total dose of 5500 rads. A combined series of chemotherapy courses was commenced on September 10, 1979. Each course consisted of: adriamycin, 80 mg on the 1st day; diamino-triazino-imidazole-carboxamide, 250 mg daily for 5 consecutive days; cyclophosphamide, 600 mg on the 1st day; and vincristine, 1.5 mg on the 1st and 5th days, respectively. An identical course was administered at four weekly intervals, to a total of five courses, when chemotherapy was discontinued because of the development of cardiotoxicity.

The patient was last seen in the outpatient clinic on December 22, 1980, 19 months following surgery. He is walking unaided; his visual condition had not improved since the oncologic therapy. A CT scan showed an enlarging mass lesion surrounding the parasellar region.

**Fig. 1.** Lateral tomogram of the sella turcica showing marked hyperostosis of the sellar and parasellar region and opacification of the sphenoid sinus and posterior ethmoidal cells.

**Fig. 2.** Computerized tomographic scan, without contrast material, showing a high-density mass lesion (attenuation values ranging from 154 to 401 Hounsfield units) in the sellar and parasellar region. *Left:* The lesion extends into the ethmoidal cells and cavum nasi. *Center:* The lesion is well demonstrated here in the sellar region. *Right:* Partial extension of the lesion to the left anterior cranial fossa.
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Discussion

Courville, et al., 3 pointed out that sarcomas that primarily invade the cranial vault are the rarest of all tumors to be seen in a lifetime of neurosurgical practice. This fact is confirmed in many published series in which bone tumors are reviewed. Of 500 cases with tumors of the skull reviewed by Geschickter, 4 only eight (1.6%) had osteogenic sarcoma; furthermore, not all of them were primary osteogenic sarcomas. Van-denberg and Coley, 11 reporting on 888 cases with primary malignant tumors of bone, found that only seven (0.8%) originated in the calvaria, and of the seven, only two were primary sarcomas of the skull.

Osteogenic sarcoma is the most frequently encountered primary malignant tumor of bone; it occurs most commonly at the growing ends of the long bones around the knee joint. This tumor affects mainly children and young adults, with a peak incidence in the second decade, and occurs most frequently in males (60%). 6 These tumors grow rapidly; the history is generally short, with pain and local swelling being the common presenting complaint. Not infrequently, metastasis to the lungs has already occurred at the time of the appearance of the first symptoms.

This tumor is roentgenographically recognized as a large osteolytic area with poorly defined margins. 8 Bone spicules radiate from the advancing margins of the osteolytic process, producing a “sunray” appearance; this typical feature is less frequently seen in the skull than in the long bones. In the skull, the spicules of bone are usually associated with thickening of the calvaria at the advancing edge of the tumor, due to subperiosteal extension. 8

Osteogenic sarcoma shows an extremely variable microscopic picture: foci of new bone formation, necrosis, hemorrhage, or a frankly sarcomatous stroma may be seen. 9 Lichtenstein, 8 in his monograph on bone tumors, stated that no two specimens of osteogenic sarcomas are exactly alike. The essential criteria for their diagnosis are: 1) the presence of a frankly sarcomatous stroma; and 2) the direct formation of tumor osteoid and bone.

In classifying sarcomas of the skull, three groups have been suggested, based on their cellular origin: 3

1) myeloblastic sarcomas, arising from the diploic marrow, corresponding to various histological types of myeloma (this form may occur in the petrous portion of the temporal bone as well as the vault); 2) periosteal or fibrous sarcomas involving the temporal bone and usually appearing behind the auricle as a firm multilobulated mass; and 3) osteogenic sarcomas, in which sarcomatous tumor osteoid is directly produced by the tumor cells (the presence of osteoid tumor establishes the diagnosis, even if only a small quantity is present).

Osteogenic sarcoma, in general, carries a bad prognosis, with a 5-year survival rate not greater than 10% with any form of treatment. 6,8 Thompson, et al., 11 have one patient who is well more than 10 years following excision of a primary osteogenic sarcoma of the calvaria; this is the first reported long-term survival of a patient treated by radical surgery.

Secondary osteogenic sarcomas of the skull vault arising from preexisting Paget’s disease of the skull is more frequent than primary osteogenic sarcoma. 9 Estimates of the incidence of sarcomatous degeneration in osteitis deformans range between 10% and 15%. 9

Occasional cases have been reported as late results of radiation therapy to the skull for intracranial tumors. All the reports quoted above concerning osteogenic sarcoma of the skull refer to tumors of the cranial vault. However, osteogenic sarcoma of the skull base has been reported only once. Kleinsasser and Albrecht reported a 48-year-old woman who had suffered from severe headache for 1½ years. A few days after her admission, she developed anesthesia over the left side of her forehead and face and a left abducens nerve palsy. X-ray films of the skull showed extensive, ill defined bone destruction of the base of the skull, affecting the sphenoid sinus, the sella turcica and the parasellar area, the left side of the floor of the middle fossa including the petrous pyramid, and the sphenoid ridge. Pneumoencephalography and carotid angiography were normal. A left temporal craniotomy revealed compression of the temporal lobe by a lesion that had invaded the base of the left temporal fossa extending intradurally. The lesion was bony hard in consistency and could only be partially resected. The patient died 6 days postoperatively. Autopsy revealed an extradural lesion in the sellar region, extending forward from the clivus as far as the frontal paranasal sinus, replacing the sphenoid bone and encroaching on the petrous pyramid bilaterally. Microscopic examination showed tumor osteoid, confirming the diagnosis of an osteogenic sarcoma.

This case, together with the one we have described, appear to be the only two reported instances of osteogenic sarcoma of the base of the skull, both cases affecting the sellar and parasellar regions. Both cases were characterized by a history of headache, with subsequent development of diplopia due to the lesion encroaching on the sixth cranial nerves. The case of Kleinsasser and Albrecht also showed involvement of the fifth cranial nerve, while our patient, after surviving operation, went on to develop involvement of both optic nerves.

The surgical removal of malignant tumors of the base of the skull, involving the middle fossa, presents almost insuperable difficulties. At the present time, the only possible therapeutic modalities are irradiation and/or chemotherapy. In view of the bad prognosis and our therapeutic helplessness in this condition, the establishment of an accurate diagnosis is of considerable importance. It seems that the constellation of symptoms and signs noted in the two cases described above, together with the findings on the skull x-ray films, bone scan, and CT scan, should suggest this, albeit rare, entity in the differential diagnosis with a reasonable degree of probability.

Unlike the improved prognosis of lesions of the anterior skull base, which today are dealt with by a combined intracranial-facial approach, malignant tumors of the middle fossa have a very bad prognosis and are not yet amenable to curative resection by any route. However, biopsy via a transnasal approach would seem to offer a reasonably safe and efficient method of establishing the diagnosis.

References

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