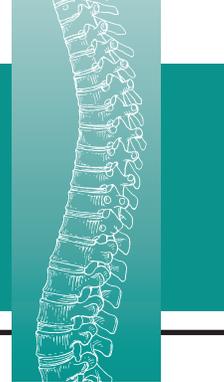


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Osseous Spinal Tumors Benign Neoplasms: Part II

Justin Munns, BS, Daniel K. Park, MD, and Kern Singh, MD

LEARNING OBJECTIVES: After participating in this activity, the surgeon should be better able to:

1. Describe the diagnostic work-up of primary tumors of the osseous spine.
2. Explain differential diagnosis of benign osseous spinal lesions.
3. Recall the common treatment principles for benign spinal tumors.

Tumors of the osseous spine represent unusual but important causes of back pain with or without neurologic symptoms. The severity and site of primary osseous spinal tumors vary immensely, demanding a high index of suspicion by the treating physician to ensure

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that disease progression is minimized. The following assessment will review the common clinical features, radiographic findings, treatment, and clinical course of the most commonly encountered primary benign neoplasms of the osseous spine.

CLINICAL APPROACH

The most common clinical finding in patients with primary tumors of the spinal column is back pain, although neurologic deficits and skeletal deformity can also occur. Pain often progressively worsens and is unrelieved by non-steroidal anti-inflammatory drugs (NSAIDs). Night pain remains a concerning symptom. Weakness and bowel or bladder dysfunction typically occur late in the course of progression.

Imaging remains critical in offering the patient a timely diagnosis. Anteroposterior (AP) and lateral x-rays can facilitate lesion identification, although radiographic evidence of bone destruction is not usually visible until 40% of trabecular bone has been destroyed.¹ Multiple patterns of bone destruction exist (geographic, moth-eaten, permeative), although a geographic pattern suggests a slower-growing benign lesion. CT is most effective in detecting cortical or intramedullary invasion. MRI, however, represents the preferred modality for detecting soft tissue invasion from an osseous lesion.

After imaging, obtaining an appropriate biopsy represents a critical step in spinal tumor management. Biopsies are not always necessary as radiograph-

ic and other staging procedures frequently allow the surgeon to perform definitive surgical therapy. If a biopsy is indicated, however, an incisional technique is most often used to assess the neoplasm. A direct incision within the potential field of surgery is critical so that the area can be incised during the definitive surgery. A variety of principles should be observed to ensure careful resection: transverse incisions must be avoided; tissues should be handled carefully; hemostasis should be maintained meticulously to prevent tumor spread; and bone should not be removed or windowed unless absolutely necessary.

A more detailed review of clinical management of benign osseous tumors of the spine has been described in part I of this article. Vascular and fibrous tumors, in particular, will be the focus of the current review, as bony and cartilage tumors were covered at length in part I.

BENIGN TUMORS

Table 1 lists the various benign osseous spinal lesions.

Giant-Cell Tumor

Giant-cell tumors (GCTs) are uncommon, locally aggressive tumors that are named for the osteoclastic giant cells that are intermixed with spindle cell stroma on pathologic samples. These tumors account for approximately 13% of benign osseous lesions of the

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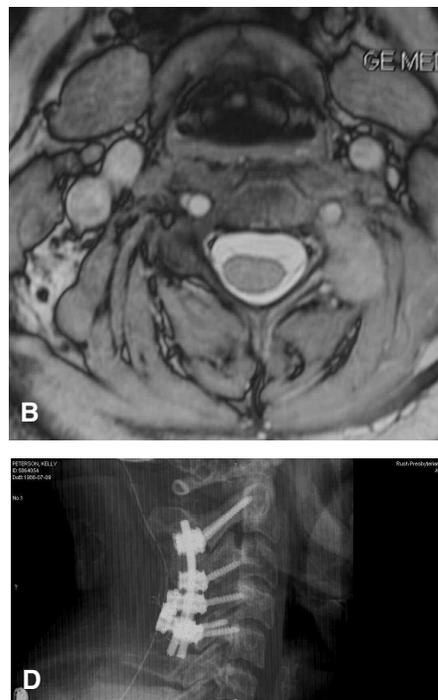
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Fig. 1 *A and B, 17-year-old patient with a giant cell tumor involving the entire left C3 and C4 lateral mass with extension into the left vertebral artery (arrow). Preoperative embolization was performed to occlude the left vertebral artery. The left C3–C4 lateral mass and vertebral artery were removed en bloc. C and D, The spine was reconstructed with a posterior spinal fusion from C2–C4.*

