Review

**Rare Solitary Primary Osseous Lesions of the Spine in Adults; Challenges in CT and MR Imaging Diagnosis With Pathological Correlation**

Zafer Orkun TOKTAS¹, Baran YILMAZ¹, Akin AKAKIN¹, Mustafa Kemal DEMİR², Ozlem YAPICIER³, Elif ONAT³, Kamran URGUN¹, Deniz KONYA¹

¹Bahcesehir University Medical School, Neurosurgery, Istanbul, Turkey ²Bahcesehir University Medical School, Radiology, Istanbul, Turkey ³Bahcesehir University Medical School, Pathology, Istanbul, Turkey

**Abstract**

This pictorial essay is a review of the computerized tomography and magnetic resonance imaging of a few solitary primary osseous lesions encountered in the adults. The lesions discussed include giant cell tumor, Langerhans cell histiocytosis, Paget's disease, plasmacytoma, fibrous dysplasia and osteoblastoma. Challenges in computerized tomography and magnetic resonance imaging diagnosis of these lesions are mentioned with clinico-radiological differential diagnosis, and include pathological correlation. Although active diagnosis and radiological familiarity of these lesions is crucial for preventing unnecessary examinations or procedures, pathological evaluation is mandatory to establish final diagnosis.

**Keywords:** Spine; primary solitary lesions, bone tumors, computerized tomography, magnetic resonance imaging

**INTRODUCTION**

A wide variety of benign and malignant primary tumors and tumor-like lesions can involve the spine and sacrum which are rare solitary lesions when compared to metastatic disease, multiple myeloma, and lymphoma. The clinical information, patient's age and location of the lesion with the characteristic imaging findings may
help in making an accurate diagnosis. In this pictorial essay, we discuss and illustrate the imaging findings and differential diagnosis of several pathologically proved solitary spinal bone lesions including giant cell tumor, Langerhans cell histiocytosis, Paget's disease, plasmacytoma, fibrous dysplasia and osteoblastoma.

**Giant cell tumor**

Giant cell tumors (GCTs) are the most challenging benign bone tumor because of their tendency for bone destruction, local recurrence, malignant transformation and occasionally metastasis to lung and/or lymph nodes. Sacrum is the most common site of involvement in the spine. Lesions typically occur in the vertebral bodies. The tumor may extend across the sacroiliac joint, disk space, paraspinal soft tissues or into the adjacent vertebrae, which is unusual for most other bone tumors. Spinal GCTs usually affect patients between 20 and 40 years old. The main presenting symptoms are pain and neurologic deficits. The imaging findings may strongly suggest the diagnosis (Fig 1). On computerized tomography (CT) the tumor appears as a well-defined lesion with soft tissue attenuation and demonstrates an absence of matrix calcifications. Expansion or collapse of the vertebral body may be seen as well. GCTs generally show low to intermediate signal intensity to intervertebral disc on T1 and heterogeneous signals on T2 weighted (W) magnetic resonance (MR) images due to areas of previous hemorrhage with hemosiderin or collagen content. However, the MR signal may be variable and cystic areas or fluid-fluid levels representing associated aneurysmal bone cyst may also be seen. The lesion shows prominent enhancement on contrast enhanced MR images. The differential diagnosis of sacral CGTs based on imaging findings may include metastatic disease, plasmacytoma, chordoma, and schwannoma\(^{(12,14)}\).

![Figure 1a-e: Giant cell tumor of the sacrum S1 and S2 levels in a 28 year-old male patient with low back pain. (a) Axial non-contrast CT image shows a sacral body lytic lesion with geographic nonsclerotic margins and without internal mineralization or matrix (star). (b) Fat saturated sagittal T2W MR image shows homogeneous hyperintense sacrum S1 mass which extends across the intervertebral space involving sacrum S2 (arrow). (c) The lesion is isointense to disc on sagittal T1-weighted MR image (arrow). (d) It shows marked enhancement on sagittal T1-weighted fat-saturated MR image following gadolinium administration (arrow). (e) Pathologic examination shows multinucleated giant cells interspersed with spindled mononuclear cells (H&E ×400).](image)
Microscopically GCT is composed of large numbers of osteoclast-like multinucleated (10-50 nuclei) giant cells and epithelioid to spindle shaped mononuclear stromal cells. Secondary changes may be present like osteoid deposits, foci of fibrosis, collections of foamy cells or aneurysmal bone cyst-like degeneration. Conventional mitotic figures are restricted to mononuclear cells (Fig 1e). Giant cell containing numerous histopathological entities may be considered in the differential diagnosis such as aneurysmal bone cyst, benign fibrous histiocytoma, non-ossifying fibroma, chondroblastoma, giant cell reparative granuloma and giant cell rich osteosarcoma

