The case:

A 9-year-old boy presented with a 3-month history of cervical pain, limited range of motion, and a palpable “bump” in his posterior neck. After treatment with antibiotics for presumptive adenopathy and progression of the palpable finding, radiographs were obtained.

Figure: Anteroposterior (A) and lateral (B) cervical spine radiographs.

Your diagnosis?

For answer see page 141
Diagnosis: Aneurysmal Bone Cyst Involving the C2 Vertebra

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Clinical Presentation
A 9-year-old boy with a 3-month history of cervical neck pain and limited range of motion had an aneurysmal bone cyst (ABC) involving the C2 vertebra. Aneurysmal bone cyst (ABC) is a rare, non-neoplastic condition of bone that is named after its expansive blood-filled cavities. Aneurysmal bone cysts account for 1% to 2% of all primary bone tumors, having an estimated annual incidence of 0.15 case per 1 million individuals.1-3 The peak occurrence of ABC is during the first 2 decades of life (as in this 9-year-old boy), with 50% to 70% of ABCs occurring during this time.4,5 Most lesions occur eccentrically in the metaphyseal medullary canals of long bones (especially the distal femur and proximal tibia), and the spine—most commonly the cervical spine—is frequently involved.5 When the blood-filled cavities of the ABC quickly expand, osseous expansion and/or compression of surrounding tissues may lead to pain, swelling, and palpable findings.3,6 Lesions involving the cervical and thoracic spine can produce symptoms of spinal cord compression, prompting emergent clinical evaluation, imaging, and treatment.5

Pathology/Classification
The exact causation and pathogenesis of ABC is not clear.1 Aneurysmal bone cysts can develop independently (primary ABCs) or can originate from preexisting bone lesions (secondary ABCs). Primary ABCs account for approximately 70% of diagnosed cases of ABC. Aneurysmal bone cysts are grossly well-circumscribed lesions composed of blood-filled cystic spaces. Histological features include fibrous septa composed of dense cellular proliferation and reactive woven bone by osteoblasts.3 Secondary ABCs account for approximately 30% of diagnosed cases, with the majority of lesions occurring in conjunction with neoplasms such as giant cell tumor, osteoblastoma, chondroblastoma and osteosarcoma, or fibrous dysplasia.4,5,7,8 The most noteworthy genetic feature of ABCs is the characteristic rearrangement of the short arm of chromosome 17, most commonly occurring as a balanced translocation.3

Diagnostic Imaging
In general, radiography is the initial imaging technique for the diagnosis of bone tumors.8 Typical radiographic features for ABCs include a lucent lesion that “balloons” the bone (expansile remodeling), has a sclerotic rim, has no significant matrix mineralization, and is located in the eccentric metaphyseal medullary canal of a long bone or posterior elements of a vertebra (Figure 1).6 When ABC margins have a narrow zone of transition without a sclerotic margin, cortical loss, or extrasosseous soft tissue extension, the differential diagnosis may include giant cell tumor, aggressive osteoblastoma, and low-grade chondrosarcoma.4,5,8 When imaging features are inconclusive or a lesion involves the spine or a site with potential neurovascular implications, computed tomog-
Figure 1: Aneurysmal bone cyst. Anteroposterior (A) and lateral (B) radiographs of the cervical spine demonstrating a lucent lesion in the C2 vertebra (arrows) with diffuse expansile remodeling and sclerotic margin, but no cortical destruction or extraosseous soft tissue mass. The lesion more extensively involves the left portion of the C2 vertebra.

Figure 2: Change after aneurysmal bone cyst resection, C2-C4 anterior corpectomy, diskectomy, and fusion, and posterior occipital-cervical fusion. Lateral radiograph showing expected changes after C2 corpectomy for aneurysmal bone cyst, as well as C2-C4 anterior corpectomy, diskectomy, and fusion with bone grafting (asterisks), multilevel posterior decompression, and occipital-cervical fusion (arrows).

Figure 3: Change on computed tomography after aneurysmal bone cyst resection, C2-C4 anterior corpectomy, diskectomy, and fusion, and posterior occipital-cervical fusion. Sagittal 2-dimensional reconstructed (A, B) and 3-dimensional surface rendered computed tomography images in the frontal (C) and lateral (D) projections showing expected changes after definitive resection of the entire C2 body, solid incorporation of the cadaveric strut graft (asterisks), C2-C4 anterior corpectomy, diskectomy, and fusion (arrowheads), multilevel posterior decompression, and occipital-cervical fusion (arrows). No recurrent or residual osseous or soft tissue mass was present.

TREATMENT
There is no consensus on the treatment of ABCs. Several key considerations include ABC location, patient age, and the behavior of the tumor. The most common primary treatment methods are curettage or resection, in conjunction with bone grafting and/or hardware fixation. One of the main complications associated with the curettage/bone grafting method is pathological fracture due to weakened osseous infrastructure; therefore, vertebral lesions usually require fixation. Recurrence of ABCs is the other main complication. In a meta-analysis conducted by Cottalorda and Bourelle that compiled data from multiple studies involving 1134 cases of ABC, there was an average recurrence rate of 31% in curetted lesions. Adjuvant therapies have been studied with the intent of reducing recurrence rates. These include cryotherapy, heat cauterization, alcohol treatment, phenolization, radiotherapy, and argon beam coagulation. The 3 most efficient adjuvant therapies with the lowest recurrence rates are radiotherapy (13.5%), cryotherapy (12%), and argon beam coagulation (0%). Currently, radiotherapy is rarely used to treat ABCs because of possible radiation damage and known risk for secondary sarcoma formation.

Embolotherapy is much less commonly used for the treatment of ABCs. Embolization can be used selectively as the primary treatment of ABCs, for vascular occlu-
sion of lesions in anatomically restricted locations, or as a secondary treatment option in cases of recurrence. In general, embolotherapy should be avoided for vertebral ABCs because of the risk of cerebral or vertebral-ilar embolic phenomenon.

**FOLLOW-UP**

No universal protocols exist for recommended follow-up imaging, and studies are often based on patients’ symptoms. Immediate postprocedural radiographs can serve as a baseline for future comparison in cases of suspected recurrence or hardware failure. In the current case, follow-up radiographs demonstrated the expected postsurgical change: C2 corpectomy with complete ABC resection, multilevel posterior decompression, C2-C4 anterior corpectomy, diskectomy, and fusion, and posterior occipital-cervical fusion (Figure 2). Unenhanced CT was also performed and showed no recurrent or residual tumor, solid incorporation of the cadaveric strut graft, and no pathologic fracture or complication related to the hardware (Figure 3). This patient had a successful postoperative course, reported no complications, and remained free of tumor recurrence at 1-year follow-up.

**REFERENCES**