Chondromyxoid Fibroma With Secondary Aneurysmal Bone Cyst in the Cervical Spine*

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This case report describes a rare bony cervical tumor, chondromyxoid fibroma (CMF) which has features of an aneurysmal bone cyst (ABC). The true etiology of CMF and ABC is unknown. The aneurysmal bone cyst may be the result of a specific pathophysiologic change, which is probably the result of trauma or a tumor-induced anomalous vascular process. In approximately one third of cases, the preexisting lesion can be clearly identified. The most common of these is the giant cell tumor, which accounts for 19-39% of cases in which the preceding lesion is found. Other common precursor lesions include osteoblastoma, angioma, and chondroblastoma. Less common lesions include fibrous dysplasia, fibroxanthoma (nonossifying fibroma), chondromyxoid fibroma, solitary bone cyst, fibrous histiocyotma, eosinophilic granuloma, and even osteosarcoma. The treatment of the secondary ABC is based on the appropriate treatment for the underlying tumor. Complete local excision with tumor-free margins avoids the recurrence of CMF, the underlying tumor in this case report.

M.I. was a 27-year-old female administrative assistant. Her chief complaint was of right-sided neck pain with numbness and paresthesias radiating into the right upper extremity, gradually worsening over the preceding six months. The sensory abnormalities began proximally in the right shoulder and progressed to involve the right lateral arm, radial forearm, and eventually the thumb and index finger. She had sustained no memorable injury and her symptoms had failed to improve with non-steroidal anti-inflammatory agents (NSAIDs) and physical therapy. She described her neck pain as moderately severe (pain score of 4 on a visual analog scale of 0–10). The family history was significant for a malignant brain neoplasm, but the patient had an unremarkable medical history and review of systems.

Physical examination demonstrated no evidence of paraspinous muscular spasm or tenderness to palpation. Her cervical range of motion was normal and painless. Spurling’s sign was positive on the right side. Axial compression and facet loading caused no pain. Motor examination was intact. Sensory examination was intact to both light touch and pinprick. Reflexes were brisk 3+ and symmetric in both the upper and lower extremities. Tinel’s sign and Phalen’s sign were negative at both the wrist and elbow.

Imaging studies demonstrated an obvious abnormality. A lateral cervical radiograph demonstrated lucency in the base of the C6 spinous process (Figure 1). T2-weighted sagittal magnetic resonance imaging (MRI) showed lamina and right lateral mass of C6 extending into the neural foramen (Figure 2). T1-weighted axial MRI with gadolinium showed a homogenous pattern of enhancement in the C6 lamina and right lateral mass (Figure 3).

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Figure 1. Lateral cervical radiograph demonstrating lucency in the base of the C6 spinous process (arrow).
T1-weighted coronal MRI with gadolinium demonstrated contrast-enhancing epidural tumor extension (Figure 4). Computed tomography (CT) demonstrated evidence of a hypodense lesion causing diffuse expansion of the right C6 lamina with extension through the anterior cortex into the epidural space (Figure 5). There was obvious canal compromise and foraminal stenosis caused by a soft tissue mass. A chest radiograph (negative) and nuclear medicine bone scan were performed to rule out metastatic disease finding only abnormal signal uptake in the lower cervical spine.

The surgical intervention consisted of complete resection of the C6 lamina and right lateral mass, complete resection of the extradural cervical mass, posterolateral fusion at C5-C7, and posterior segmental instrumentation from C5 to C7. (Figure 6).

Coronal CT reconstructions demonstrated complete resection of the involved bony structures (Figure 7). Histopathological examination of the resected reddish purple epidural mass demonstrated no evidence of necrosis or mitotic figures on frozen section while permanent section showed groups of spindle-shaped or stellate cells
with abundant myxoid or chondroid intercellular material. Microscopic evaluation further identified a pale blue myxoid matrix containing intertwined strands of spindle and stellate cells with bland nuclei, finely dispersed chromatin, and inconspicuous nucleoli. These were rimmed by hypercellular areas that contained similar fusiform to spindle cells mingled with variable numbers of osteoclast-like giant cells. In certain areas, it appeared that there were small hemorrhagic cystic and cavernous spaces surrounded by fibrous septa composed of mildly to moderately mitotically active spindle cells intermixed with scattered osteoclast-like multinucleated giant cells. These findings were consistent with a chondromyxoid fibroma with cystic change and, in focal areas, degenerative conversion to aneurysmal bone cyst (Figures 8–10).