spine; the vast majority arise in the sacrum, where they account for 71% of benign sacral tumors.^{2,3} GCTs frequently affect the vertebral body more often than the posterior spinal elements. Women are affected two times more fre-



quently than men, and patients are typically between 20 and 40 years of age at presentation (Figure 1). Nonspecific pain is characteristic, but radicular symptoms have been described in 33% of cases.⁴ In patients with spinal and sacral GCTs, there is often a significant delay between onset of symptoms and diagnosis, due to the lesion's slow-growing nature. Hormonal stimulation is thought to contribute to the dramatic increase in size that accompanies GCTs that occur during pregnancy.⁵ Spinal and sacral GCTs often grow to large sizes and appear very vascular, which complicates resection.

X-rays often demonstrate an expansive, lytic lesion without mineralization or a sclerotic margin. Sacral lesions might be difficult to visualize on x-rays. Because of their large size and soft tissue component, they can be mistakenly interpreted as malignant neoplasms.⁶ Compression fractures are often present with vertebral lesions above the sacrum.⁷ On MRI scans, the lesion shows heterogeneous signal intensity, with low to intermediate signal intensity on both T1- and T2-weighted images due to the presence of collagen and hemosiderin. A low-signal pseudocapsule and areas of hemorrhage or cyst formation are common findings.⁶ CT scans can assist in identifying bony invasion in preparation for surgery. As many tumors



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appear similar in appearance on images, CT-guided core biopsy is indicated before excision.

First-line treatment for GCTs is en bloc surgical resection with wide margins, although spinal (especially sacral) GCT lesions are notoriously difficult to resect. Preoperative angiographic embolization is usually undertaken, given the

notable vascularity of these tumors.^{8,9} For unresectable lesions, radiation therapy and embolization are chosen, although newer treatment methods, including stereotactic radiosurgery, are still being investigated.⁹⁻¹¹ In a literature review, Leggon and colleagues noted that in a series of 159 patients, 51% experienced tumor recurrence when treated

Table 1. Benign Osseous Spinal Tumors

Tumor	Nature	Location	Level	Frequency	Peak Age (years)	Sex	Common Presentation	Imaging Findings	Treatment
Osteochondroma	Benign	Usually posterior elements	C>T	Rare (4% of solitary spinal lesions)	20–40	M>F	Myelopathy, mass	Continuity of lesion with marrow and cortex of underlying bone	Surgical excision; cartilage cap removal to prevent recurrence
Osteoid osteoma	Benign	Posterior elements (75%)	L>C>T>S	Very common in long bones, 10% occur in spine	10–20	M>F	Pain worse at night but relieved by aspirin	Round, discrete radiolucent nidus surrounded by variable sclerosis	Aspirin/NSAIDs for pain; surgical excision for definitive treatment
Osteblastoma	Benign	Usually posterior elements	C, L	10% of primary osseous spine lesions	10–30	M>F	Dull pain, neurologic deficits	>2.0 cm expansile lesion with bone destruction, variable appearance on imaging	Surgical excision; percutaneous resection, RF ablation, alcohol ablation if safely accessible
Chondroblastoma	Benign	Both anterior and posterior; spinal invasion common	C>L, T, S	Rare	10–40	M>F	Pain, spinal stiffness	Aggressive osteolytic lesion w/surrounding sclerosis, calcifications	Curettage, resection (total vertebrectomy most common)
Giant-cell tumor	Benign	Sacrum, vertebral body> posterior elements	S>L, T, C	13% of benign osseous spine lesions	20–40	F>M	Pain, radiculopathy	Expansile, osteolytic lesion w/o calcification	Resection ± radiation; preoperative embolization
Vertebral hemangioma	Benign	Vertebral body	T>L>C>S	10%–12% of adult population	20–50	M>F	Pain, radiculopathy, myelopathy	Corduroy pattern on x-ray; spotted appearance on CT	None if asymptomatic; surgical decompression, vertebroplasty, embolization, radiation, or ethanol if symptomatic
Aneurysmal bone cyst	Benign	Posterior elements, with frequent expansion	L>C, T	15% of primary osseous spine tumors	<20	F>M	Progressive pain, palpable mass	Well-defined, radiolucent expansile lesion on x-ray; egg-shell calcification on CT; fluid-fluid levels on CT/MRI	Resection ± radiation, embolization, injection of fibrosing agents
Eosinophilic granuloma	Benign	Vertebral body	T>L, C	70% cases of LCH	<20	M>F	Pain, neurologic deficits, systemic symptoms	Vertebra plana; lytic lesion leading to vertebral collapse	Observation, resection, radiation, chemotherapy; results similar regardless of therapy

