A rare case of psammomatoid ossifying fibroma in the sphenoid bone reconstructed using autologous particulate exchange cranioplasty

Case report

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Psammomatoid ossifying fibroma (POF), a variant of ossifying fibroma, is a benign fibroosseous lesion typically arising within the nasal cavity, paranasal sinuses, and orbit. Cranial vault involvement is exceedingly rare, with very few cases reported in the literature. The authors report a case of POF in the neurocranium of an 11-year-old child, 4 years after chemotherapy and radiation therapy for acute lymphoblastic leukemia. This case is reported in view of its rarity, novelty of presentation, and the difficulty in diagnosis due to its radiological resemblance to aneurysmal bone cyst or monostotic cystic fibrous dysplasia, further aggravated by the clinical scenario. A novel technique of cranial reconstruction called autologous particulate exchange cranioplasty was used following tumor excision.

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Key Words • fibroosseous lesion • psammomatoid ossifying fibroma • tumor

**Ossifying** fibroma is a rare fibroosseous lesion that most commonly occurs in the craniofacial skeleton, with particular predilection for the nasal cavity, paranasal sinuses, and the orbit. Psammomatoid ossifying fibroma, also known as juvenile (active) ossifying fibroma, is a variant of ossifying fibroma containing numerous calcified “psammomatoid” ossicles that histologically resemble psammoma bodies. A more common and related variant is the cementifying or cementoossifying fibroma, which occurs in gnathic bones.

Very few cases of POF have been described in the literature; most of them involve the paranasal sinuses or the orbit. A POF primarily involving the neurocranium or the sinonasal region with intracranial extension is extremely rare. Clinical presentation of ossifying fibromas includes proptosis, facial swelling, nasal obstruction, visual or ocular motility disturbance, headache, and sinusitis, reflecting its frequent occurrence in the sinonasal location.

We describe an unusual case of POF involving the sphenoid bone in a child who presented with swelling in the left temporal region. The appearance of the lesion and its location presented a diagnostic challenge. We also present a novel technique of autologous cranial bone that provides many advantages over more conventional methods.

**Case Report**

**History and Examination.** This 11-year-old girl presented to our center with painless swelling in the left temporal region of the scalp that had continued for 2 weeks. She had no other complaints. Her medical history was significant for high-risk acute lymphoblastic leukemia occurring at the age of 6 years, which was treated with chemotherapy and 1200 cGy of cranial radiation. She had had no documented recurrence and had been asymptomatic. Examination revealed a hard, nontender, nonpulsatile swelling in the left temporal region. She had normal vision and globe position, and conjugate ocular motion.

**Neuroimaging.** Computed tomography scanning revealed a 3.3 × 3.6 × 3.5–cm expansile, lytic lesion in the left superolateral orbit involving the greater wing of the left sphenoid bone. The mass contained irregular hyper-
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Fig. 1. Bone window of a CT scan (A), and T2-weighted (B) and postcontrast (C) axial MR images showing a 3.3 × 3.6 × 3.5-cm expansile, multicystic, lytic lesion involving the greater wing of the left sphenoid bone and containing irregular hyperdense elements, predominantly hyperintense on T2-weighted imaging, with internal septations that have peripheral and septal enhancement and no intracranial or intraorbital extension.

Biopsy Findings. The patient underwent incisional biopsy to clarify the diagnosis. Histological studies revealed the presence of a POF characterized by a benign fibrous proliferation with admixed calcified “psammomatoid” ossicles. The cells were negative for EMA and S100 (markers for the histologically similar psammomatous meningioma) and had a low (< 3%) Ki 67 proliferative index (Fig. 2). Although the lesion was benign, a complete excision and osseous reconstruction were planned to prevent further expansion and to restore her normal facial contour.

Operation. The tumor was exposed through a coronal incision to allow complete access to the lesion and

Fig. 2. Photomicrographs of sections showing a benign fibrous proliferation with numerous calcified “psammomatoid” ossicles (A and B), a low Ki 67 proliferative index (C), and absence of EMA immunoreactivity (D). H & E staining (A and B), Ki 67 immunostaining (C), and EMA immunostaining (D).
full exposure of the contralateral parietal bone donor site. Intraoperatively, there was a completely extradural bony tumor containing a cavity filled with viscous material. The tumor was completely excised, with clean bone margins. The resection created a large bone cavity in the temporal area, and there was intact exposed dura mater (Fig. 3). We prefer autogenous bone reconstruction in the pediatric craniofacial skeleton to the use of titanium or synthetic constructs. Because this defect required a properly contoured structural graft to create symmetry in this visible area, we elected to repair the defect by using a modification of our previously described cranioplasty technique.9 The advantages of this approach have been previously discussed, but a major consideration here was the difficulty inherent in the successful creation of a split-thickness bone graft with a curved piece of bone in a relatively young child.

