Isolated calvarial aneurysmal bone cyst in a pediatric patient: illustrative case

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BACKGROUND Aneurysmal bone cysts (ABCs) are benign, osteolytic lesions that can occur in long bones, vertebrae, or rarely, the skull. Here the authors present the case of a 15-year-old male with a primary ABC of the left frontoparietal skull along with a review of the literature to provide insight into the nature of this rare disease.

OBSERVATIONS An otherwise healthy 15-year-old male presented with a tense, painful lesion of the left frontoparietal scalp. He could not identify any inciting trauma, but first noted the lesion less than 2 weeks prior to presentation with progressive enlargement. Cranial imaging revealed a lytic skull lesion with fluid-fluid levels suggestive of ABC. Curative therapy was provided via wide excision of the lesion and calvarial reconstruction of the resultant skull defect. This was performed without complication, and histopathological evaluation confirmed the diagnosis of primary ABC.

LESSONS ABCs of the skull are rare entities and most often arise in the skull base versus the calvaria. Typically, these lesions are associated with an underlying bone pathology (secondary ABCs) but can be rarely seen as isolated lesions (primary ABCs). Clinical management consists of excision and adjuvant therapy for underlying pathology where appropriate.

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KEYWORDS aneurysmal bone cyst; calvarial lesion; lytic bone lesion; pediatric neurosurgery

Aneurysmal bone cysts (ABCs) were first described by Jaffe and Lichtenstein in 1942.1 They comprise 1% to 2% of primary bone tumors and are benign, osteolytic vascular lesions that grow rapidly.2 ABCs are most likely to arise in the second decade of life, with a mean age of 13 years, and occur equally in males and females.3 Most ABCs arise in the metaphysis of long or flat bones or within the vertebrae. They can also arise in the calvaria at a reported rate of 3% to 6%, most commonly in the skull base.1 Although the cause of ABC is unknown, trauma, underlying neoplasms, and cytogenetic abnormalities often contribute to the calvarial presentation.4 Here, we report the rare case of a rapidly progressive calvarial ABC without underlying pathology (primary ABC). A literature review is also presented to give context to this clinical presentation, etiology, and natural history of this rare disease.

Illustrative Case

A 15-year-old, otherwise healthy, male presented as an outpatient with a palpable lesion of the frontoparietal region of the skull. This lesion was first noted 1 to 2 weeks prior to presentation, and the patient denied any history of trauma. He endorsed pain on palpation of the lesion and with Valsalva maneuvers, chewing, or smiling. At the time of presentation, he and his family noted that the lesion had increased in size since first detection. The patient denied pain worsening at any particular time of day and had not tried over-the-counter pain medication for relief.

On examination, the patient had an approximately 1.5-cm raised lesion of the left parietal bone, which was firm but tender to palpation. No other palpable step-offs or cranial defects were detected. No changes in the overlying skin were noted. The lesion was nonpulsatile. He had an otherwise normal neurological and physical examination. Magnetic resonance imaging (MRI) of the brain had already been performed for further workup (Fig. 1), revealing a cystic expansile lesion within the diploic space of the left frontoparietal skull with inward bowing and thinning of the calvaria and possible complete erosion of the skull. There was no evidence of intradural extension; however, surrounding edematous changes in the overlying scalp and
temporalis muscle were noted. Given these findings, resection of the lesion with cranioplasty was recommended.

Three weeks after presentation, the patient was taken to the operating room for resection of the lesion and cranioplasty of the skull defect. Of note, the lesion was significantly larger and more tender and tense upon presentation to the operating room compared to his outpatient visit. The area was marked and prepared for resection, which was conducted without complication (Fig. 2). The lesion was noted to be cystic and filled with acute-on-chronic blood products. The lesion was subperiosteal in location, elevating the superior aspect of the temporalis muscle, and had completely eroded through the skull without invading the underlying dura. Bone edges immediately surrounding the defect were irregular and dysplastic, so the craniotome was used to expand the skull defect to more normal appearing bone. The residual 4 × 4 cm skull defect was repaired with Cranios Reinforced Fast Set Putty mix (Depuy Synthes), set over a Rapidsorb Rapid Resorbable Fixation System mesh plate (Depuy Synthes) that was positioned in the epidural space (Fig. 2). The patient tolerated the procedure without complication. Postoperative imaging was performed, which revealed complete resection of the cystic calvarial lesion and adequate repair of the calvaria (Fig. 3A–C). Pathological evaluation of the lesion demonstrated features compatible with primary ABC and associated skull remodeling (Fig. 3D). The patient was discharged on postoperative day 2. He was seen for follow-up 4 months postoperatively and was doing well with no complications and no evidence of recurrence.