C, cervical; F, female; L, lumbar; LCH, Langerhans cell histiocytosis; M, male; NSAID, nonsteroidal anti-inflammatory drug; RF, radiofrequency; T, thoracic; V, vertebral.



Fig. 2 CT scan of a vertebral hemangioma identified in a 29-year-old woman after a fall. *A*, Sagittal view demonstrates a space-occupying lesion in the T12 body and posterior elements, leading to pathologic fracture. *B*, Axial view shows “polka-dot” appearance in the vertebral body typical of hemangiomas. (Images courtesy of Kern Singh, MD.)

with radiation alone compared with 49% when incomplete resection was combined with radiation therapy.¹²

Vertebral Hemangioma

Vertebral hemangiomas are self-involuting tumors of endothelial cells that are localized to the vertebrae, often in the lower thoracic and upper lumbar spine, in 10% to 12% of the adult population. Most of the hemangiomas represent incidental findings on imaging, as only 1% of lesions are symptomatic.^{13,14} Patients present most often during the fifth decade of life, and there is a 2:1 male-to-female predominance.¹⁵ Pregnancy, however, is a recognized risk factor for a rapid onset of symptoms from these normally silent lesions.¹⁶ Most hemangiomas affect the vertebral body, rarely with posterior extension. Symptoms include local pain, radicular pain, myelopathy, and biomechanical instability from vertebral destruction.

The classic radiographic appearance of vertebral hemangiomas includes diminished vertebral density along with coarse vertical trabecular striation, forming a “corduroy” pattern.¹⁵ Enlargement of trabeculae along lines of stress in the vertebral body is responsible for this pathognomonic appearance. On axial CT scans, the thickened trabeculae in the vertebral bodies produce a series of dots, forming the classic “polka dot” appearance

(Figure 2). On MRI scans, the thickened trabeculae can be seen as linear low-signal areas oriented vertically on all pulse sequences. Hemangiomas enhance with contrast and have a high signal on T1-weighted images, because of fat within the lesion, and high signal on T2-weighted images from slow flow through the vascular component.¹⁷

Treatment varies according to the presenting symptoms. Asymptomatic hemangiomas form the vast majority of lesions and do not require treatment. Symptomatic pain without nerve root or spinal cord compression can be treated by a variety of therapies including transarterial embolization, radiation, direct ethanol injection, or vertebroplasty.¹⁸ If impingement is present, however, decompressive surgery with laminectomy is warranted (Figure 3).¹⁹ Significant bleeding represents a frequent complication.²⁰ Embolization can be used to reduce intraoperative bleeding, and radiation therapy given postoperatively can reduce the risk of recurrence.

Aneurysmal Bone Cysts

Aneurysmal bone cysts (ABCs) are expansile, osteolytic, blood-filled lesions that usually arise in long bones but originate in the spine in 9% to 20% cases and account for 15% of primary spine tumors.^{21–23} ABCs have been likened to blood-filled sponges, as the lesions consist almost entirely of multiloculated spaces replete with blood. The lesions are typically solitary, and they occur in patients under age 20 in 80% of cases.²⁴ A slight female predominance exists. Lesions most often affect the thoracic or lumbar spine, often arising from the posterior osseous elements. Notably, between 30% and 50% arise from within other preexisting lesions (GCT, chondrosarcoma, etc.).⁶

