RADIOLOGIC CASE STUDY

Figure 1 and Figure 2 are spinal radiographs representing two different patients with the same condition. Your diagnosis?

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Aneurysmal Bone Cyst

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Figures 1 and 2 are different examples of aneurysmal bone cysts of the spine. In Figure 1 the right pedicle of L4 is expanded, whereas in Figure 2 the right pedicle of T12 is not visualized and appears to be destroyed.

Figures 3 to 8 are examples of aneurysmal bone cysts in various locations. Figures 3A through 3D are aneurysmal bone cysts in the spinal appendages. Figures 3C and 3D show an aneurysmal bone cyst in the spinous process of T6. In Figure 3C the spinous process is not visualized on the plain film but is markedly expanded on the CT scan as compared to Figure 3E, a CT scan of T5 on the same patient. The diagnosis of aneurysmal bone cyst must always be entertained in children and young adults, particularly on encountering a bone lesion that is markedly destructive, expansile and at the same time, fairly well demarcated.

Aneurysmal bone cyst, described originally by Jaffe and Lichtenstein,1,2 is characteristically an expanding “blow-out” lesion. The major long bones are commonly involved, particularly the femur and tibia. Although the diaphysis of a long bone may be affected (Fig. 5), the lesion is usually found near or in the metaphysis of the bone but it does not usually extend to the articular cortex.3 (Fig. 4). Flat bone lesions occur somewhat less frequently than in the long bones, common sites being the spine (mainly appendages) (Fig. 1, 2, 3), talus and calcaneus (Fig. 7), and clavicles (Fig. 8). Lesions occur rarely in the short tubular bones of the hands and feet (Fig. 6).

At the Cardinal Glennon Memorial Hospital for Children, in St. Louis, Missouri, a study of 20 surgically proven cases of aneurysmal bone cyst revealed an age range of five to 16 years with equal distribution between the sexes. Seventy-five percent of the lesions affected the major long bones, pelvic bones and spine. Equal numbers (four each) occurred in the femur, tibia and spine. These findings are similar to those described by other authors. Aneurysmal bone cysts are extremely uncommon over the age of 25 years.

Radiological Features

The radiological features may vary but the basic pattern usually consists of a benign, usually solitary, expansile, rapidly growing, lytic area in the involved bone, sharply demarcated by a very thin subperiosteal shell. The zone of transition between normal and abnormal bones is usually, but not invariably, narrow. A large, soft tissue mass may be present, with well-defined margins. Such changes are characteristic in the major long bones and in such flat bones as the innominate. In a long bone, the lesion tends to be eccentric, with a predilection for maximum growth beyond the outer margins of the bone. Occasionally, the expanding lesion may be central in location. A “soap-
Fig. 3A

Fig. 3B

Fig. 3C: Plain radiograph of an aneurysmal bone cyst in the spinous process of T6.

Fig. 3D: CT scan of an aneurysmal bone cyst in the spinous process of T6.
bubble” pattern is often observed within the lesion, caused by residual or newly formed ridges of cortex. A thin sclerotic margin may be present or occasionally a wide, densely sclerotic border may be observed.4

Lesions in the spine (the thoracic and lumbar spine are most frequently involved) tend to affect the appendages—lateral masses, pedicles, laminae, spinous and transverse processes frequently are markedly expanded. The vertebral appendage may be destroyed and the lesion may extend into the vertebral body (Fig. 1, 2, 3). Extradural blocks are an infrequent but feared complication of spinal lesions. Some observers feel that lytic, expanding lesions of the spine, diagnosed histologically as giant cell tumor, are in reality aneurysmal bone cysts.5

