ANEURYSMAL BONE CYSTS OF THE NEURAL AXIS

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In recent years, aneurysmal bone cysts have become recognized as rare but distinct clinical and pathologic entities that frequently involve the cerebrospinal axis. The benign nature and usually favorable response of this lesion even to incomplete removal make the diagnosis of aneurysmal bone cyst important so that overtreatment and an unnecessarily sinister prognosis may be avoided.

In 1942, Jaffe and Lichtenstein introduced the term “aneurysmal bone cyst” to designate a lesion previously called “atypical giant-cell tumor,” “angioma,” “pulsating benign giant-cell tumor,” “subperiosteal giant-cell tumor,” “benign bone aneurysm,” “hemorrhagic osteomyelitis,” “osteitis fibrosa cystica” or “central giant-cell tumor.”

Aneurysmal bone cysts are rare. They account for 1.5 per cent of all primary tumors of bone seen at the Mayo Clinic. They are located most commonly in the shafts of long bones and in the vertebrae, but they have been reported in practically all parts of the skeleton. In 9 of the 61 cases recorded at the clinic, the lesions affected vertebrae. The peak incidence is in the second decade of life. The incidence according to sex in reported cases is about equal.

The complaints elicited most commonly are mild localized pain, tenderness, limitation of movement and an enlarging mass that may or may not be tender. Examination often reveals a mass fixed to the underlying bone.

The roentgenologic findings in aneurysmal bone cysts in the vertebral column above the sacrum are typical as a rule. Part or all of the vertebral body may be affected, showing central rarefaction of the bone, which is expanded and shows only a thin shell of cortex remaining. The expansion, or widening, of the bone is often a striking feature and is evident even when great collapse of the vertebral body is present. Similar involvement often is seen in the laminae. When the spinous process is the site of the lesion, it often is greatly expanded and in some respects resembles a large egg, since the margin of the tumor consists of an extremely fine shell of bone. As the result of the collapse of the vertebral body and involvement of the neural arch, concomitant subluxation of the vertebra often occurs, and a gibbus may be present. When the sacrum is affected, the roentgenologic features are not characteristic as a rule, and the diagnosis depends on biopsy.

Giant-cell tumors are extremely rare in the spinal column above the sacrum, and any lesion in this location that looks as if it might be a giant-cell tumor is almost always an aneurysmal bone cyst or some other giant-cell “variant.”

Since a thin shell of bone almost always remains over the tumor, these lesions should not often be mistaken for malignant tumors. An interesting feature of aneurysmal bone cysts of the spinal column is that occasionally they affect adjacent vertebrae. The involvement of one vertebra is usually so great that it is easy to overlook the fact that adjacent vertebrae also are affected.

One tumor in this series involved the right half of the body of the 10th thoracic vertebra. The pedicle on that side was obliterated, and the right side of the vertebral body was completely destroyed without the usual thin
The gross surgical features of an aneurysmal bone cyst consist characteristically of an expansile lesion with a thin bony shell that, when removed, reveals a honeycombed mass of spaces filled with blood that usually is unclotted. Bleeding is brisk, but spurting is noticeably absent. Fleshy, granular or somewhat fibrous material may make up a prominent portion of the lesion.

Microscopically, the typical finding is blood-filled cavernous spaces that usually are not lined with endothelium and do not contain elastic laminae and muscle, as are seen normally in blood vessels (Fig. 2). The walls separating the spaces are fibrous and frequently contain osteoid or “fiber bone” and varying numbers of multinucleated giant cells. Benign giant cells may be especially numerous in the noncavernous zones, and their presence commonly has led to an erroneous diagnosis of “giant-cell tumor.”

As already emphasized, bona fide giant-cell tumors rarely are encountered in the vertebrae above the sacrum. Of 123 histologically verified giant-cell tumors in our files, 11 involved the sacrum but only 2 arose in the other vertebrae. Moreover, 90 per cent of giant-cell tumors occur in patients more than 20 years old, whereas most patients who have aneurysmal bone cysts are less than 20 years of age.

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Fig. 1. Typical aneurysmal bone cyst. An eccentric, well-circumscribed, osteolytic area is present in the involved bone, extending into the surrounding soft tissue. The lesion is limited at its periphery by a thin shell of subperiosteal bone. (Reproduced with the kind permission of the publishers from Adson.)

shell of bone being visible. A paravertebral soft-tissue mass was noted to the right of T10, and the logical roentgenologic diagnosis of a malignant tumor was made. The correct diagnosis could not have been made without biopsy.

Fig. 2. Aneurysmal bone cyst showing fibro-osseous trabeculae and large blood-filled spaces. Other zones contained numerous benign giant cells in the fibroblastic tissue (hematoxylin and eosin, X 50).