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Osseous Spinal Tumors Benign Neoplasms: Part I

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

LEARNING OBJECTIVES: After reading this article, the participant should be 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 that disease progression is minimized. We will review the common clinical features, radiographic findings, treatment, and clinical course of

the most commonly encountered primary benign neoplasms of the osseous spine.

GENERAL CONSIDERATIONS

Primary tumors originating in the spine are much less common than metastatic lesions, which account for more than 97% of all spinal column tumors.¹ Neoplasms of the spine occur in patients of all ages but are most common in the fourth, fifth, and sixth decades of life.¹ The spine represents an unusual location for bony tumor growth, only accounting for 3.4% to 9% of all primary skeletal neoplasms.² Primary tumors are broadly classified as benign or malignant. Because of the non-specific nature of symptoms, timely diagnosis is difficult. Neurologic symptoms, usually occurring late with disease progression, are much more common in children.²

Primary tumors most commonly affect the lumbar and thoracic regions of the spine, although all regions of the spine and sacrum are affected.^{3,4} Furthermore, location of the lesion within the vertebra often holds prognostic significance: the majority (75%) of malignant tumors—both primary and metastatic—originate anteriorly in the vertebral body and involve one or more pedicles, while posterior localization is associated with malignancy in only 32% of cases.³ Imaging findings on x-rays, CT scans, and MRI scans can greatly assist in timely diagnosis, even before a diagnostic biopsy is performed.

CLINICAL PRESENTATION

Patients with primary tumors of the spinal column most commonly suffer from

back pain, although neurologic deficits and skeletal deformity are also frequent symptoms. In a series of 82 patients presenting with both benign and malignant primary neoplasms of the bony spine, the most common symptoms were pain in 84%, subjective weakness in 42%, and the presence of a mass in 16% of patients.³ Less than 10% of patients initially present with spinal deformity, which often elicits back pain through paraspinous muscle spasm.⁵

Back pain due to neoplasm typically begins intermittently but progressively worsens. Pain is worse at night and at rest, and it is not improved by nonsteroidal anti-inflammatory drugs (NSAIDs). The pain is not closely associated with activity, which distinguishes it from the more typical mechanical back pain.⁶ Typically, pressure or percussion over the involved segment can localize the pain.

Lower-extremity weakness can occur, but it rarely represents the first symptom seen. Finally, bowel and bladder dysfunction may develop before diagnosis is confirmed in up to half of patients with cord compression from tumor growth. Isolated sphincter dysfunction can develop from pressure at the level of conus medullaris, although lower-extremity impairments are more common with a tumor at this location. A thorough neurologic examination is critical to assess for deficits and evaluate for bladder and bowel function if warranted.

IMAGING

Plain films should be the first diagnostic imaging test performed if a spinal tumor is suspected. Anteroposterior (AP)

Mr. Munns is Medical Student and Research Associate, Dr. Park is Resident, and Dr. Singh is Assistant Professor, Department of Orthopaedic Surgery, Rush University Medical Center, 1725 West Harrison Street, POB 1063, Chicago, IL 60612; E-mail: kern.singh@rushortho.com.

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and lateral films often provide considerable information about the nature of the lesion and assist in identifying characteristic tumor types. Geographic patterns of bone destruction suggest a slowly expanding lesion that is often benign; a moth-eaten appearance is typical of more rapidly growing tumors; and a permeative pattern of bone destruction is emblematic of highly malignant, aggressive lesions with the most rapid rate of growth.⁷ Early in the course of growth, however, a lesion of the vertebral body might be very difficult to detect as radiographic evidence of bone destruction is not visible until 30% to 50% of trabecular bone has been destroyed.⁸ The most visible sign on AP x-rays is the typical "winking owl" sign, which represents a unilateral loss of the bony pedicle ring from invasion by a radiopaque tumor mass. Vertebral body collapse from bony erosion is also commonly seen.

A 99m technetium bone scan can also assist in identifying a neoplastic process in the bony spine. Technetium scans are sensitive to areas of increased osteoid formation. They can identify lesions as small as 2 mm, provided an osteoblastic response occurs in the surrounding bone. The high sensitivity but low specificity of the scans makes them ideal for diagnosing symptomatic patients with negative or equivocal x-rays and for determining the spread of a known systemic disease.⁹ When an isolated lesion is identi-

fied, the differential diagnosis remains broad, including fracture, infection, neoplasm, and local soft-tissue inflammation.