A template was made of the calvarial defect by using sterile cardboard. The template was then placed over an area of intact bone that has a similar contour, in this case the contralateral parietal region, and marked with a sterile pencil. The bone was removed in standard fashion. Particular bone was then harvested by using a hand-turned Hudson brace and a D’Errico drill bit. Graft material was obtained by drilling partial-thickness bur holes from the endocortex of the removed segment and the ectocortex of the adjacent parietal bone. The tip should be flat to prevent bone penetration. The bit is placed firmly against the stabilized bone surface and, as it engages the cortex, the axis of the brace is rotated in a gradually larger angular motion (precession) to produce more widening and to minimize the depth of the partial-thickness cortical defect. The graft is placed in a container with blood. The full-thickness parietal bone flap was then contoured to fill the defect perfectly, and stabilized using 1-mm-thick titanium miniplates. The harvested corticocancellous bone graft was then placed over the exposed dura at the bone donor site (at a depth of 5 mm) and stabilized with fibrin glue (Fig. 4).

Postoperative Course. The patient had an excellent outcome, with very good cosmetic reconstruction of the skull defect. Follow-up imaging demonstrated no recurrent disease and excellent healing at the donor site, with full thickness of the calvaria (Fig. 5).

Discussion

Diagnosis of an ossifying fibroma can be difficult, radiologically as well as pathologically. On CT scans, ossifying fibroma appears as a well-demarcated, expansile lesion of mixed lytic and sclerotic density with variable internal consistency, which may be multiloculated depending on the relative content of the mass, cystic changes, and the osteoid components.4,15,26 Similarly, the MR appearance of these lesions can vary depending on the internal architecture, with enhancement of the solid portions of the lesion.5,13,28 Whereas the sclerotic variant may be most difficult to distinguish from the monostotic form of fibrous dysplasia, the cystic one may closely resemble aneurysmal bone cysts.4,13,15,17,25,26 The presence of fluid levels in aneurysmal bone cysts, nonetheless, may be a useful differentiating feature, because these are usually absent in ossifying fibroma, as seen in the present case.17 Our patient had further diagnostic considerations due to the rarity of involvement of cranium by POF, and her history of leukemia and cranial irradiation, which further increased the diagnostic dilemma. Radiation has not been implicated in the literature as a risk factor for developing nonossifying fibromas. However, given that this mass developed directly in the field of radiation therapy (performed 4 years earlier), it is tempting to speculate that radiation may have played a role in the development of this lesion.

The histological appearance of POF is that of a benign fibroosseous proliferation composed of fibrous stroma, admixed bony spicules, and the presence of calcified “psammomatoid” ossicles that can vary in size and frequency. In contrast to fibrous dysplasia, osteoclasts and osteoblasts typically line the trabeculae, which are composed of entrapped lamellar bone. The stroma is frequently cellular, with fascicular-to-storiform groups of round-to-spindled cells that can have prominent nucleoli and scant cytoplasm. Mitoses are rare, and atypical mitotic figures are usually absent. The main differential diagnosis for ossifying fibromas is fibrous dysplasia, which in gnathic bones can be difficult to distinguish due to overlapping histological appearance; therefore, careful examination is required because prognosis and treatment differs. Absence of activating missense mutations of the GNAS1 gene in POF, which is seen in virtually all cases of fibrous dysplasia, can be of diagnostic significance and provides further evidence that these lesions are distinct entities.2,11,19 A POF can occasionally be mistaken for meningioma; however, the absence of EMA positivity, characteristic whorl formation, and intranuclear pseudoinclusions would not support a diagnosis of meningioma.21
Cases of EMA-positive POF have been described in the literature in combination with careful histological examination and further immunohistochemical studies; these entities can be easily distinguished. Secondary lesions, such as aneurysmal bone cysts, have been described in POF. Although the Ki 67 proliferation index is low in cases of ossifying fibromas, as was seen our case, it does not correlate with tumor behavior, because POFs have been known to behave aggressively.