FIG. 1. Preoperative coronal (A) and axial (B) T2-weighted MRI sequences, axial T1-weighted MRI sequence (C), and axial susceptibility-weighted imaging (D) demonstrated findings of a cystic calvarial lesion of the left frontoparietal skull. The lesion is expansile within the diploic space and demonstrates a blood-fluid level.

FIG. 2. A and B: Preoperative image demonstrating gross appearance of the progressively enlarging skull lesion. C: Intraoperative image of the resected cyst capsule. D: After the cyst was resected, an irregular skull defect was seen, with an intact underlying dura. E: Intraoperative photograph demonstrating the skull defect postreconstruction, prior to resuspension of the temporalis muscle and scalp closure.
Patient Informed Consent

The necessary parental informed consent was obtained in this study.

Review of the Literature

A PubMed search from database inception through February 2024 was conducted using combinations of the search terms “primary aneurysmal bone cyst” AND “skull” OR “calvarium/calvarial” OR “cranial.” Full-text articles published in the English literature were included if they reported on patients with primary ABCs located within the cranial vault. Reports of secondary ABCs or ABCs of the skull base were excluded (Fig. 4). This process yielded five articles reporting on six patients (age range 17 months to 27 years). Patients commonly presented with progressive painless swelling of the area, headaches, or mass effect from a hemorrhage after blunt trauma. Diagnostic workup generally consisted of computed tomography (CT) or MRI of the brain. Two articles described the use of angiography in three cases, with the use of preoperative embolization in one case. Details of the management of these patients and their outcomes are summarized in Table 1.

Discussion

ABCs of the calvaria are a rare entity. Approximately 30% of calvarial ABCs have been reported to occur in conjunction with associated pathology (secondary ABC), including osteoblastoma, chondroblastoma, giant cell tumors, osteogenic sarcoma, chondromyxoid fibroma, Langerhans cell histiocytosis, or fibrous dysplasia. Although primary ABCs are benign tumors of the bone, they can destroy surrounding bone tissue and enlarge rapidly, as in our illustrative case. ABCs of the skull present with varying symptoms depending on their anatomical location; an enlarging palpable mass, focal tenderness, headache, cranial nerve palsy, seizure, otitis media, vision changes, plosis or exophthalmos, cerebellar signs, bacterial meningitis, or increased intracranial pressure and hydrocephalus have all been reported in the literature. Reports of frontal, temporal, occipital, and skull base lesions have all been described, with skull base lesions more commonly reported.

Observations

ABCs have a characteristic appearance on imaging studies. On radiographs, an ABC will demonstrate a cystic, expansile, "blown out" appearance with a sclerotic rim. CT scans can show the bony anatomy in more detail, as well as any intracranial extension or associated findings. On CT, ABCs appear as multiloculated, osteolytic lesions with a classic "soap-bubble" appearance and fluid-fluid levels. On MRI, the multiloculated nature of the lesion is more apparent, with fluid-fluid levels within these cystic structures and a hypointense rim. Because the cyst contents consist of blood products at different points in degradation, these fluid components will appear with different signal intensities. The differential diagnosis for bony lesions with intracystic blood products includes chondroblastoma, giant cell tumor, osteosarcoma, and malignant fibrous

FIG. 3. Coronal (A) and axial (B) T2-weighted MRI sequences demonstrating complete resection of cystic lesion and repair of calvaria. Lateral plain radiograph of the skull (C) demonstrating the calvarial reconstruction at the site of resection of the lesion. D: Histopathological evaluation of the lesion revealed an irregular cystic space with a wall composed of some reactive fibroblast, multinucleated giant cells, and fragment of calcified reticulated martial. Hematoxylin and eosin, original magnification ×200.
Because calvarial ABCs are avascular, they typically appear angiographically occult, though reports of preoperative angiography and lesion embolization appear in the literature. ABCs also demonstrate a characteristic pathological appearance on histological evaluation (Fig. 3D). These multicystic, hemorrhagic, and well-circumscribed lesions will have multiple blood-filled cysts with fibrous septa made of spindle cells and osteoclast-type giant cells. Infiltration with inflammatory cells and the presence of immature bone formation are commonly observed. There is typically no evidence of malignant transformation, as these are benign lesions.

