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Primary Osteogenic Sarcoma of the Calvaria

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Osteogenic sarcoma of the skull is rare [1–7], particularly as a primary development in the later decades of life without associated Paget disease or history of irradiation. Because of the paucity of cases, the radiologic findings of calvarial osteogenic sarcoma have not been described in detail. Previous reports have dealt mainly with plain skull radiographs for the diagnosis of this tumor before biopsy. We present two cases of primary osteogenic sarcoma of the calvaria in 11- and 71-year-old patients who did not have predisposing conditions. They illustrate characteristic neuroradiologic findings which we believe could provide accurate preoperative diagnosis and guide surgical management of these tumors.

Case Reports

Case 1

An 11-year-old boy had had a painless swelling on the right forehead for about 1 year. There was no history of trauma. A 2 x 3 cm firm, nontender mass was noted on physical examination. The neurologic examination was completely normal.

Radiologic evaluation included plain skull radiography, bone scanning, and cranial computed tomography (CT) (figs. 1A–1F). Cerebral angiography showed a large vascular mass with primary blood supply from the superficial temporal and meningeal arteries. Displacement of the superior sagittal sinus was also noted. Meningioma was considered as the preoperative diagnosis, and frontal craniotomy was performed with subtotal resection of the tumor. Pathology showed osteogenic sarcoma with tumor present at the surgical margins as well as focally invading the dura. However, the underlying brain parenchyma appeared to be intact. Postoperatively, the patient received chemotherapy with methotrexate, vincristine (four courses), bleomycin, Cytoxan, and actinomycin D, followed by two additional courses of methotrexate and vincristine.

Five months after subtotal resection of the tumor and 10 weeks after initiation of chemotherapy, he was noted to have enlargement of a bony ridge along the surgical defect. CT showed increased intracranial growth of the tumor mass. Repeat cerebral angiography (figs. 1G and 1H) again revealed a large, highly vascular mass. Occlusion of the superior sagittal sinus by tumor was noted at this time. Bone scanning and chest CT showed no evidence of distant metastasis, and cerebrospinal fluid cytologic examination was negative for tumor cells. After radical resection of the tumor with a large margin of normal calvaria, the patient was given two doses of cisplatin and actinomycin. No radiation therapy was given. About 8 months later, a third surgical procedure was performed for local recurrence of tumor. No further tumor recurrence or metastases have been found during 12 months of follow-up by cranial CT and bone scanning.

Case 2

A 71-year-old woman had had a painless enlargement of the right side of her face, especially over the zygoma, for 6 months. She had had a cerebrovascular accident 12 months earlier, with residual right facial and extremity weakness. On physical examination, there was obvious swelling on the right side of the face, lateral to the orbit and just above the zygoma. There was no skin erythema or tenderness about the mass. Movement of the temporomandibular joint was intact. Neurologic examination revealed nonfluent aphasia, right homonymous hemianopsia, and mild right facial weakness. These neurologic abnormalities, including a mild limp on the right, were thought to be related to the previous cerebrovascular accident.

Neuroradiologic studies obtained on admission are illustrated in figs. 2A–2F. On review of the CT scan obtained 1 year earlier during her hospitalization for cerebrovascular accident, the mass was noted to be present in the bone but without intracranial involvement, suggesting a slow-growing mass originating in the bony calvaria. The preoperative differential diagnosis included osteochondroma, chrondrosarcoma, and osteogenic sarcoma. Craniotomy was performed with removal of the tumor, which proved to be osteoblastic osteogenic sarcoma by histopathologic examination. Postoperatively, the patient received neither chemotherapy nor radiation treatment.

Three months after surgery, she was readmitted through the emergency room with a traumatic nasal fracture. A cranial CT scan (fig. 2G) obtained for evaluation of possible traumatic intracranial complications revealed recurrence of the tumor. Extensive bony destruction of the sphenoid wing was also seen. Further surgery was not performed. Currently, the patient is being followed and the tumor mass is growing extracranially.