POF is locally aggressive, with higher rates of recurrence compared with other variants. Although management of POF consists of total excision or enucleation, complete excision should be performed whenever feasible, considering the high recurrence rates, especially after subtotal excision, that have been reported in cases described in the literature. The tumor is said to show no radiosensitivity, and no malignant transformation has been reported. In fact, the tumor in our case developed after radiation: this brings into question the role of radiotherapy in its management. Whether association with aneurysmal bone cyst alters the prognosis is not known due to the rarity of such cases in the literature.

We resorted to biopsy sampling of the lesion initially, because a diagnosis of POF was not entertained preoperatively, especially in the setting of a previous malignancy treated with chemoradiation. The lesion was completely excised following the diagnosis of POF. We now have 1 year of follow-up, with no evidence of recurrence.

Reconstruction of the cranial defect following removal of the bony tumor is an important aspect of management. The options include autogenous bone graft using split calvarial graft, rib and iliac bone, alloplastic materials (for example, methyl methacrylate, porous polyethylene, titanium mesh), bone substitutes (for example, Norion, Bone Source), and biological options such as de-mineralized bone matrix and bone morphogenetic protein. Alloplastic materials or bone substitutes are not generally recommended for use in the pediatric craniofacial skeleton. First, these materials never osseointegrate, and do not grow with the cranium. Thus, they can dislodge or become unstable with the patient’s growth. Second, they are less able to resist compressive load than intact cranial bone. Third, they have a higher

Fig. 4. Intraoperative photographs showing the technique of exchange cranioplasty. A: Harvested parietal bone the size of the template. Harvesting of endocortical (B) and ectocortical (C) corticocancellous bone. Final view of the reconstructed skull defect at the site of tumor accomplished using normal parietal bone and miniplates (D), and filling of the parietal skull defect with corticocancellous bone graft (E).

Fig. 5. CT images demonstrating preoperative (left) and 3-month postoperative (right) coronal reconstructions that reveal resection of the tumor (arrowheads indicate the operative site) on the postoperative study and excellent, full-thickness reconstitution of the calvaria at the donor site (red circled area).
risk of infection, and some patients can have allergic reactions to these materials. Fourth, they are very expensive, and may not be an option in economically disadvantaged areas of the world. Biological materials are not recommended for large cranial defects, are relatively unproven in the craniofacial skeleton, can cause allergic reactions, and are extremely expensive. Autologous bone avoids most of the problems of other reconstructive techniques, but the lack of available bone in children, coupled with additional donor site morbidity, has dampened enthusiasm for this method. Split cranial bone is difficult to harvest in very young children, who do not have a diploic space, and split bone remains thinner and structurally weaker than full-thickness bone grafts. Other options, such as rib and iliac crest, are known to have more resorption than cranial bone.

The reconstruction described in this report is a modification of an autogenous cranioplasty technique we have used for some time. The advantages of this method are as follows: there is no additional donor site morbidity; the theoretical quantity of donor bone available is nearly double the area of the intact cranium; and no specialized expensive equipment is required, so there is no additional cost—thus, it can be used anywhere in the world. Technically, it is easier to perform this technique than to create a split-thickness graft, especially using curved pieces of bone and in young children. To date, there are no reported infections or allergic reactions, both donor and particulate graft sites heal to a thickness of normal bone (unlike split cranial bone), and the procedure can be used in very young infants even before the formation of the diploic space. The concept of exchange cranioplasty performed using particulate bone graft allows any recipient defect to be repaired with full-thickness structural bone without having to leave a large donor defect. We have previously used this technique with great success to repair large defects in situations in which the recipient site dura mater or operative site tissues were grossly abnormal, heavily scarred, or irradiated. In this case, we required a structural bone of sufficient strength and with the appropriate contour to fill the defect. The exchange allowed us to harvest the desired full-thickness parietal bone graft and still effortlessly fill the donor defect with nearly one-half centimeter of autologous graft. Further details of the technique and outcomes in craniofacial procedures can be found elsewhere.

Conclusions

POF remains a rare diagnosis in expansile bony lesions of the neurocranium. Awareness of its neurocranial presentation, and radiological and pathological findings may facilitate diagnosis and appropriate management. Good skull reconstruction following tumor excision is equally important.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Smith, Kasliwal, Rogers, Ramkisson, Kurek. Acquisition of data: all authors. Analysis and interpretation of data: Smith, Rogers, Ramkisson, Kurek. Drafting the article: Smith, Kasliwal, Rogers, Ramkisson, Kurek. Critically revising the article: Smith, Kasliwal, Rogers, Ramkisson, Kurek. Reviewed final version of the manuscript and approved it for submission: all authors. Administrative/technical/ material support: Smith, Moses-Gardner.

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