Benign fibrous histiocytoma of the skull with increased intracranial pressure caused by cerebral venous sinus occlusion

Case report

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The authors present a very rare case of benign fibrous histiocytoma of the skull with increased intracranial pressure caused by sinus occlusion. A 33-year-old woman was referred for investigation of a right occipital protrusion with tenderness and double vision. She had only mild divergence insufficiency and bilateral papilledema neurologically. Imaging findings showed that the skull tumor was located at the right occipital bone with bone disruption and a compressed right sigmoid sinus. When planning the resection, caution was required to spare the collateral flow so as to manage the intracranial pressure. Immunohistochemical analysis showed that the tumor was positive for CD68, α1-antichymotrypsin, and α1-antitrypsin. From these findings, the tumor was diagnosed as a primary benign fibrous histiocytoma of the skull. (DOI: 10.3171/2008.11.JNS081206)

Key Words • benign fibrous histiocytoma • increased intracranial pressure • sinus occlusion • skull tumor • venous congestion

Primary BFH of bone is a rare skeletal tumor accounting for ~ 1% of surgically managed benign bone tumors. Fewer than 100 cases have been reported in the literature. To our knowledge, there have been only 5 case reports of a BFH in cranial bone and no cases of BFH in the neurocranium. The present case was also characterized by increased intracranial pressure accompanied by sinus occlusion caused by this skull tumor. We present the first reported case of BFH in the neurocranium, including its management in the perioperative period, and a review of the literature.

Case Report

History and Examination. This 33-year-old woman developed headache and right-sided tinnitus over a period of 2 months. She was referred to our hospital because of the additional symptoms of right occipital protrusion with tenderness and horizontal diplopia. The cutaneous veins of the bilateral temporal area were visibly dilated from the surface of the skin. Neurological examination revealed bilateral papilledema, dilation of the retinal arteries, and bilateral abducens nerve palsy. Computed tomography scanning revealed that the tumor was a homogeneously enhancing mass with bone destruction (Fig. 1A–C). The present case was also characterized by increased intracranial pressure accompanied by sinus occlusion caused by this skull tumor. We present the first reported case of BFH in the neurocranium, including its management in the perioperative period, and a review of the literature.

Abbreviations used in this paper: BFH = benign fibrous histiocytoma; ICP = intracranial pressure; SSS = superior sagittal sinus.
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Operation. Prior to the operation, we performed a compression test for collateral veins around the patient’s scalp and confirmed that no symptoms were produced. A semicircular skin incision was made using the StealthStation TREON Navigation system (Medtronic, Inc.) to detect and spare cutaneous and subcutaneous veins that could be collateral veins caused by sinus occlusion (Fig. 3 left). Total tumor removal was achieved after induction of general anesthesia. The tumor mass was separated from the subcutaneous tissue and dura mater with little adhesion. The tumor was cream-yellowish, elastic, hard, and located in the skull bone (Fig. 3 right). The patency of the transverse and sigmoid sinuses was confirmed by micro-Doppler ultrasonography after removal of the tumor.

Histological Findings. Histological examination revealed the proliferation of histiocytic foamy cells and spindle-shaped fibroblasts arranged in a storiform pattern (Fig. 4A). There were no malignant appearances, such as nuclear atypia, pleomorphism, or atypical mitosis. Immunohistochemical analysis was performed on the formalin-fixed, paraffin-embedded sections by using the streptavidin-biotin-peroxidase complex labeling method. The tumor cells were almost all positive for CD68 (KP1, dilution 1:50; Dako) (Fig. 4B), α1-antitrypsin (Fig. 4C) (dilution 1:300; Dako), and α1-antichymotrypsin (Fig. 4D) (dilution 1:800; Dako), and partially positive for S100 protein (dilution 1:50; Dako) and α-smooth muscle actin (dilution 1:50; Dako). On the other hand, tumor cells were

Fig. 1. A: Bone-window CT scan revealing bone defect in the supramastoid occipital region. B and C: Computed tomography studies showing low density (B) with a homogeneously enhancing mass lesion (C). D and E: Axial Gd-enhanced T1-weighted (D) and coronal (E) MR images showing the skull tumor with homogeneous enhancement on the right sigmoid sinus. F: An FDG-PET study showing moderate uptake (arrow) in the tumor body.

Fig. 2. Right internal carotid artery angiograms (anteroposterior [left] and lateral [right] views) showing the occlusion of the junction between the right transverse and right sigmoid sinuses and hypoplasia of the left transverse sinus, which caused cutaneous or subcutaneous collateral veins (thick arrows) flowing into the external jugular vein (thin arrows).
negative for cytokeratin (AE1/AE3, dilution 1:50; Dako), desmin (dilution 1:80; Dako), and CD34 (dilution 1:50; Nichirei, Tokyo, Japan). These findings strongly indicated a histiocytic origin tumor. The MIB-1 (Ki 67, dilution 1:100; Dako) labeling index was calculated as the percentage of MIB-1–positive nuclei in total nuclei under a microscope, and was < 1% (data not shown).

Postoperative Course. Six months later, the patient had no symptoms, including double vision and headache. Neurologically, her abducens palsy and bilateral papilledema and the dilation of her retinal arteries were completely improved. Postoperative MR imaging and MR angiography findings showed that the tumor was completely eradicated and the patency of the right transverse-sigmoid sinus was preserved (Fig. 5).

Discussion

Primary BFH in bone is rare, with < 100 reported cases according to the WHO Classification of Tumors. It accounts for ~ 1% of all surgically treated benign bone tumors. The age of patients with BFH has been reported to range from 5 to 75 years, but the disease is most frequently seen in young adults. No sex predilection has been reported.

