Primary osteogenic sarcoma of the brain

Case report

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osteogenic sarcoma involving the brain is exceedingly rare. We are presenting such a case treated by excision and radiation. The patient died as a result of a second recurrence of the tumor.

Case Report

This 53-year-old right-handed Caucasian man was admitted to the Huntington Memorial Hospital complaining of right-sided headaches, dizziness and left arm weakness of 10 days’ duration. He had previously been in excellent health.

First Admission. He was alert and oriented in all spheres. General physical examination was unremarkable. There were no gross deformities of the long bones. Examination of the head revealed normal symmetry of the calvaria. The pupils were 2 mm and equal and there was an obvious right ptosis, bilateral papilledema and a left central facial palsy. He had a left hemiparesis, the arm being much weaker than the leg. On sensory examination, decreased cortical association and projection sensory function of the left side of the body was found. Deep tendon reflexes were slightly hyperactive on the left. There was a negative Babinski response bilaterally. An echoencephalogram showed a significant midline shift of 7 mm from right to left. On brain scanning a 5-cm-round area of increased uptake was seen in the right midparietal region (Fig. 1). Treatment was started with dexamethasone, and cerebral angiography the following day revealed a right posterior temporoparietal space-occupying lesion (Fig. 2).

First Operation. A craniotomy was performed immediately and a seemingly well-encapsulated cystic tumor mass totally removed. On microscopic examination the tumor was found to be osteogenic sarcoma. A subsequent technetium isotopic bone survey of the entire body was unremarkable, as were x-ray films of both humeri, both radii and ulnae, both tibiae and fibulae, skull, spine, and...
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Laboratory data were also unremarkable, including a normal calcium and phosphorus content, a normal alkaline phosphatase, and an erythrocyte sedimentation rate of 12. The patient’s postoperative course was uneventful and he was discharged approximately 2 weeks after admission with a remarkable improvement in his left-sided weakness and right ptosis.

Second Admission. The patient was readmitted approximately 6 weeks later, with a chief complaint of increasing left-sided weakness. General physical examination again was within normal limits. Neurological examination now revealed a left homonymous hemianopia. Repeat skull films were normal except for the craniotomy defect. Echoencephalography again showed a 7 mm right-to-left shift. Repeat brain scan showed probable tumor recurrence. The chest film was normal. Angiography again confirmed a right posterior temporoparietal mass lesion. Prominent feeding vessels to the tumor were now seen (Fig. 3), strongly favoring tumor regrowth as the likely lesion.

Second Operation. Subsequently, repeat right temporoparietal craniotomy was undertaken with complete resection of the recurrent tumor, which was grossly cystic and apparently well-encapsulated as before. Postoperatively, radiation therapy was administered with a treatment course of 6000 rads over a 6-week period. The patient was discharged approximately 3 weeks after admission and after approximately 1 week of radiation therapy with gradual improvement to normal in the left arm drift.

Third Admission. The patient did well for 2 months following discharge. He was then readmitted following a focal seizure that involved the left upper extremity. He complained of increasing weakness of the left arm. General physical examination was again within normal limits. He still had a left hemiparesis primarily involving the arm. There was decreased sensation to pinprick on

FIG. 1. $^{99m}$Tc rectilinear brain scan, right lateral projection, showing large right temporoparietal lesion.

FIG. 2. Preoperative angiogram, right retrograde brachial injection. Note large, relatively vascular, temporoparietal mass.

FIG. 3. Angiogram taken on second admission; right retrograde brachial injection. Prominent feeding vessels to the tumor can now be seen.
the left. The patient was again hyperreflexic on the left as compared to the right. His mental status deteriorated rapidly despite treatment with dexamethasone, and he became comatose and died 4 days later.

