Giant ossifying fibroma of the skull

Case report

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A case of giant ossifying fibroma of the cranial vault is reported. The rapid extensive growth of this large parietal bone tumor inaccurately suggested malignancy. The literature relative to this benign bone tumor as well as its surgical management and follow-up findings are presented.

KEY WORDS • skull tumor • ossifying fibroma • bone tumor • parietal bone

Ossifying fibroma is a benign bone tumor that rarely involves the skull. This report illustrates that this benign bone tumor may rapidly reach gigantic size, and have radiological characteristics suggestive of malignancy. In the case presented here, the tumor was completely resected.

Case Report

This 22-year-old man was admitted to the Montreal Neurological Hospital in June, 1979, for treatment of a large parietal mass. He had noted a swelling over his left parietal region 1 year before this consultation. Thereafter the lesion had increased markedly in size. For 2 months prior to admission, the patient had suffered from throbbing headaches, brought on by exercise, and blurring of vision associated with rapid movements of the head. For a few months he had been bumping into objects and people on his right side. His personal and family medical histories were unremarkable.

Examination. Physical examination showed a large mass over the left parietal region, measuring about 15 cm in diameter, extending from 1 cm anterior to the coronal suture all the way to the lambdoid suture. The mass crossed the midline by about 3 cm and extended laterally to about 2 cm above the left ear; it measured 15 cm in height. It had a firm rubbery consistency and was not painful to palpation. Although the mass was fixed to the skull, the overlying skin was mobile and appeared normal.

Neurological examination showed a normal mental state, and bilateral papilledema with constricted visual fields. There were no motor, sensory, reflex, or coordination abnormalities. Considering these clinical findings, a primary malignant bone tumor of the skull was suspected.

Skull x-ray films showed a large mass destroying the cranial vault over the left parietal bone and extending across the midline. The mass was characterized by partly lytic and blastic processes, and loss of cortical outline of the inner and outer tables of the skull (Fig. 1). Computerized tomography showed the intracranial extent of the tumor and a shift of the brain to the right (Fig. 2). Bilateral selective internal and external carotid angiograms revealed that the vascular supply to the tumor was derived mainly from the left middle meningeal artery and the superficial temporal artery. These vessels were embolized preoperatively (Fig. 3). Except for uptake by the skull lesion, the bone scan was normal. The biochemical investigation was normal, with the exception of a slight elevation of alkaline phosphatase levels.

Operation. A diagnosis of malignant tumor of the skull was suspected, and a bilateral craniotomy was planned. At surgery, two large barn-door skin flaps showed the tumor to be well delineated from the skin. Although firmly adherent to the dura mater and the superior sagittal sinus, the tumor did not invade them, but it did depress the dura and the sinus by about 2 cm (Fig. 4). The tumor was yellowish in color, with a cartilaginous consistency and spicules of bone throughout. It was rather firm and avascular. Piece-meal removal was carried with cutting loops, a knife, and rongeurs. Although the edge of bone immediately
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adjacent to the tumor was soft, the bone became pinker and harder away from the tumor. One centimeter of what appeared to be normal bone was removed at the periphery. The tumor was completely removed. Examination of a frozen section resulted in a provisional diagnosis of a fibrous benign tumor. To provide covering and good cosmetic appearance, an autogenous split-rib cranioplasty was carried out by Dr. R. Daniels from the plastic surgery service of the Royal Victoria Hospital.

Fig. 1. Skull x-ray films, anteroposterior (left) and lateral (right) projections. A large tumor is seen on the cranial vault, partly lytic, partly blastic, destroying the outer and inner tables.

Fig. 2. Computerized tomography scan with contrast material showing a large area of increased density and an abnormal cranial vault. The falx is displaced to the right.

Fig. 3. Left external carotid angiogram, lateral projection, showing the vascular supply to the tumor through the superficial temporal and middle meningeal arteries.
FIG. 4. Left internal carotid angiogram, lateral projection, venous phase. A large avascular mass is visible over the parietal region, depressing the superior sagittal sinus without occluding it.