Although spontaneous regression of primary ABCs has been reported, clinical management of ABCs is usually necessitated by their associated symptoms, with the approach determined by the location of the lesion. The most common approach is resection with a wide margin, and this can be curative in easily accessible lesions. Preoperative embolization has been reported for large lesions, though this is typically not needed. Complete excision is often challenging with larger and skull base lesions. In these cases, other modalities, including postoperative radiotherapy, endovascular embolization, or sclerotherapy, may also be considered. Surgical reconstruction of the resultant skull defect must also be considered when planning the extent of lesion excision.

**Lessons**

Here, we presented the case of a 15-year-old male with a primary ABC managed with curative therapy via wide excision of the lesion and calvarial reconstruction of the resultant skull defect. This report highlights a rare pathology that should be considered when evaluating skull lesions in pediatric patients. ABCs of the calvaria are rare, lytic, multicystic lesions that can occur in isolation (primary ABCs) or in association with underlying bone pathology (secondary ABCs). Although the most common tender skull lesion in children is eosinophilic granuloma, ABCs should be considered in the differential diagnosis of lytic skull lesions in children, and underlying pathology should always be suspected until proven otherwise. Specifically, a tender mass lesion of the scalp of a child should be worked up with skull radiography, followed by CT if needed. MRI can be valuable in surgical planning, as it can provide

![Flow diagram](https://example.com/flowchart.png)
<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs)/Gender</th>
<th>Location</th>
<th>Presentation</th>
<th>Diagnosis/Workup</th>
<th>Management</th>
<th>Histology</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Phan et al., 2023&lt;sup&gt;8&lt;/sup&gt;</td>
<td>1.4/F</td>
<td>Lt orbit</td>
<td>2 wks Lt upper eyelid swelling, 1 day of esotropia &amp; epiphora</td>
<td>CT–osteolytic lesion w/ peripheral enhancement &amp; heterogeneous internal enhancement MRI–multiple cysts &amp; fluid levels w/ internal enhancing septae PET–increased uptake</td>
<td>Biopsy followed by GTR via bicoronal approach</td>
<td>Cystic hemorrhagic mass w/ intralesional solid &amp; cystic areas filled w/ blood &amp; lined by stromal &amp; osteoclastic cells; cellular component contained mononuclear stromal cells w/ both spindle &amp; ovoid nuclei &amp; multinucleate giant cells; no evidence of necrosis or malignancy; FGFR1-USP6 fusion noted on FISH</td>
<td>At 6 mos, mild Lt ptosis</td>
</tr>
<tr>
<td>Borni et al., 2022&lt;sup&gt;5&lt;/sup&gt;</td>
<td>7/M</td>
<td>Rt frontal</td>
<td>3 mos progressive painless swelling</td>
<td>CT–expansile extraaxial osteolytic heterogeneous mixed density mass MRI–multilocular cystic mass w/ soap-bubble appearance w/ fluid levels, no restricted diffusion, heterogeneous enhancement DSA–supply from ant &amp; pst frontal branches of STA</td>
<td>Planned GTR followed by cranioplasty</td>
<td>NA</td>
<td>Surgery postponed due to COVID-19; patient seen 2 wks later w/ regression of cyst &amp; 3 mos later, patient symptom free w/ no evidence of recurrence</td>
</tr>
<tr>
<td>Hermann et al., 2018&lt;sup&gt;7&lt;/sup&gt;</td>
<td>27/F</td>
<td>Rt frontal</td>
<td>2 mos progressive painless swelling w/ headache</td>
<td>CT–expansile extraaxial lytic lesion, multiloculated cysts w/ significant intracranial extension &amp; mass effect MRI–soap-bubble appearance w/ hemosiderin deposit &amp; intense septal enhancement PET–no enhanced metabolism</td>
<td>Frontal craniectomy w/ bone cement cranioplasty for GTR</td>
<td>Multiple cystic cavities containing variable numbers RBCs &amp; lined by fibrous septa; fibrous septa composed of loose fibrous tissue containing uniform population of plump fibroblasts or spindle cells; some osteoblast-like multinucleate giant cells also present in connective tissue; rearrangement of USP6 gene locus by FISH</td>
<td>Not reported</td>
</tr>
<tr>
<td>Garber &amp; Riva-Cambrin, 2015&lt;sup&gt;4&lt;/sup&gt;</td>
<td>3/F</td>
<td>Occipital</td>
<td>Fall, emesis, altered mental status, pst fossa EDH</td>
<td>CT–Lt pst fossa EDH overlying transverse sinus w/ mass effect; irregular bone loss around pst margin of foramen magnum &amp; occipital bone w/ associated soft-tissue mass</td>
<td>Emergent pst fossa craniectomy w/ GTR of lesion w/ evacuation of EDH</td>
<td>Large cystic spaces filled w/ blood &amp; separated by fibrous septa, alternating w/ more solid areas; clusters of multinucleated giant cells w/ loose, spindly stroma; no malignant osteoid &amp; no atypia</td>
<td>Neurologically intact at 1 &amp; 6 mos postop w/ no recurrence</td>
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TABLE 1. Literature review of cranial aneurysmal bone cysts

