Aneurysmal Bone Cysts of the Neuraxis: Part 1—Clinical Presentation and Pathogenesis

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Learning Objectives: After participating in this CME activity, the neurosurgeon should be better able to:
1. Evaluate the nature and pathogenesis of aneurysmal bone cysts (ABCs).
2. Identify the clinical presentation of ABCs.
3. Interpret the radiologic and histopathologic findings of ABCs.

Aneurysmal bone cysts (ABCs) are rare, benign, nonneoplastic lesions that occur in long bones such as the femur and humerus, or in the pelvic bones. They may also be seen in the vertebral column, but cranial (calvarial or cranial base) ABCs are rare. ABCs were first described as a distinct pathologic entity by Jaffe and Lichtenstein in 1942, after whom they were initially eponymously known. ABCs represent approximately 1% of all bone tumors, with an incidence of 0.14 per 100,000 individuals. They are usually sporadic, although rare familial cases have been reported. They may arise de novo or evolve from other preexisting lesions. ABCs are most commonly seen in patients younger than 20 years and are slightly more prevalent in women. Orthopaedic surgeons are familiar with these lesions and usually treat patients with a combination of curettage and allograft or autograft bone instillation into the cavity, with relatively good results. ABCs that involve the cranial bones or spine may necessitate a more complex management plan, particularly when there is epidural extension and neurologic compromise. In this 2-part series, we review ABCs from a neurosurgical perspective and discuss management options germane to these areas.

Pathogenesis

An ABC is an expansile, multiloculated, blood-filled lesion within bone that consists of anastomosing cavernous spaces, separated by thin walls that are composed of fibroblasts, macrophages, osteoclast-like giant cells, and islands of bone. ABCs weaken and deform the bone they occupy and may break through the bone cortical mantle impinging on surrounding structures. Sporadic ABCs usually form de

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appearance of an expanded contour is the result of bone pro-

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no novo (primary ABC), that is, in the absence of any precursor lesion within the bone, and they account for 70% of reported cases.

In the remaining 30% to 35% of patients, ABCs form as a result of a secondary vascular phenomenon superimposed on a preexisting lesion such as a giant cell tumor or osteoblastoma (secondary ABC). Other precursor lesions for secondary ABCs include tumors such as osteosarcoma, angiomia, chondroblastoma, fibrosarcoma, hemangioendothelioma, or metastatic tumors. In some instances, benign processes such as fibrous dysplasia, a solitary bone cyst, eosinophilic granuloma, radiation osteitis, or trauma may be implicated. The theory is that the primary or precursor lesion initiates formation of an intraosseous arteriovenous fistula or malformation, which via hemodynamic forces creates a secondary reactive bone remodeling process forming the ABCs. Some authors doubt this postulate, and there is some controversy regarding this theory of the origin of secondary ABCs, as it is rare to find neoplastic elements in an ABC at primary surgery and even at the time of recurrence.

Whether primary or secondary in origin, the formation of an ABC involves mechanical and vascular vectors, cystic degeneration of bone, and a remodeling process. It is hence not considered a neoplastic process. It is likely that intralesional hemorrhage contributes to further growth and initiates formation of reactive, poorly formed fibrous and osseous tissue that weakens the structure of the bone.

Four stages characterize the development of an ABC, and these may be visible on radiologic imaging studies. The first phase involves the development of a demarcated area of osteolysis, with discrete elevation of the periosteum. This is followed by a growth phase, in which the lesion grows rapidly with progressive destruction of bone and development of the characteristic “blown-out” radiologic appearance. The appearance of an expanded contour is the result of bone production by the periosteum, stimulated by the underlying inflammatory and reactive cells. The third phase is a period of stabilization, in which the lesion matures with formation of a bony shell; on imaging studies, this appears as a characteristic “soap-bubble” appearance. The final stage is one of healing and remodeling of bone characterized by progressive calcification and ossification, transforming the lesion into a dense bony mass.

**Clinical Presentation**

ABCs can grow rapidly and be locally destructive, compromising the structural integrity of the bone involved. Patients commonly present with pain and/or swelling in the region of the lesion. Symptoms and signs are usually secondary to bony decompensation and secondary compression of adjacent structures. Age at diagnosis or sex does not seem to influence the clinical presentation or natural history. With long-bone ABCs, patients may present with a pathologic fracture. This is more common in weight-bearing bones such as the femur or tibia. A similar process can also occur in the spine. Growth abnormalities may also be seen, as ABCs near the metaphysis may disrupt the growth plate.

