Aneurysmal bone cysts (ABCs) are expansile, vascular bony lesions. The typical location for an ABC is the metaphyses of the long bones, although approximately 20% present in the spine. Calvarial ABCs are very rare and represent only 3%–6% of these vascular bony lesions overall. Aneurysmal bone cysts are most common in patients younger than 20 years, and a slight female predominance has been reported in the literature. Several theories exist regarding the origin of ABCs, with no one unifying theory present to explain the cause of these rare lesions. To our knowledge, a ruptured ABC as a result of head trauma has not been reported in the pediatric literature. In this case report, we review the imaging findings, natural history, clinical course, and treatment of these rare lesions.

Case Report

History and Presentation

A previously healthy 3-year-old girl was brought to our institution after falling off the monkey bars equipment at a playground and striking the back of her head. The patient vomited twice on the scene, and she was lethargic with a Glasgow Coma Scale score of 10 on presentation, with reactive pupils and symmetrical movements to stimulation. A CT scan obtained without contrast revealed a left-sided posterior fossa epidural hematoma (EDH) overlying the transverse sinus. There was effacement of the fourth ventricle and dilation of the temporal horns (Fig. 1). Irregular bone loss around the posterior margin of the foramen magnum and occipital bone, with an associated soft-tissue mass, was also noted (Fig. 2). The family was unaware of this lesion prior to this event. The CT scan was reviewed by a neuroradiologist and thought to represent an ABC, and because of the patient’s neurological compromise, an emergency MRI study was not obtained. Because of the often bloody nature of these lesions, the operating room staff was notified of blood type and screening results before the patient was taken to the operating room, and unmatched O-negative packed red blood cells were made available in case of urgent need.

Operation

The patient underwent an emergency posterior fossa craniectomy with gross-total resection (GTR) of the bony lesion and evacuation of the EDH. Intraoperatively, the mass had eroded the margins of the foramen magnum and was removed en bloc, although there was brisk bleeding...
necessitating blood transfusion. Once the EDH was evacuated, we closely inspected the transverse sinus for tears; no injury to the sinus was found as an alternative source for the EDH. All bleeding from the edges of the craniotomy was stopped with bone wax to help prevent air embolism. An intraoperative ultrasound confirmed that there was no subdural or intraparenchymal hemorrhage, nor was there obvious thrombosis within the venous sinus.

Postoperative Course

Postoperatively, the patient was extubated and taken to the pediatric ICU in stable condition. An MRI study of the brain and cervical spine was performed on postoperative Day 1, which was consistent with a GTR of the mass and decreased effacement of the fourth ventricle (Fig. 3). The patient was discharged home on postoperative Day 2 neurologically intact. On pathological examination, the lesion showed large cystic spaces filled with blood and separated by fibrous septa, alternating with more solid areas. There were also clusters of multinucleated giant cells with loose, spindly stroma, but no malignant osteoid and no atypia (Fig. 4). The final pathological diagnosis was a ruptured ABC. The patient has been seen at 1- and 6-month follow-up and remains neurologically intact. Follow-up CT scans and MRI studies confirm GTR and no recurrence at 6 months.

Discussion

Origin of ABCs

Several theories exist to explain the origin of ABCs. One theory hypothesizes that an inciting trauma induces a local circulatory disturbance that leads to the development of an ABC. Another theory is that ABCs are actually neoplasms with unique cytogenetic abnormalities. Approximately 50% of patients with these lesions show an abnormality on chromosome 16q or 17p. This is typically a chromosomal translocation that causes upregulation of ubiquitin-specific proteases, leading to increased cell-cell adhesion. A third theory posits that ABCs arise as a result of an underlying neoplasm that induces a vascular process such as a venous obstruction or an arteriovenous fistula. In fact, up to 30% of ABCs are associated with underlying...
ing tumors such as giant cell tumors, chondroblastoma, osteoblastoma, osteosarcoma, and fibrous dysplasia. A hormonal influence on the growth of these tumors has also been proposed, because they do tend to have a slight female predominance and have been shown to increase in size during pregnancy; however, these lesions have displayed negative results on immunohistochemical investigation for estrogen or progesterone receptors. Given that our patient had no history of previous head trauma and no underlying neoplasm was found on pathological or radiological examination, it is possible that the second theory may be most applicable in this case.