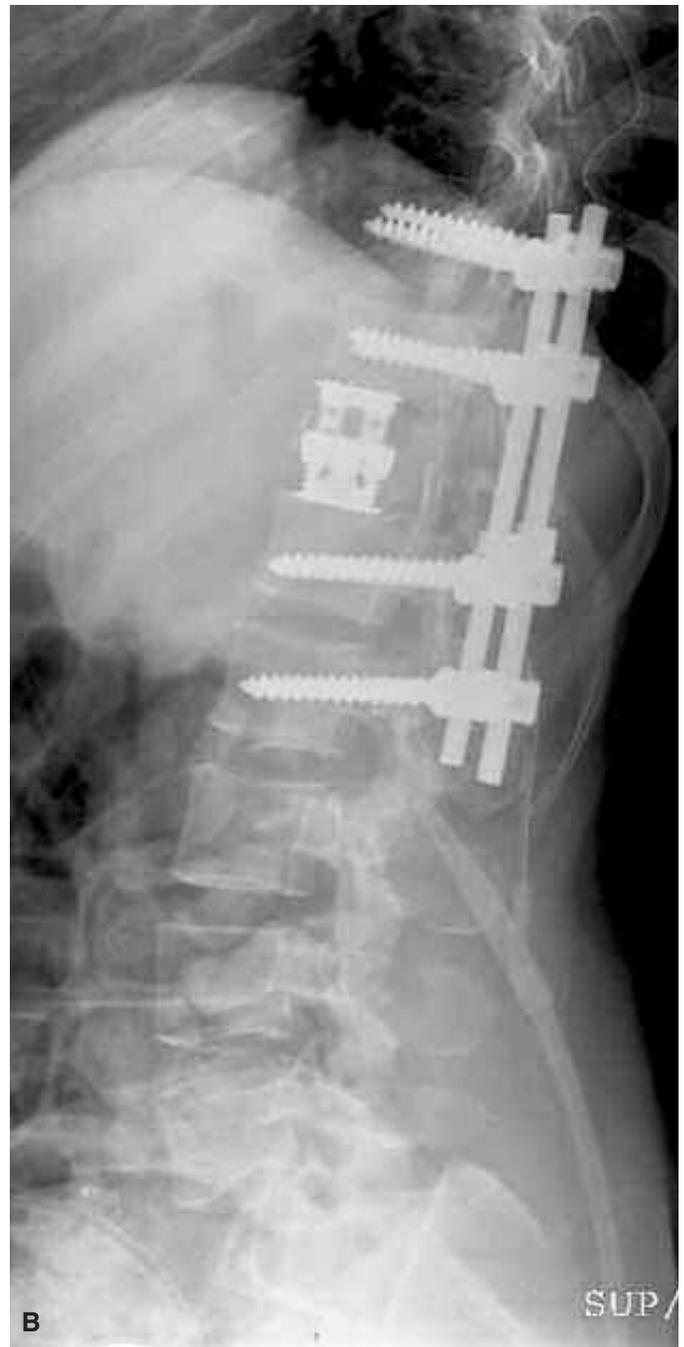
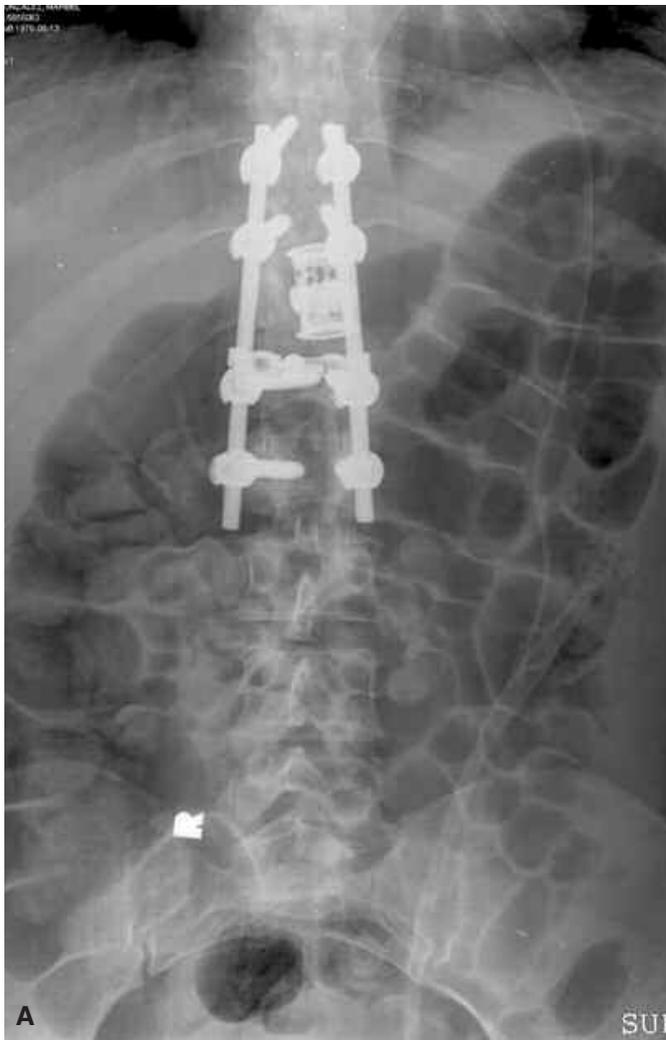