CT is best used to characterize cortical lesions and mineralized matrix, which are disturbed with neoplastic invasion.¹⁰ It can effectively identify early changes to cortical bone before further destruction occurs or before intramedullary invasion occurs. CT also effectively images the spinal cord, alone or in combination with a myelogram.

MRI is the method of choice to evaluate the spine given its superior sensitivity and specificity. It delineates soft tissue and spinal cord invasion more effectively than CT. MRI is able to detect tumors because the increased cellularity and extracellular water content of bone marrow with neoplastic growth provide a different signal intensity compared to surrounding tissues and nearby non-affected bone.¹¹ Gadolinium can assist in increasing contrast between tumor tissue and normal bone marrow, allowing use of MRI to visualize tumor growth earlier than other modalities.¹²

BIOPSY

Obtaining a suitable biopsy specimen remains a critical step in the staging, diagnosis, and treatment of a spinal tumor. Care must be taken to ensure that the biopsy is performed correctly, as the incision site for the biopsy often determines the surgical approach. The incidence of inadequate or inappropriate extremity biopsy that significantly alters a patient's care is greater than one in three overall, a figure that could be even higher for spinal lesions.¹³ This risk is minimized when the biopsy is performed by the treating physician. Biopsies are not always necessary as radiographic and other staging procedures frequently allow the surgeon to perform definitive surgical therapy.

When indicated, however, biopsies can be performed in three distinct forms: excisional, incisional, and needle. Excisional incision is occasionally called upon in the evaluation of a spinal lesion (e.g., if the lesion is posteriorly located, away from neural structures); otherwise, needle and incisional biopsy techniques are preferred. Needle biopsies are the least invasive technique but are the most prone to sampling errors.



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Because of this, needle biopsies are used for confirmation of disease—such as recurrence of a known lesion, metastatic spread of a known malignancy, or sarcomatous histology of a known osteosarcoma.¹⁴ In most other cases, incisional biopsy is performed immediately before definitive surgical therapy. The incision should be as direct as possible, and it should lie with-

in the field such that it 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.

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.

BENIGN TUMORS

Table 1 lists the various benign osseous spinal lesions.

Osteochondroma

Osteochondromas account for 30% to 40% of benign bone tumors, although they are seen uncommonly in the spine, representing only 4% of all solitary spinal lesions.^{15,16} Commonly, patients are male (2.5:1 predominance) and are frequently diagnosed in the third or fourth decade of life—although patients with multiple hereditary exostosis syndrome present earlier.^{17,18} Rarely, osteochondromas arise at a site of prior radiation therapy.¹⁹ Lesions are typically solitary, occur in the posterior elements, and commonly occur in the cervical spine (in 50% cases), especially C2.^{17,20} Myelopathy is the most common presenting symptom (34%) overall in patients with a solitary lesion.² When the mass arises anteriorly in the cervical spine, however, dysphagia and hoarseness may be presenting symptoms.²¹

Radiography has limited diagnostic significance, as 15% of x-rays appear normal, and a definitive diagnosis from the image can be made in only 21% of cases.² The pathologic/radiologic hallmark of osteochondromas is cortical and medullary continuity with the underlying bone. Thin-section CT, however, is the modality of choice to assess the osseous nature of the lesion and detect the marrow/cortical continuity. MRI can assist in diagnosis by revealing the contiguity of yellow marrow with high signal intensity on T1-weighted images and intermediate signal intensity on T2-weighted images centrally, and low signal intensity peripherally.²² MRI also can delineate the thickness of the cartilage cap, which may signify the possibility of malignancy. A well-known complication of osteochondroma is malignant transformation to chondrosarcoma (in 1% to 5% of patients with a solitary lesion), an event suggested by the presence of a hyaline cap larger than 3 cm in thickness, recurrence of a lesion after resection, and new-onset pain or lesion growth.²³

Treatment involves surgical resection. Successful removal of the cartilaginous cap is required to prevent recurrence.²⁴ Incomplete resection can prompt disease recurrence, but complete surgical excision is usually curative; 89% of patients experience improvement postoperatively. Surgical treatment should be tailored according to the tumor: lesions confined to the posterior elements are commonly treated with conventional laminectomy. Facetectomy without laminectomy might be required for strictly articular involvement, whereas extensive lesions might require spinal fusion with instrumentation.²⁵