**Imaging Findings**

On CT scans, patients with ABCs of the cranium may demonstrate thinning of the cortical bone surrounding the lesion, resulting in focal cortical destruction. The cysts present in an ABC are separated by septae of varying thicknesses, and fluid-fluid levels are often seen because of hemorrhage inside the cysts. The intensity of an ABC on MRI varies depending on the stage of hematoma degradation. These cysts can also demonstrate peripheral enhancement with the administration of intravenous contrast.

**Presentation and Treatment**

Typically, ABCs present in patients during the 1st or 2nd decade of life. The most common presenting symptom is pain at the site of the ABC. When an ABC presents in the spine, epidural compression and cortical erosion can lead to scoliosis, fractures, or neurological symptoms from root or cord compression. To our knowledge, the patient described in this case report did not complain of any posterior neck pain or headaches localizing to the site of her ABC prior to her minor fall.

Hemorrhagic skull lesions in the pediatric population are extremely rare. Both intraosseous hemangiomas and ABCs can present as benign, enlarging skull masses, although typically intraosseous hemangiomas are painless, whereas ABCs are painful to the touch. Osteoid osteomas, which represent 10%–12% of osseous tumors in the pediatric population and are distinguished from osteoblastoma by size (< 1.5 cm), are also composed of highly vascular connective tissue, although these lesions are more typically found in the posterior elements of the cervical spine. As far as more malignant skull lesions are concerned, osteosarcomas of the skull base can present with hemorrhage, although these are more common in older patients (3rd–4th decade of life), whereas osteosarcoma of the long bones tends to present in adolescents. The rate of hemorrhage of these lesions is largely unknown.

The treatment of an ABC consists of GTR whenever possible. There is a reported 3% rate of malignant degeneration into osteosarcoma, and incomplete resection is associated with a recurrence rate of 71%. When treating an ABC of the spinal column, resection may be accompanied by spinal fusion to address destabilization of the spine after lesion removal; however, this is not relevant to our suboccipital case. Preoperative embolization is also an option because of the vascular nature of these lesions and their propensity for intraoperative bleeding. Because of the rare possibility that an ABC may degenerate into a more malignant entity such as an osteosarcoma, radiation alone is no longer recommended as a sole treatment for these lesions. Partial excision with adjuvant radiation may be required for lesions that are not amenable to GTR, such as ABCs located in the orbit, paranasal sinuses, or petrous temporal bone.

Other modalities that have been used with varying success for the treatment of ABCs include CT-guided percutaneous lesional injection of calcitonin and methylprednisolone. Calcitonin inhibits osteoclastic activity and promotes trabecular bone formation. Methylprednisolone has an antiangiogenesis effect. Additionally, the use of sclerosants such as alcohol has been tried; these act by direct damage to the endothelial lining, triggering a coagulation cascade and thrombotic occlusion of blood vessels. Because our patient displayed no residual lesion on postoperative imaging, the current plan is observation without adjuvant therapy. Another CT scan will be performed at an 18-month follow-up visit.
Conclusions

Aneurysmal bone cysts are rare lesions that typically present in the long bones or the spine in young patients. Several theories exist regarding the origin of these lesions, and treatment consists of GTR whenever possible. The presence of an ABC in the cranium is extremely rare in both the pediatric and the adult populations, and a ruptured ABC after head trauma has not been reported previously. We do not know whether these lesions are capable of spontaneous rupture, although there is an association with the development of ABC at the site of a prior trauma. Knowledge of the possibility that an ABC may rupture is useful for early recognition and diagnosis of this entity and prompt surgical treatment.

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References


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