Postoperative Course. The postoperative course was uneventful. Clinical and radiological follow-up examination 2 years postoperatively showed no evidence of recurrence of the tumor; there was mineralization of the cranial vault at the site of the cranioplasty (Fig. 5).

Pathological Examination. Multiple fragments of the tumor were submitted for histological examination. Microscopically, the mass consisted largely of a proliferation of fibrous tissue and spicules of bone. The fibrous component was poorly cellular, varying from early fibroblastic proliferation to a hyalinized zone (Fig. 6 left). The spicules of bone were interspersed with fibrous proliferation; some were rimmed with osteoblasts and others with osteoclasts (Fig. 6 right). Woven bone in the process of transforming to lamellar bone was noted, as were many dilated vessels in the fibrous component. Mitotic figures were rare, and no atypical cellular features could be visualized. On one fragment, a small area of benign cartilaginous proliferation could be seen.

A small fragment of tissue prepared for ultrastructural study revealed mainly fibroblasts, occasional cells containing calcification in their cytoplasm, and many mature collagen fibers. The benign fibroblastic nature of the tumor, the amount of osteoid material and occasional cartilaginous metaplasia, the lack of any inflammatory process, and the absence of extension from or to the dura all supported the diagnosis of ossifying fibroma.

Discussion

Ossifying fibroma is a benign, usually monostotic, fibro-osseous tumor. Although this tumor is typically found in the maxillary sinus and mandible, it has also been reported in the paranasal sinuses, frontal, ethmoid, and sphenoid bones, and orbital roof. It has occasionally been encountered in the long bones. Involvement solely of the cranial vault is extremely rare.

The tumor usually develops slowly but, as this case illustrates, it may grow very rapidly. It is usually well delineated by a thin shell of bones that it expands outward, but it can also be invasive. In our particular case, the tumor was thought to be malignant. The tumor had a cartilaginous consistency and was rather avascular. This avascularity may have resulted in part from the preoperative embolization. The tumor had the usual gross characteristics: it was firm, gritty, tan-yellow in color, and well circumscribed. It did not invade the dura, and its gross appearance suggested a benign process.

Although the pathogenesis of the tumor is not clear, a few theories have been proposed. Trauma has been suggested as an etiological factor by Smith and Zavaleta, who observed a history of trauma at the tumor site in four of 20 cases. A developmental abnormality has also been proposed, since this tumor usually arises from membranous bone and occurs in patients in a young age group. On the basis of electron microscopic findings of bone formation and resorption, Kempson proposed that the tumor results from excessive fibrous repair of resorbed bone due "to defective bone formation or excessive osteoclastic activity."

Review of the literature uncovered few well documented cases involving the cranial bones alone. More cases compromised the frontal bone than any
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Other, but these were usually associated with frontal, ethmoid, or sphenoid sinus involvement or were located supraorbitally. Smith and Zavaleta reported four cases of ossifying fibroma of the skull, but only one involved the skull alone; its exact location was not specified. Thomas and Kasper reported one case of frontal bone involvement that did not seem associated with the sinuses. In 1966, Hiranandani, et al., reported one case of exclusive frontal bone involvement. More recently, Yamashita, et al., reported on a young boy with an occipital ossifying fibroma, and Seitz, et al., described a case in which a small tumor involved the parietal bone.

The usual radiological characteristics of ossifying fibroma (a rather well demarcated lesion, circumscribed by the thin shell of bone that the mass displaces outward) were not present in this case. The disappearance of the inner and outer tables of the skull, the blastic and lytic portions shown by radiology, and the tumor extension over the midline suggested a much more aggressive process. These characteristics were likely secondary to the tumor's large size and very rapid growth, resulting in different parts of the tumor being at different stages of maturation, with bone formation, bone resorption, and fibrous proliferation.

This case illustrates that this rare benign tumor of the skull may grow very rapidly and be associated with neurological signs due to an intracranial mass effect. It also illustrates that, even though gigantic in size, the tumor need not invade the dura and superior sagittal sinus, and that removal can be complete. To prevent recurrences, it is recommended that a margin of normal bone and peristome also be removed.

References

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