Osteoid Osteoma

Osteoid osteomas are benign bone lesions less than 2 cm in diameter that arise from cancellous bone. The tumors arise most often in long bones, but they occur in the spine in approximately 10% of cases. The lumbar spine is the most frequent target, followed by cervical, thoracic, and sacral segments.²⁶ The posterior elements are involved in 75% of cases.²⁷ Patients—typically male—present between 5 and 25 years of age with symptoms of dull back pain that worsens at night and is relieved by salicylates or NSAIDs. Painful or dextroscoliosis concave to the side of the tumor is a less common

feature of osteoid osteoma. Painful deformity is a notable finding because idiopathic scoliosis, a competing diagnosis, demonstrates a thoracic levoscoliosis that is typically painless. Neurologic compromise is even less frequent.²²

Radiographically, the classic appearance of osteoid osteoma is a round to oval, discrete radiolucent center (the nidus) surrounded by a variable amount of sclerosis and occasional mixed calcifications. Central calcification may be present, although the complex anatomy of the spine often obscures the nidus.¹⁶ Technetium bone scan is recognized as the most accurate means of localizing the tumor.²⁸ CT achieves higher sensitivity than x-rays, and it clearly delineates bony involvement, although the placement of CT cuts may cause the lesion to be missed.² On MRI scan, the nidus appears as low-intermediate signal intensity on T1-weighted images and intermediate-high signal intensity on T2-weighted images. Gadolinium contrast provides focal enhancement within the nidus. Soft tissue extension is uncommon, but a parosseous reaction may occasionally be visualized.²⁹

Treatment of osteoid osteoma includes the use of salicylates or NSAIDs as initial conservative therapy, as these lesions can resolve spontaneously. Proper localization remains crucial for complete excision, which provides definitive cure and pain relief. An instrumented fusion may be indicated if the lesion is associated with scoliosis, which occurs less commonly than in osteoblastoma.²⁸ Recent evidence suggests the nidus can be treated successfully with radiofrequency ablation, laser therapy, or alcohol injections.³⁰ Surgical excision, however, remains the gold standard.³¹

Osteoblastoma

Although similar in some ways radiographically and pathologically to an osteoid osteoma, an osteoblastoma represents a distinct entity with a different clinical presentation. Osteoblastomas account for only 1% of primary bone tumors, but they are notably more prevalent in the spine and are responsible for 10% of primary spinal neoplasms.³² Osteoblastoma often presents in the second or third decade of life, with a 2.5:1 male predominance.³³ Osteoblastoma is classically defined as a vascular, osteoid, and bone-forming tumor that contains numerous benign-appearing osteoblasts, although it is sometimes histologically indistinguishable from osteoid osteoma. Osteoblastoma has a predilection for the posterior elements of the spine, with distribution varying widely by study.² Dull localized pain present for more than a year is the most common presenting symptom. Neurologic deficit occurs in up to 29% of patients, as expansile growth can cause cortical destruction and invasion of nearby nerve roots.³³

X-rays typically demonstrate an expansile lesion that is osteolytic, osteosclerotic, or mixed, with calcifications arising from the posterior elements.³⁴ Although the tumor might demonstrate stippling, a soap-bubble appearance to these lesions is rare.³⁵ On CT scans, lucent areas of geographic bone destruction are visualized along with replacement by bone osteoid.³⁶ MRI details soft tissue invasion and nonspecific signaling, with low to intermediate signal intensity on T1-weighted images and intermediate to high intensity on T2-weighted images.³⁷ Treatment of osteoblastoma is complete surgical excision via en bloc resection. Frequently, lesions are

multilobular with soft-tissue extension, requiring intralesional resection.²⁸ These intralesional resections are associated with the recurrence of aggressive osteoblastomas in up to half of cases.³¹

Osteoblastoma has been grouped with osteoid osteoma in some descriptions due to their histologic resemblance, but numerous features can differentiate them. Osteoid osteomas are generally considered to be less than 1.5 to 2.0 cm in diameter. Clinically, the pain in osteoid osteomas is relieved by NSAIDs or salicylates, but only 27% of patients with osteoblastoma experience pain relief from these medications.³³ Osteoid osteomas are nonprogressive (and sometimes undergo spontaneous regression), unlike osteoblastomas, which often expand to encroach upon surrounding structures.³⁸ Vertebral body extension occurs in 42% of osteoblastoma cases. Recurrence rate is also significantly lower in osteoid osteomas (4.5%) compared with osteoblastomas (9.8%).³⁹