Spinal ABCs are more common in adolescents and young adults. They are slightly more common in the lumbar spine than in the cervical and thoracic spine. The posterior elements of the spine (lamina, pedicle, spinous process, and transverse process) are commonly involved. Although extension into the vertebral body may be seen, the sole involvement of the vertebral body in the absence of posterior element involvement is rare. There is a risk of a pathologic fracture and a predilection to develop spinal deformities such as scoliosis and kyphosis. If the lesion breaks through the cortex into the epidural space, it may cause spinal cord or nerve root compression, resulting in neurologic symptoms such as paresis, paresthesia, or numbness. Involvement of 2 adjacent vertebrae may be seen at times. A solid variant of ABC in the vertebral column is also described, which may appear different from radiologic
perspective but they behave in a similar fashion from a clinical standpoint.

ABCs of the skull or cranial base are much less common, and they are less predictable in their presentation. Frequently, they are asymptomatic and may be incidentally discovered during imaging studies obtained for an unrelated event such as trauma. In other instances, they may present with scalp and skull bone pain and swelling. When located in the structures such as the orbit or mandible, they may present with proptosis or teeth numbness, respectively. Epidural extension can result in dural and cerebral or cerebellar cortex compression that may or may not cause neurologic symptoms.

Radiographic Findings

Radiographic findings of ABCs reflect the pathophysiologic process underlying their formation. Plain x-rays typically show an eccentric, osteolytic lesion with an expansile, remodeled “blown-out” contour surrounded by a thin shell of sclerotic bone representing normal bone cortex, which is thinned out over the lesion. Internal septa with fluid levels and sedimentation reflect hemorrhage within the lesion. Characteristically, ABCs are solitary lesions seen at the metaphyseal region of long bones or in the spine or pelvis. Spinal lesions generally involve the posterior elements but may extend through the pedicle into the vertebral body and intrude into the epidural or paraspinal space.

CT provides better anatomic delineation and visualization of intraliteral characteristics. Findings characteristically seen on plain radiographs are better demonstrated (Figure 1A). On CT scans, the density of fluid within the cyst is usually heterogeneous because of admixtures of degraded blood products and serous fluid. The expansile appearance and demarcation from surrounding soft tissue is well visualized, as are the thin septa and loculations lending to a classically described “soap-bubble” appearance. MRI also shows an expansile, septated intraosseous lesion with fluid levels and thin demarcation from surrounding soft tissues. This demarcation is low signal on both T1- and T2-weighted sequences, which pathologically correlates with fibrous tissue that surrounds the lesion (Figure 1B). Bone scintigraphy shows increased radionuclide uptake, and arteriography demonstrates hypervascularity.

Although these findings are highly suggestive of an ABC, they are not pathognomonic, and definitive confirmation relies on surgical biopsy and histopathologic correlation. Other lesions that must be included in the differential diagnosis are telangiectatic osteosarcoma, giant cell tumor, osteoblastoma, eosinophilic granuloma, unicameral bone cyst, fibrous dysplasia, chondroblastoma, chondrosarcoma, chondromyxoid fibroma, or Ewing tumor. In older patients, metastatic carcinoma or myeloma may present with similar findings.

A solid variant of ABC was first described in 1983 by Sanerkin et al. as characterized by florid fibroblastic or fibrohistiocytic proliferation, osteoblastic differentiation with osteoid production, areas rich in osteoclast-type giant cells, aneurysmal sinusoids, and occasional foci of degenerate calcifying fibromyxoid tissue. This solid variant of ABC is rare, accounting for 3.4% to 7.5% of all ABCs, and may be easily misdiagnosed as an osseous neoplasm. It has also been noted to occur in the spinal vertebral column, almost exclusively in children. On plain x-rays, it is indistinguishable from the classic ABC demonstrating an osteolytic, expansile lesion. Contrast-enhanced CT and MRI are more useful; although heterogeneous fluid-fluid levels may be seen, the septations are usually lacking; rather, a homogeneous pattern of enhancement may be observed.

Figure 1. A, Axial CT scan demonstrating the expansile appearance, and demarcation from adjacent brain parenchyma, of a left posterior fossa aneurysmal bone cyst. B, Axial T2-weighted MRI study of the brain demonstrating a well-demarcated, expansile, septated lesion within the posterior fossa with the characteristic soap-bubble appearance of an aneurysmal bone cyst, with no evidence of vasogenic edema or brain invasion.
One tumor that deserves special mention in the context of ABCs is osteoblastoma, which can mimic an ABC on imaging studies and also tends to occur in younger patients in long bones and the vertebral column. Within the spine, both ABC and osteoblastoma form expansile lesions that involve the dorsal vertebral elements. An osteoblastoma is simply a larger (>2 cm) variant of an osteoid osteoma; both are benign bone-forming tumors differentiated only by size. The trabeculae in osteoblastomas are interconnected and arranged in a haphazard fashion and surrounded by reactive bone. Central calcification may be seen, and the stroma between the trabeculae are filled with vascular loose connective tissue containing dilated capillaries, but without the large cavernous spaces seen in ABCs.