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs)/Gender</th>
<th>Location</th>
<th>Presentation</th>
<th>Diagnosis/Workup</th>
<th>Management</th>
<th>Histology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arthur &amp; Brunelle, 1988</td>
<td>6/F</td>
<td>Lt occipital</td>
<td>Rapidly enlarging swelling noted after minor head injury</td>
<td>Radiography–large osteolytic defect of occipital bone, diploic expansion, thinning of outer table, distorted foramen magnum CT–multiloculated intradiploic lesion w/ fluid levels &amp; enhancement of septae DSA–vascular mass supplied by muscular &amp; meningeal branches of lt vertebral &amp; lt occipital arteries</td>
<td>Surgical removal</td>
<td>Diagnosis of ABC confirmed</td>
<td>Not reported</td>
</tr>
<tr>
<td>9/M</td>
<td></td>
<td>Rt frontal</td>
<td>Hard, nontender swelling w/ rt ophthalmoplegia &amp; exophthalmos, papilledema, possible seizure</td>
<td>Radiography–large lytic defect of rt frontal bone w/ marked diploic expansion intracraniually CT–multiloculated mass w/ internal bony &amp; soft tissue septae, fluid levels; significant intradiploic expansion of lesion intracranially w/ mass effect &amp; obstructive hydrocephalus DSA–highly vascular lesion supplied by terminal branches of internal maxillary artery</td>
<td>Preop embolization &amp; surgery for GTR</td>
<td>Diagnosis of ABC confirmed</td>
<td></td>
</tr>
<tr>
<td>Present case</td>
<td>15/M</td>
<td>Lt frontal</td>
<td>1–2 wks progressive swelling &amp; pain</td>
<td>MRI–cyst expansile lesion over lt frontoparietal region w/ internal fluid level</td>
<td>Cranectomy for GTR w/ excision of narrow margin of normal bone w/ bone cement cranioplasty</td>
<td>Irregular cystic space w/ wall composed of some reactive fibroblast, multinucleated giant cells, &amp; fragment of calcified reticulated martial</td>
<td>No evidence of recurrence at 4 mo postop visit</td>
</tr>
</tbody>
</table>

ant = anterior; COVID-19 = coronavirus disease 2019; DSA = digital subtraction angiography; EDH = epidural hematoma; FISH = fluorescence in situ hybridization; GTR = gross-total resection; NA = not available; PET = positron emission tomography; pst = posterior; RBC = red blood cell; STA = superficial temporal artery.
information on the degree of lesion vascularity. Wide excision can be curative, but treatment is influenced by patient age, anatomical location of the lesion, and lesion size. In cases in which complete resection is not possible, adjuvant therapies can be considered.

References


Disclosures

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

Conception and design: Li, Ruge. Acquisition of data: Li, Saratsis. Analysis and interpretation of data: Saratsis. Drafting the article: Li, Saratsis. Final version of manuscript: Li, Saratsis. Approved the final version of the manuscript on behalf of all authors: Li. Pathology images: Asado.

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