Chondroblastoma

Chondroblastomas are rare benign tumors of immature cartilage that typically arise in the epiphyses of long bones, representing 1% of benign primary bone tumors. Localization to the spine is rare (1.4%) and occurs most frequently in the cervical spine, although less than 50 cases have been reported in the literature.⁴⁰ These lesions usually present in adolescence, with a 2:1 male predominance. Chondroblastomas can often arise from both anterior and posterior elements, with spinal invasion common.^{40,41} Pain and spinal stiffness represent the most common symptoms, although neurologic involvement frequently occurs by the time of diagnosis. Vertebral lesions are typically more aggressive than those located in the appendicular skeleton.⁴²

Imaging findings in the spine are nonspecific. On x-rays and CT scans, chondroblastomas appear as aggressive osteolytic lesions with areas of calcification, as well as cystic and hemorrhagic areas. The appearance might be indistinguishable on images to that of a chondrosarcoma, making definitive diagnosis challenging.⁴³ MRI can show invasion of soft tissue or vertebral canal, with low signal on T1-weighted images and intermediate signal on T2-weighted images.⁴⁰

Treatment options include curettage or local resection. Due to the aggressive nature of the lesion and potential for recurrence, resection with total vertebrectomy is common. Specific resection strategies should be tailored to radiographic and clinical findings. Local recurrence rates range from 24% to 100%, and aggressive recurrence types have been recorded.^{40,44}

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 the basic findings of osteochondromas, osteoid osteomas, osteoblastomas, and chondroblastomas as well as their appropriate clinical management. Treatment methods are rapidly evolving and should be tailored toward the specific clinical scenario presented by the patient. The range of primary benign osseous tumors is vast; therefore, a full discussion of fibrous and vascular neoplasms will be covered parts II and III of this article.

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1. The radiographic pattern most suggestive of an aggressive spinal lesion is
 - A. geographic
 - B. permeative
 - C. moth-eaten
 - D. osteoblastic
2. How much trabecular bone destruction must occur for an osseous tumor of the spine to be visible on x-rays?
 - A. 20%
 - B. 40%
 - C. 30%
 - D. 80%
3. A 10-year-old boy reports 3 months of dull back pain that worsens at night. X-rays reveal a discrete lesion with a radiolucent center, surrounded by sclerosis and calcifications. Treatment options could include
 - A. salicylates
 - B. laser therapy
 - C. radiofrequency ablation
 - D. all of the above
4. An 18-year-old woman with dextroscoliosis presents with occasional back pain. The pain may be associated with
 - A. osteoid osteoma
 - B. osteochondroma
 - C. idiopathic adolescent scoliosis
 - D. aneurysmal bone cyst
5. All of the following statements regarding differentiation of osteoid osteomas from osteoblastomas are true, *except*
 - A. Osteoid osteomas do not frequently progress, whereas osteoblastomas are often progressive.
 - B. Osteoblastomas are typically larger than osteoid osteomas.
 - C. Nonsteroidal anti-inflammatory drugs or salicylates usually relieve pain associated with osteoblastoma, but not osteoid osteoma.
 - D. The recurrence rate is higher in osteoblastomas compared with osteoid osteomas.
6. A 40-year-old man with a history of deformity of his extremities presents with myelopathy. He has a family history of EXT gene mutation. A CT scan demonstrates an expansile lesion arising in the posterior elements. The likely diagnosis is
 - A. cervical myelopathy secondary to degenerative disc disease
 - B. osteoblastoma
 - C. metastatic disease
 - D. chondroblastoma
7. A 10-year-old boy presents with nonmechanical back pain. MRI demonstrates destruction of the vertebral disc and the adjacent vertebra. The likely diagnosis is
 - A. osteosarcoma
 - B. neuroblastoma
 - C. hemangioma
 - D. infection
8. A technetium bone scan is effective in identifying neoplasm by detecting
 - A. osteolytic areas of lesions
 - B. osteoblastic areas of lesions
 - C. osteolytic response of surrounding bone
 - D. osteoblastic response of surrounding bone
9. The most common presenting symptom of a patient with osteochondroma is
 - A. local pain
 - B. radiculopathy
 - C. dysphagia
 - D. myelopathy
10. Biopsy for diagnosis of osteochondroma should be performed
 - A. with transverse incisions
 - B. with adequate hemostasis
 - C. preferably via CT-guided needle biopsy
 - D. with all of the above