Discussion

Osteogenic sarcoma is primarily a disease of the long bones, with greatest predilection for the metaphyses, especially around the knee joint–distal femur/proximal tibia. It is the only malignant tumor derived from bone, originating from

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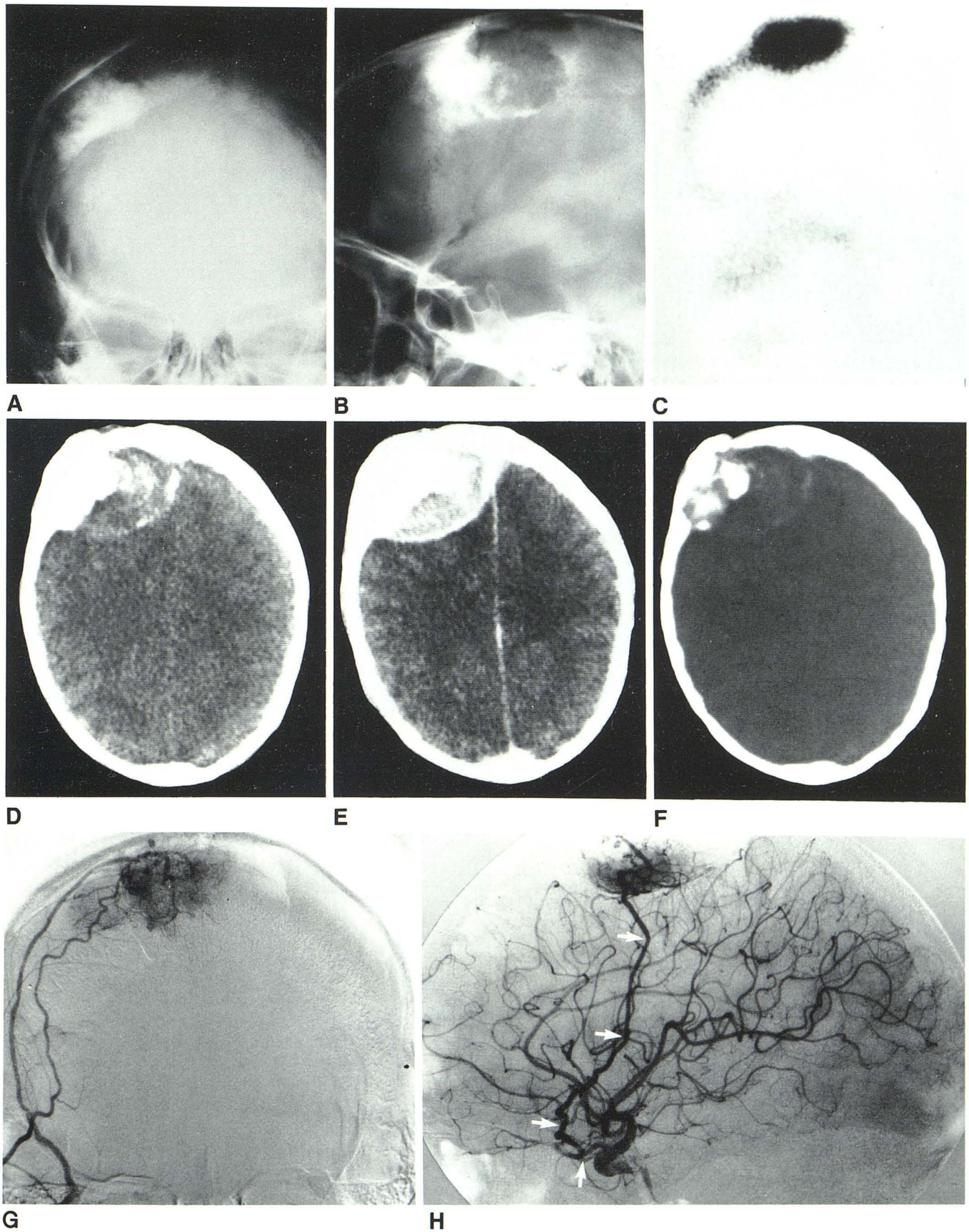

Fig. 1.—Case 1. Admission studies: Anteroposterior (**A**) and lateral (**B**) plain skull radiographs show large area of osteolytic and blastic changes in right parietal bone. **C**, ^{99m}Tc bone scan, right lateral view. Markedly increased uptake of radionuclide. Cranial CT scans without (**D**) and with (**E**) contrast material and with window setting adjusted for bone image (**F**) show irregular bony densities within enhancing soft-tissue mass, with apparent bony destruction of calvaria. Sharply delineated inner border of mass is rather characteristic for extraaxial mass. Postoperative studies: Right external (**G**) and left internal (**H**) carotid angiograms, anteroposterior and lateral views, respectively. Major blood supplies to tumor mass are from middle meningeal artery and superficial temporal branch of right external carotid artery. There is marked enlargement of recurrent meningeal artery (**H**, *arrows*), which arises from ophthalmic artery on left side.