**Histopathologic Characteristics**

ABCs consist of multiple cystic blood-filled spaces lined by endothelium and separated by thin septa composed of spindled fibroblasts, osteoclastic giant cells, and reactive woven bone (Figure 2). This woven bone is surrounded by a rim of osteoblasts arranged in a rather regular pattern that follows the contours of the fibrous septa. The bone produced is basophilic and appears as a bluish osteoid matrix in either a trabecular or lace-like pattern. The presence of proliferating fibroblasts and inflammatory cells suggests a reactive process. The stroma of an ABC contains spindle cells with osteoclast-like giant cells distributed sporadically around channels that resemble dilated capillaries; they contain an endothelial lining but lack the muscular or elastic walls seen in larger blood vessels.

The osteoclast-like giant cells in an ABC seldom contain more than 50 nuclei, differentiating those from a giant cell tumor that may have up to hundreds of nuclei. Mitotic figures and atypical cells are rare, supporting the hypothesis that ABCs are a reactive vascular rather than a neoplastic process. The presence of atypical cells or mitotic figures should increase suspicion for osteoblastoma or osteosarcoma. Calcification is seen in about one third of ABCs and as mentioned above. In the solid variant of ABC, the typical fibrogenic stroma with osteoid production and benign giant cells is seen but the cavernous blood-filled channels are attenuated or nonexistent.

Although these features are suggestive of an ABC, they are nonspecific. Other lesions may mimic these findings. The notable ones to carefully distinguish are the unicameral bone cyst, giant cell tumor, and osteoblastoma, as these can have a similar radiographic appearance. A unicameral bone cyst contains constituents of the surrounding tissue and lacks giant cells lining the cavernous areas. Giant cell tumors may contain a substantial amount of grossly cystic tissue with typical ABC features, but are such a lesion is considered a giant cell tumor with secondary ABC changes rather than a secondary ABC. Osteoblastomas can be hemorrhagic and show secondary ABC changes; conversely, because primary ABCs can show extensive solid components and reactive new bone production, it can become difficult to make a distinction. Osteoblastomas usually demonstrate a discrete area of the tumor without cystic changes, which gives a clue to the true lesion etiology. These areas will have a different pattern of osteoid deposition. However, there may be cases in which the secondary ABC has obliterated the primary tumor, making it impossible to tell them apart. In either event, both lesions are benign and treated in the same fashion, thus the differentiation is of little clinical significance.

**Conclusion**

ABCs are benign, nonneoplastic lesions of the appendicular skeleton that may be encountered occasionally in the neural axis. They may cause pathologic fractures or extend into the epidural space to cause symptomatic compression of neural structures. As the radiologic and histopathologic findings of ABC may be mimicked by other, similar entities, careful interpretation of these studies, complemented by clinical correlation, is essential. This part I provides the reader with a basic understanding of the pathogenesis and clinicopathologic features of ABCs. In part II, we discuss the management of ABCs that involve the neuraxis.

**Readings**


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1. ABCs are composed of fibroblasts, macrophages, osteoclastic-like giant cells, and islands of bone.  
   True or False?

2. All ABCs form as a result of a secondary vascular phenomenon superimposed on a preexisting lesion such as a giant cell tumor or osteoblastoma.  
   True or False?

3. Spinal ABCs are generally more common in the lumbar region and tend to involve the posterior elements of the spine.  
   True or False?

4. On CT scans, the density of the fluid within the cyst is usually homogeneous because of admixtures of degraded blood products and serous fluid.  
   True or False?

5. On MRI sequences, an expansile, septated intraosseous lesion with fluid levels and thin demarcation from surrounding soft tissues is seen with ABCs.  
   True or False?

6. Radiographic imaging studies conclusively distinguish ABCs from other lesions such as giant cell tumor, osteoblastoma, or unicameral bone cyst.  
   True or False?

7. An osteoblastoma is simply a larger (>2 cm) variant of an osteoid osteoma; both are benign bone-forming tumors differentiated only by size.  
   True or False?

8. Mitotic figures and atypical cells are frequently observed in ABCs.  
   True or False?

9. Giant cell tumors may have histologic features similar to ABCs.  
   True or False?

10. On plain radiographs, ABCs appear as eccentric, osteolytic lesions with an expansile, remodeled “blown-out” contour surrounded by a thin shell of sclerotic bone.  
   True or False?