Fig. 3 Postoperative AP (A) and lateral (B) x-rays following decompressive surgery for vertebral hemangioma. Instrumentation spans the site of resection at the T12 vertebral body, where an expandable cage was placed via an all-posterior extra-cavitary approach. (Images courtesy of Kern Singh, MD.)

Presenting symptoms include progressive pain, palpable mass, and neurologic deficits.²⁵ Although they are benign lesions, ABCs often expand into the vertebral body, soft tissue, and adjacent vertebral bodies to cause radiculopathy or myelopathy.

On radiographic images, ABCs demonstrate a well-defined, radiolucent, expansile, and often trabeculated lesion surrounded by a thin sclerotic margin. Soft tissue extension or vertebral body compression fracture is possible. CT scans reveal a typical expansile, lytic lesion with a classic “eggshell” layer of surrounding cortical bone.²² Both CT and MRI detail the cystic nature of the lesion, with single or multiple fluid–fluid levels that indicate hemorrhage with sedimentation. Fluid–fluid levels are sensitive (87.5%) and specific (99.7%) for ABCs on CT scans, but not on MRI scans (77% and 67%, respectively).^{26,27}

Because of the aggressive nature of lesion growth, timely management is essential. Treatment of ABCs depends on size

and location of the lesion. Choices include arterial embolization, percutaneous intralesional injection of a fibrosing agent, radiotherapy, intralesional curettage (with or without bone grafting), and en bloc resection. No prospective studies have proven the superiority of one method over another, and most series describe a combination of these therapies.^{23,28} Careful preoperative arterial embolization is frequently accomplished due to the high vascularity of these lesions and their potential for high intraoperative blood loss. Radiation is provided if intralesional resection must be performed, as failure to do so is associated with higher recurrence rates.²⁹ Recurrence rates are variable but lowest for those undergoing en bloc resection, as reported by Mankin and colleagues in a series of 150 cases (Figure 4).²⁸