The radiological appearance of an aneurysmal bone cyst may, because of its markedly expansile, aggressive and destructive quality, simulate a malignant neoplasm. This is particularly true because, as in other potentially aggressive, benign lesions (giant cell tumor, pseudotumor of hemophilia), the zone of transition may be wide, and the large size of the bone lesion and its related soft tissue mass may suggest a malignant process.6 However, careful evaluation of the radiological appearance, particularly with reference to: 1) the preservation of a very thin, eggshell-like bony margin around the periphery of an aggressive, lytic expanding lesion; 2) the lack of a permeating destructive pattern; and 3) the presence of a large soft tissue mass with well-delineated borders, should suggest the correct diagnosis. Visualization of the thin shell makes the diagnosis almost certain; lack of it may cause this benign condition to simulate a sarcoma.
Clinical Symptoms and Signs

The clinical symptoms and signs of aneurysmal bone cysts are mainly those of pain at the affected site, generally of a relatively acute onset, with rapid increase in severity over a short period of time. Pathological fractures are frequent, particularly with markedly destructive and expanding lesions. An aneurysmal bone cyst in the spine may produce neurological deficits and even complete paraplegia. Myelographic examination will generally demonstrate an extradural defect and in advanced cases a complete "block."

Etiology

The etiology of aneurysmal bone cysts is unknown, but proposed explanations include: 1) arteriovenous communications within the bone; 2) development of a cyst in preexisting bone disease; and 3) subperiosteal hemorrhage with organization. An antecedent history of trauma to the affected part is not infrequently found in a young patient presenting with an aneurysmal bone cyst. A history of trauma is not uncommon with any bone tumor so that its importance is questionable. The opinion does exist that some cases of aneurysmal bone cyst may represent instances of post-traumatic hemorrhage. Yet, it is generally felt that this lesion is a distinct entity. Trauma could conceivably play a role in isolated instances, but the relationship to trauma is at best highly speculative and certainly unproved.

Pathological Findings

The gross pathological findings consist generally of a bulging shell of bone, lined by periosteum and even new
bone, containing large, distended, thin-walled, blood-filled, cystic cavities of varying sizes.

Microscopically, the blood channels are composed of fibrous connective tissue, osteoid, granulation tissue and multinucleated giant cells. They are usually devoid of muscular coats but are lined by endothelial cells (Fig. 9, 10). Phagocytes containing hemosiderin are often abundant. The histologic pattern is varied and some portions may be devoid of cystic spaces. The solid areas may be misinterpreted as giant cell tumor, fibrosarcoma or osteosarcoma, depending upon the predominant histologic component. Aneurysmal bone cyst-like areas are found in various benign lesions such as giant cell tumor, chondroblastoma, chondromyxoid fibroma and fibrous dysplasia and also in malignant tumors including osteosarcoma and chondrosarcoma.8,9

Conversely, telangiectatic osteosarcoma mimics aneurysmal bone cyst when viewed at low magnification, while higher magnification discloses the malignant cells.8,9 These pitfalls may be avoided by examination of adequate material and by correlation with radiographic and clinical data.

Treatment

Aneurysmal bone cysts are extremely vascular; however, the term aneurysmal refers to its gross appearance and not to its vascularity. If an aneurysmal bone cyst is surgically opened, a rapid, nonpulsatile flow of blood is obtained. Hemostasis can be extremely difficult and these lesions should not be biopsied or operated upon unless an adequate amount of blood for transfusion is available.

In general, aneurysmal bone cysts are treated by curettage and, if possible, packing with bone chips. Some, particularly those in the extremities, have been treated cryosurgically, by freezing with liquid nitrogen. Sometimes the mere performance of a biopsy may be sufficient to stop the rapid growth of an aneurysmal bone cyst and, in fact, result in the establishment of healing with new bone formation. Recurrence of aneurysmal bone cyst after treatment occurs in 10% to 20% of patients.9

References
Figs. 6, 7, 8: Statistically relatively infrequent sites for an aneurysmal bone cyst, although the appearances of the tumor are rather characteristic.

Fig. 6A: (Reproduced by courtesy of Dr. B. Akbarnia and Dr. F. Ghebadi.)

Fig. 6B

Fig. 7


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Fig. 8

Fig. 9: Blood-filled spaces are lined by flattened endothelial and multinucleated giant cells.

Fig. 10: Fibrous stroma displaying a fascicular pattern, and containing a few multinucleated giant cells.