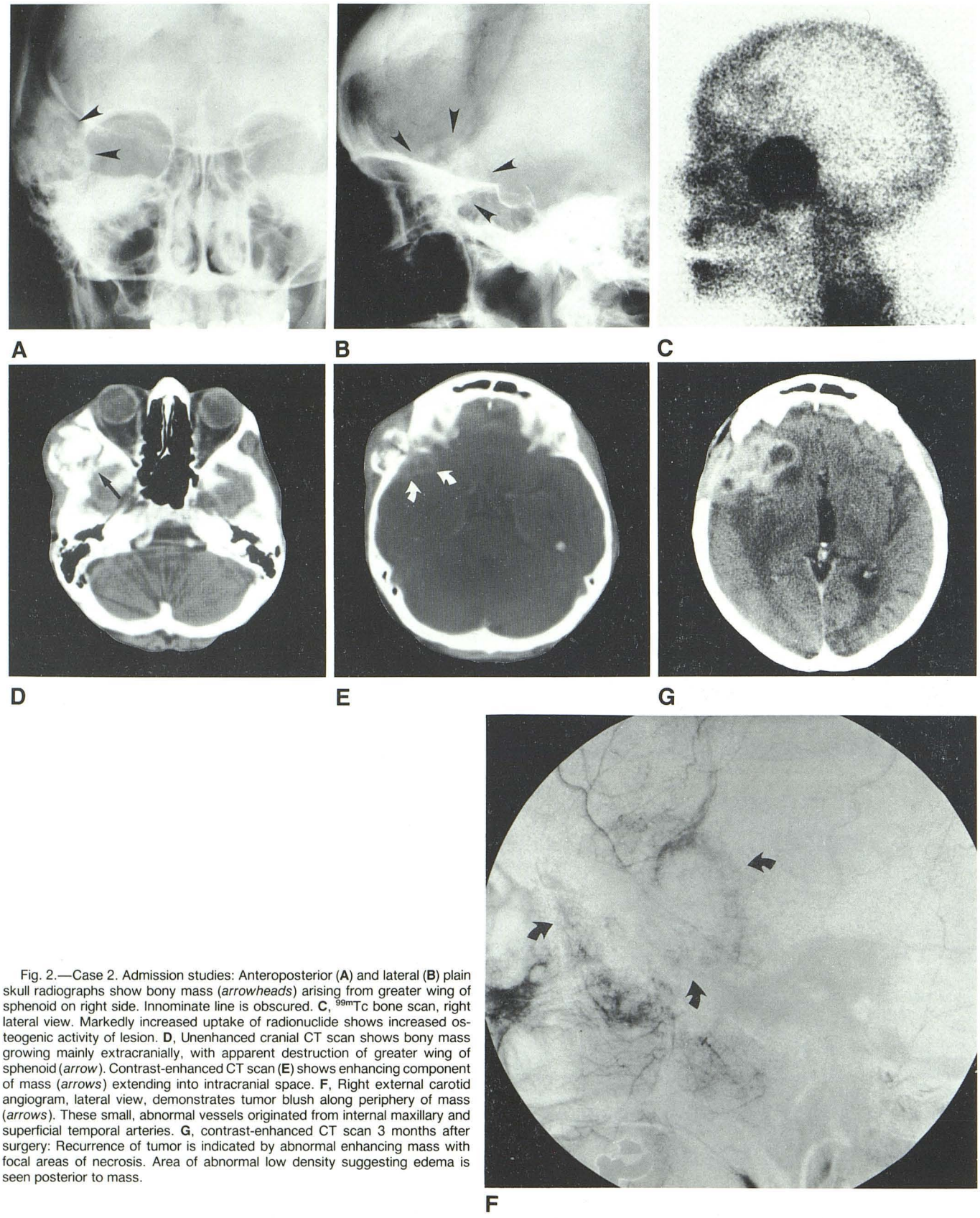

Fig. 2.—Case 2. Admission studies: Anteroposterior (**A**) and lateral (**B**) plain skull radiographs show bony mass (*arrowheads*) arising from greater wing of sphenoid on right side. Innominate line is obscured. **C**, ^{99m}Tc bone scan, right lateral view. Markedly increased uptake of radionuclide shows increased osteogenic activity of lesion. **D**, Unenhanced cranial CT scan shows bony mass growing mainly extracranially, with apparent destruction of greater wing of sphenoid (*arrow*). Contrast-enhanced CT scan (**E**) shows enhancing component of mass (*arrows*) extending into intracranial space. **F**, Right external carotid angiogram, lateral view, demonstrates tumor blush along periphery of mass (*arrows*). These small, abnormal vessels originated from internal maxillary and superficial temporal arteries. **G**, contrast-enhanced CT scan 3 months after surgery: Recurrence of tumor is indicated by abnormal enhancing mass with focal areas of necrosis. Area of abnormal low density suggesting edema is seen posterior to mass.