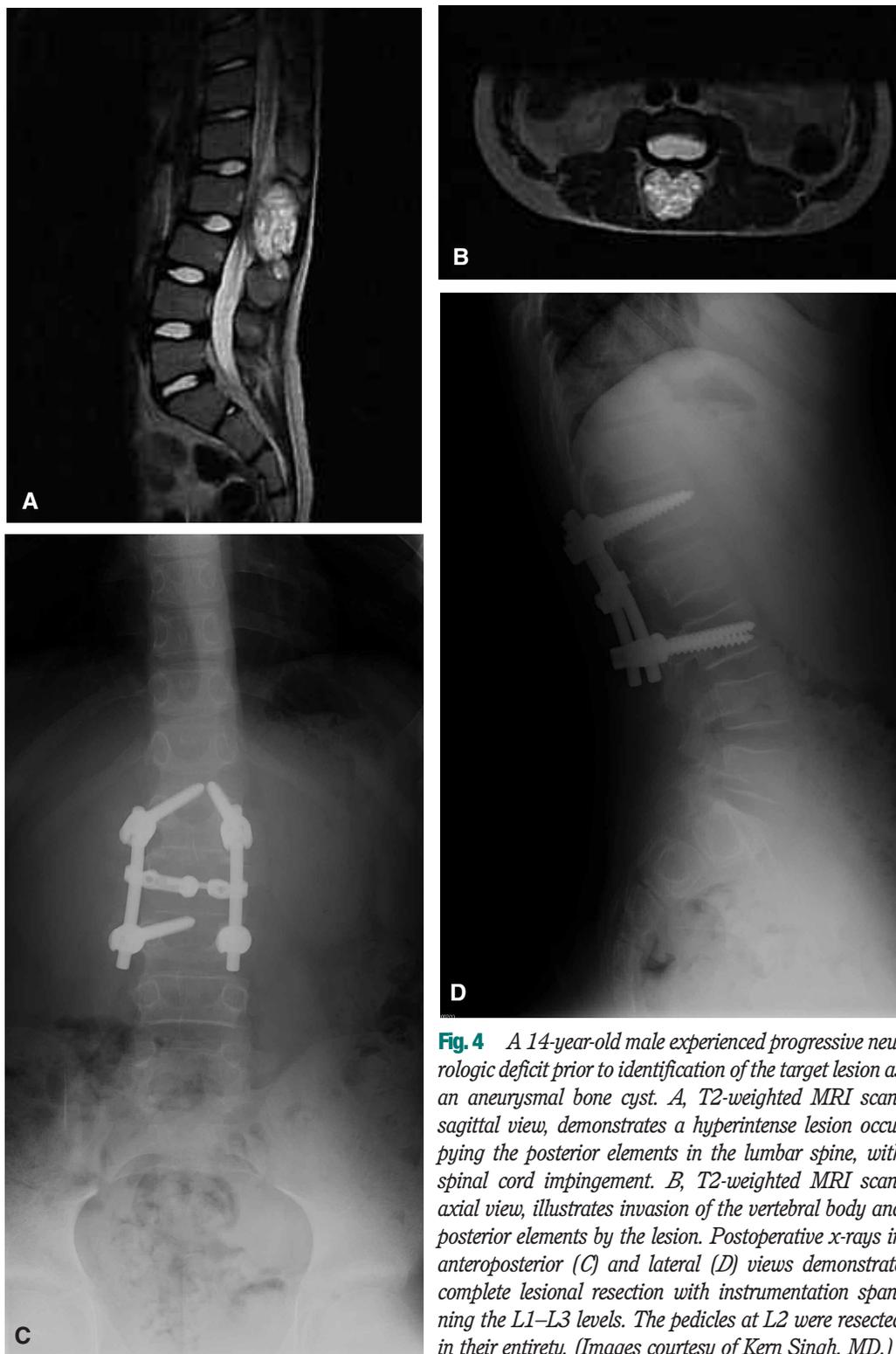


Fig. 4 A 14-year-old male experienced progressive neurologic deficit prior to identification of the target lesion as an aneurysmal bone cyst. *A*, T2-weighted MRI scan, sagittal view, demonstrates a hyperintense lesion occupying the posterior elements in the lumbar spine, with spinal cord impingement. *B*, T2-weighted MRI scan, axial view, illustrates invasion of the vertebral body and posterior elements by the lesion. Postoperative x-rays in anteroposterior (*C*) and lateral (*D*) views demonstrate complete lesion resection with instrumentation spanning the L1–L3 levels. The pedicles at L2 were resected in their entirety. (Images courtesy of Kern Singh, MD.)

Eosinophilic Granuloma

Eosinophilic granuloma (EG) of the bone is the most benign form of Langerhans cell histiocytosis (LCH). Unlike the other two syndromes associated with LCH, lesions in EG are confined to the bone and are frequently solitary. The ver-

tebrae are the primary location in 20% of cases.⁶ Children under the age of 15 years are most often affected, typically boys. Lesions are commonly asymptomatic, but growth often leads to fracture and symptoms of pain, swelling, and deformity.³⁰ EG of the spine commonly causes neurologic deficits that may result in spinal instability, but the most common symptom is pain.^{31,32} Tumor growth often originates in the vertebral bodies, and posterior element involvement is rare.³³ EG lesions are typically rapidly growing lesions that lead to progressive vertebral collapse with preservation of adjacent disc space, causing local edema and hemorrhage.⁶