Nine months postoperatively, all numbness and paresthesias in the right upper extremity had subsided. The patient still experienced intermittent right neck soreness (which she attributed to fatigue and weather changes), but the prior neck pain had disappeared. Solid bony fusion was documented without any evidence of tumor recurrence.

DISCUSSION

Chondromyxoid Fibroma

Chondromyxoid fibroma (CMF), a rare benign primary bone neoplasm, was first described in 1948. It is
believed to be one of the least common bone tumors, accounting for less than 0.5% of all primary bone neoplasms. CMF may affect any bone in the body but typically involves the metaphysis of long bones, most commonly the proximal tibia. CMF is uncommon in the spine, with only 8 to 12% of all lesions located in the spine. A recent review found a total of 42 spinal CMF cases reported in the modern English literature. In the cervical spine, benign primary bone tumors most commonly occur at C2, C4, & C7.

The radiographic appearance of metaphyseal CMF is typical: oval, eccentric, with scalloped and sclerotic margins. However, the typical CMF radiographic appearance may not apply to vertebral lesions. Vertebral CMF causes extensive erosion of the cortex and extends beyond the peristeme into the spinal canal or surrounding soft tissue. The vertebral posterior elements are most commonly affected.

CMF histopathology shows “lobulated areas of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material separated by zones of more cellular tissue rich in spindle-shaped or round cells with a varying number of multinucleated giant cells of different sizes.”

The typical treatment of CMF by curettage has a 20–25% recurrence rate, possibly due to unremoved tumor lobules. Compared to curettage alone, curettage with concurrent bone grafting reduces the recurrence rate to 7%. Resection provides lower recurrence rates but is not always feasible depending on the location of the lesion. Spinal CMF with extensive local invasion and cord compression requires extensive surgery and tends to have a higher recurrence rate.

Aneurysmal Bone Cyst

Aneurysmal bone cyst (ABC) is also a rare benign condition, accounting for about 2.5% of all primary bone tumors. ABCs appear in any bone but most are found within the spinal column and large long bones. Within the spinal column, the posterior elements and the pedicles are affected first and, in 60 to 70% of the cases, the lesion extends to the vertebral body. The sacrum is affected in 13% of the cases, the lumbar spine in 31%, the thoracic spine in 34%, and the cervical spine in 22%.

In 20 to 30% of ABC cases, it is associated with an underlying skeletal lesion, such as giant cell tumor, osteoblastoma, hemangioma, chondroblastoma, nonossifying fibroma, fibrous dysplasia, CMF, telangiectatic osteosarcoma, and brown tumor of primary hyperparathyroidism. In large case series, areas of ABC were found in 7 to 8% of CMF cases. ABC causes cortical expansion but CMF does not necessarily do so.
Expansion of the bone is typically evident on x-rays. ABCs appear as blood-filled cavities separated by thin, fibrous septa made of fibroblasts, myofibroblast, multinucleated osteoclast-like giant cells, blood vessels, hemosiderin deposits, and fields of osteoid and woven bone.11

As was the case for CMF, the treatment choice is dependent on the location of the lesion and recurrence rates vary considerably with the type of treatment: 19% recurrence for curettage alone, 25% for subtotal excision and 60% for en bloc excision. Total excision must remove the entire cyst wall, all abnormal tissue that feels spongy, and bone surfaces that are lined with fragile and hypervascular membranes. Recurrence is due to the incomplete removal of the lesion, including the cyst wall.11

REFERENCES