Postmortem Examination. A complete autopsy was undertaken. Significant pathological findings were restricted to a second recurrence of the osteogenic sarcoma in the right temporoparietal lobe and mild bronchopneumonia. Gross and microscopic examination of all major organs revealed no evidence of osteogenic sarcoma. Microscopic examination of all long bones and the spine failed to show the neoplasm. Examination of the brain revealed a gritty tumor, 5.0 × 4.5 × 4.5 cm in size, buried deeply within the right temporoparietal area. The tumor was sharply circumscribed from the surrounding brain, and there was no observable association with any meningeal elements. Microscopic examination showed primarily a pleomorphic, bone-forming sarcoma containing predominantly spindle cells and without true encapsulation. There were numerous focal areas of well-organized bone formation (Fig. 4).

Discussion

The term “osteogenic sarcoma” supposedly was first used by Ewing.1 It is important to recognize that osteogenic sarcoma is the only malignant tumor derived from bone or bone-forming tissue, as distinct from associated structures such as bone marrow or the endothelial cells of the blood vessels.1 The primary cell type is spindle-shaped; however, there may be many giant cells present in the stroma, as was true in this case.

Osteogenic sarcoma is primarily a disease of the skeleton. The greatest incidence is in the growing end of the long bones, and most occur around the knee joint. The disease is more common in males, with 60% of cases, and in the second decade of life, with 80% of all patients between the ages of 10 and 30 years. According to Cade,1 in patients over the age of 30 the disease is exceedingly rare, and from age 50 onward it never occurs except in instances of pre-existing bone disease such as benign chondroma, Paget’s disease, and post-radiation osteitis. The natural history of the disease is one of gradual de-
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terioration and death, usually from pulmonary metastases. These metastases are the rule and are found in nearly every patient on postmortem examination. Visceral metastases other than to the lungs are relatively rare. There is occasional invasion of surrounding lymph nodes. Metastatic spread to other bones including the skull and vertebrae occurs in a few patients, and cerebral metastases have been reported. According to Courville, et al., sarcomas involving primarily the cranial vault are the rarest of all the tumors to be seen in a lifetime of neurosurgical practice. Cade’s review of osteogenic sarcoma, based on a personal experience of 133 patients, noted only two cases of skull involvement.1 Coley’s series lists 12 cases affecting the skull in a total of 985 cases of sarcoma of the bone.2

According to Courville, et al., and others,3-7 the sarcomatous change secondary to Paget’s disease causes the majority of cases of osteogenic sarcoma of the skull. Courville divided the tumors of the cranial vault into three groups based on their cellular origin: the myeloblastic sarcomas that arise from the marrow of the diploë, the periosteal sarcomas located most commonly on the temporal bone, and the osteogenic sarcomas of the cranial vault, usually secondary to Paget’s disease. It is unlikely that our case could be explained as a metastatic lesion, as shown by the negative autopsy findings. The absence of a primary lesion of the skull, including Paget’s disease, is substantiated by the lack of abnormal findings in the skull x-ray films, bone scans, surgical and autopsy findings. Furthermore, it is known that even with involvement of the cranial bones with Paget’s sarcoma, actual invasion of the dura occurs very infrequently and invasion of brain tissue even after penetration of the dura is likewise extremely uncommon.4

The time-honored, almost universally accepted method of treatment for osteogenic sarcoma of the long bones is amputation followed by radiotherapy. Most patients succumb to pulmonary metastases regardless of early amputation which suggest that these patients have often already developed pulmonary metastases when first treated. It is for this reason that some patients are now given radiation for the purpose of palliation of symptoms without the amputation of limbs. New modes of therapy have recently been described which seem to be superior to classical methods of treatment for osteogenic sarcoma, especially in preventing pulmonary metastases. Treatment with adriamycin, methotrexate or immunotherapy seem especially promising.2

The origin of the tumor in our case must remain speculative. A small, primary tumor could of course have been missed at autopsy, since serial microscopic sections of every long bone and the skull were not done. However, this possibility seems unlikely in view of the negative radiographic findings. We assume, therefore, that the tumor arose from a multipotential mesenchymal cell in the brain, or perhaps was metastatic from an unrecognized focus elsewhere.

References

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