The common radiographic presentation of EG is that of a lytic lesion featuring vertebra plana—symmetric flattening of the vertebra with preservation of intervertebral disc space and sparing of the posterior elements.³⁴ Vertebra plana lesions in a young male are considered pathognomonic for EG until proven otherwise.³⁵ These lesions usually can be identified on x-rays, although EG can occur in the absence of vertebra plana. Other pathologic processes, such as tuberculosis and Ewing's sarcoma, can also cause vertebra plana. Radionuclide bone scanning has been used in the past to detect lesions, although it has low sensitivity. A skeletal survey also may be used to investigate for additional tumors.³⁶ CT and MRI scans outline extension of the disease. Lesions are low intensity on T1-weighted MRI scans and high intensity on T2-weighted images, without any characteristic features.⁶

Treatment of EG is variable and depends on lesion size and location. Treatment methods include observation; surgical intervention (incisional biopsy, curettage, or total excision); radiation therapy; chemotherapy; or some combination of these techniques.^{37–39} Isolated bone lesions typically have

an excellent prognosis and may undergo spontaneous healing. Two major studies show that the outcome is unaffected by treatment method.^{40,41} Most series report an approximate 80% response rate, regardless of treatment method chosen.³⁸ Lesions rarely recur, and patients typically remain disease-free for the remainder of their lifetimes after treatment.⁴²

CONCLUSION

Because of the large number of patients that present to clinicians with back pain, weakness, and spinal deformity, familiarity with the basic characteristics of the primary benign osseous tumors of the spine is critical. We reviewed the basic findings of giant cell tumors, vertebral hemangiomas, aneurysmal bone cysts, and eosinophilic granulomas to assist in effective and timely management. Treatment methods are evolving, and they should be tailored toward the patient's specific clinical scenario.

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- A patient diagnosed with diabetes insipidus reports low back pain. Examination reveals that the patient also has exophthalmos. X-rays will likely demonstrate
 - osteoblastic reaction
 - lytic destruction
 - striated vertebra
 - vertebra plana
- A 12-year-old boy presents with a several-month history of progressive back pain. X-rays reveal isolated flattening of the vertebral bodies. He is also febrile, with several scalp lesions. The likely diagnosis is
 - osteosarcoma
 - non-Hodgkin's lymphoma
 - Langerhans cell histiocytosis
 - spinal osteomyelitis
- A 10-year-old child of immigrants from Mexico presents with insidious back pain. MRI demonstrates destruction of the disc and adjacent vertebral bodies. The likely diagnosis is
 - eosinophilic granuloma
 - Pott's disease (tuberculosis of the spine)
 - vertebral hemangioma
 - aneurysmal bone cyst
- In approximately what percentage of patients are vertebral hemangiomas symptomatic?
 - 1
 - 5
 - 10
 - 15
- On images, vertebral hemangioma is defined by a
 - "corduroy" appearance from diminished vertebral density along with coarse vertical trabecular striation
 - "polka-dot" appearance due to the thickened trabeculae in vertebral bodies
 - lytic appearance with a classic "eggshell" layer of surrounding cortical bone
 - A and B
- An 18-year-old woman undergoes successful surgery for a large giant-cell tumor (GCT). The sizable growth of the tumor can be attributed to all of the following, *except*
 - hormonal stimulation
 - absence of nerve impingement by the tumor
 - delay between growth and onset of symptoms
 - sacral location of the tumor
- Studies have demonstrated that regardless of treatment method chosen, the approximate cure rate for patients with eosinophilic granuloma is
 - 40%
 - 60%
 - 80%
 - 95%
- Fluid-fluid levels are sensitive and specific findings for the diagnosis of aneurysmal bone cysts with
 - MRI alone
 - CT alone
 - Plain x-rays
 - Both CT and MRI
- Origin in the posterior elements is a common finding in patients with
 - eosinophilic granuloma
 - aneurysmal bone cyst
 - vertebral hemangioma
 - GCT
- Recurrence rates of aneurysmal bone cysts are improved if
 - preoperative arterial embolization is performed
 - radiation is provided in cases of intralesional resection
 - en bloc resection is performed
 - B and C