Benign fibrous histiocytoma usually occurs in the dermis, superficial subcutaneous tissue, and deep soft tissues and more rarely in parenchymal organs. According to several reports concerning BFH of bone, the tumors frequently arise in the pelvic bone, femur, and tibia, and less frequently in the jawbone, vertebrae, sacrum, and clavicle. To our knowledge, however, there have been only 5 reported cases of BFH of the cranial bone. There have been only 1 maxillary case and 4 mandibular cases in the viscerocranium of the skull, but there has been no report of this tumor occurring in the neurocranium. As described previously, many of these cases arose from long bones such as the femur, humerus, and tibia, and not in the metaphyseal portion.

One report mentioned that radiography showed a radiolucent lesion with extracompartmental extension and a somewhat irregular edge. Bone window CT scanning also showed a bone defect with thinning of the cortical bone. In the case reported by Katagiri et al., there was no specific uptake of FDG-PET. In our case, CT findings likewise showed a bone defect with thinning of the cortex, and also with defects. There have been no reports regarding MR imaging, enhanced MR imaging, contrast CT, or angiography findings for BFH, and there have been no reports of this tumor in the metaphyseal portion of the skull thus far, although 3 cases of MR imaging findings in the spine have been reported. In the present case, the tumor showed a homogeneous enhancement on CT scanning with contrast, Gd-enhanced MR imaging, and moderate uptake of FDG-PET. This is the first reported case providing a detailed modality evaluation of BFH of cranial bone and will be very useful for diagnosis in the future.

We had to carefully consider a surgical plan to prevent interception of collateral flow by the skin incision and craniotomy from further increasing ICP. We attempted temporary occlusion of the collateral scalp veins by manual compression of the scalp and confirmed that no symp-
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toms occurred. We used a neuronavigation system to detect subcutaneous and diploic veins, which was very helpful for making the skin incision and performing a craniotomy because it minimized the injury to the collateral scalp veins (Fig. 3 left). In fact, it was impossible to detect these veins using other tools, such as transcutaneous Doppler ultrasonography, and to feel them against the scalp. Moreover, since it was possible that the tumor was conglutinated to the surrounding tissue, such as the subcutis, skull bone, and the wall of the sigmoid sinus, we needed a meticulous strategy for safe and complete removal of the tumor. We first made a plan to expose the intact dura mater around the tumor by using the eggshell drilling technique of intact marginal bone with a diamond drill so as to detach it from the margin to the center of the tumor from the dura mater in a safer manner. We next adequately coagulated a small portion of residual tumor tissue on the sinus to avoid a recurrence. We did not try to detach the tumor by force in an attempt to avoid injury to the sinus wall because the tumor and wall were conglutinated strongly.

There have been several reports in which ICP was found to increase as a result of sinus occlusion by skull tumors such as Ewing sarcoma, neuroblastoma, carcinoma of the breast, plasmacytoma, and meningioma.9,12,15,17,18,20,22 The site of compression has been the terminal portion of the SSS and confluence of sinuses. Obstruction of these sites produces increased ICP through venous congestion throughout the length of the SSS. The increased ICP will develop if anastomotic channels between the SSS and elements of the cerebral venous system proximal to the site of obstruction are inadequate. This symptom would also occur when the tumor produces an extradural tissue mass beneath the inner table of the skull.21 In our case, although only the right transverse sinus was obstructed by the extradural bone tumor, an increase in ICP occurred because the left transverse sinus was too hypoplastic congenitally to compensate for the impaired venous perfusion, which subsequently gave rise to intracranial venous congestion. We strongly suspect that this was why collateral flow from the obstructed SSS via cutaneous, subcutaneous, and diploic veins draining into the external jugular vein developed.

It has been recognized for some years that meningiomas may occasionally produce increased ICP by occluding a venous sinus.17 It has recently been emphasized that unless the venous drainage is improved along with resecting the tumor, papilledema may persist and visual loss may continue.22 This phenomenon might correlate with the period of steadily increasing ICP. In our case, the period of increased ICP might have been relatively short, because the papilledema and bilateral abducens palsy were immediately improved after removing the tumor.

The histological characteristics of BFH in bone are similar to those of BFH occurring in soft tissue. These tumors included a proliferation of benign oval spindle cells resembling fibroblasts, intermixed with benign mononuclearized cells resembling histiocytes.24 Moreover, there is histological similarity between BFH of soft tissue or bone and nonossifying fibroma, and also between BFH and metaphyseal fibrous defect in childhood. In our case, proliferation of histiocytic foamy cells shown histologically and immunohistochemically was a predominant feature that could differentiate the tumor from a nonossifying fibroma. There was little nuclear atypia, pleomorphism, or atypical mitosis; in fact, the MIB-1 labeling index was < 1%. Moreover, intraoperative findings showed that this tumor was well demarcated from the surrounding tissue and detached with little adhesion. These findings indicated that it was benign with few invasive characteristics. Differential diagnosis from giant cell tumors, which usually show aggressive behavior and recur locally in as many as 50% of cases, was not difficult because multinucleated giant cells were not detected in this case.

No recurrence was seen 6 months after total removal of tumor. Complete tumor removal was very important to prevent tumor recurrence, because the tumor had the potential to recur locally, in which curettage and bone grafting were carried out.24 There is no reliable report in the literature in regard to a BFH becoming malignant. In the present case, although there were no malignant characteristics histologically, such as nuclear atypia, pleomorphism, and atypical mitosis, preoperative FDG-PET showed moderately strong uptake in the tumor body and tumor stain feeding from the right occipital artery on cerebral angiography, which might have been due to the high cell viability. Careful long-term follow-up clinically and radiologically is essential.

Disclaimer
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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References


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