osteoblasts and not from associated structures such as bone marrow or vascular endothelium [1]. These tumors are most common in the second decade of life: 80% of all patients are between 10 and 30 years of age. There is a male predominance of 3:2.

Primary involvement of the cranial vault, excluding the mandible and maxilla, is exceedingly rare. One of the largest reported series of osteogenic sarcoma lists only 21 cases of primary origin in the cranial vault among more than 1000 cases [2]. Caron et al. [3] cited 11 cases of calvarial osteogenic sarcoma during their 37-year study. A large proportion of patients who had osteogenic sarcoma of the calvaria showed such well known predisposing conditions as Paget disease or previous irradiation. Other preexisting factors such as fibrous dysplasia [3, 4], multiple osteochondromatosis, osteomyelitis [2], and myositis ossificans [5] also have been recorded. Our two cases are unusual in that no predisposing factors could be found, particularly in the second case, which had developed at age 70. Review of the studied cases shows that the most common presenting complaint was localized swelling, usually without associated pain at the beginning. Unfortunately, this deceptively benign prodrome deterred these patients from seeking medical attention until the mass attained a fairly large size. The size, together with the location, precluded total surgical removal in many instances.

Bony changes seen on plain skull radiographs in calvarial osteogenic sarcoma are similar to those seen in long bones, depending on the relative osteolytic and osteoblastic nature of the tumor [3, 6]. Cranial CT findings in osteogenic sarcoma of the calvaria are not well documented; theoretically, they should be similar to those seen in the extremities, just as the plain-film findings are comparable. Only one published account of calvarial osteogenic sarcoma includes CT findings [2]. Although these findings, particularly the prominent new bone formation in the soft-tissue mass, are nearly conclusive for an osteogenic tumor, the differential diagnosis of osteogenic sarcoma from osteochondroma or chondrosarcoma cannot be made readily. Similar cranial CT findings are described in cases of chondrosarcoma of the base of the skull [8]. However, the cartilaginous tumors are unlikely to be seen over the cranial vault since the calvaria undergoes membranous ossification [5]. The osteochondromas usually are sharply defined and homogeneous in density [7]. Although the meningiomas often are calcified, the pattern of calcifications is either patchy and nodular or of a psammomatous type [9].

Other radiologic methods also play a role in the evaluation of patients with suspected calvarial osteogenic sarcoma. Radioisotope bone scanning is unquestionably more sensitive than any other radiographic examination for identifying and defining an osteogenic activity of the lesion, and it is useful in revealing unsuspected polyostotic involvement [10]. Angiography may aid the surgeon in his approach, particularly by defining the extent of tumor growth into the soft tissues and major vessel involvement by tumor [11]. As seen in other extraaxial lesions, calvarial osteogenic sarcoma derives its primary blood supply from the branches of the external carotid artery, particularly the meningeal and superficial temporal arteries. Tumor blood supply from the cortical branches of the internal carotid artery probably indicates tumor invasion beyond the dura.

The optimal treatment of calvarial osteogenic sarcoma is uncertain because of its rarity [2]. Radical surgery—extirpation including a large margin of normal bone—is the current treatment of choice [2–7, 12]. However, because of the location and often the extent of the neoplasm, this is rarely feasible. Experience with chemotherapy and radiation treatment is limited; they appear to offer little but palliation at best. Local recurrence of these tumors after surgery is the major cause of death and should be treated aggressively. Distant metastases are seen only occasionally, in contrast to the high incidence of pulmonary metastases from osteogenic sarcomas originating in the long bones. The prognosis for patients with calvarial osteogenic sarcoma is significantly worse than that for patients with tumors of the long bones or facial bones. The vast majority of the calvarial lesions are histologically highly malignant. Most of the reported patients died within 1 year after initiation of treatment; the 5-year survival rate is about 10% [2, 3].

The characteristic radiologic findings of calvarial osteogenic sarcoma described and illustrated here could provide accurate diagnosis, although the true nature of the lesion must be determined by histologic examination.

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