**Langerhans cell histiocytosis**

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by idiopathic accumulation of Langerhans-type histiocytes within the various tissues. It was previously known as "histiocytosis X". LCH includes three clinical variants of the same disease, which are eosinophilic granuloma (EG) of the bone, Hand-Schuller-Christian syndrome, and Letterer-Siwe disease. EG is a benign osteolytic lesion that commonly affects the skeletal system in a unifocal or multifocal form. Spinal EG is relatively common, and most of them arise in the vertebral body. Although children are most commonly affected, it can occasionally be identified in adults. Clinical symptoms of the disease are neck or back pain, limited motion, neurologic symptoms, kyphosis and fever. However, the lesions may also be silent. The classical radiographic finding is vertebra plana. It is due to a rapidly growing lytic lesion of the vertebral body leading to progressive collapse. The disc space is preserved. Vertebral collapse may be associated with soft tissue mass. There is usually a lytic lesion on CT scans and any part of the vertebra can be involved. MR imaging findings are not specific. The lesions usually demonstrate low to intermediate signals on T1W MR images and high signal on T2W MR images. There is marked enhancement on contrast-enhanced studies (Fig 2). Since EG of the spine has many appearances that depend on the site and stage of disease, it may mimic many other conditions such as metastasis, lymphoma, leukemia, infection etc. Biopsy is required to confirm the diagnosis in all patients suspected of having this disease

Microscopically the lesion shows an intense inflammatory cell infiltration with abundant eosinophils decorated by pathognomonic Langerhans cells. Langerhans cells are polygonal cells with eosinophilic cytoplasm and oval nucleus with longitudinal grooves resembling coffee-bean. Binucleated and multinucleated forms are frequently seen. The inflammatory component often composed of abundant eosinophils admixed with neutrophils, foam cells, lymphocytes, plasma cells and histiocytes (Fig 2e). CD1a immunopositivity is characteristic of Langerhans cells. Granulomatous inflammation, Hodgkin lymphoma, Erdheim-Chester disease and Rosai-Dorfman disease should be considered in the histopathological differential diagnosis

**Monostatic Paget's disease**

Paget's disease of bone or osteitis deformans is a benign mono-ostotic or polyostotic nonhormonal osteometabolic disorder with an uncertain etiology. The disease is characterized by excessive and abnormal bone remodeling which starts with an osteolytic phase, followed by mixed and osteoblastic phases, and ending in an inactive osteosclerotic phase. Age at diagnosis is usually greater than 40 years. The lumbar spine is a very common site of involvement with multiple vertebral levels. Clinical symptoms may include pain, deformity, fracture, arthritis, spinal stenosis, cranial nerve dysfunction, and sarcomatous degeneration. However,
monostatic vertebral involvement is rare and usually asymptomatic\(^{(15)}\).

There is lateral and sagittal enlargement of the vertebral body together with the involvement of a portion of the neural arch on imaging studies. The height of the vertebra usually remains unchanged. The combination of thickening and hypertrophy of the trabecular bone at the end-plates with apposition/absorption on the periosteal and endosteal surfaces leads to the "Picture frame" sign. In the reparative stage, there is mixed sclerotic and lytic pattern, and in the sclerotic stage there may be a uniform increased vertebral density known as ivory vertebra on radiographs and CT scans. MR signal intensities may be variable due to the stage of the disease. Any disorder associated with an increase in trabecular bone mass, which is evident on radiographs and on CT scans as osteosclerotic, presents on MR imaging with low signal intensity on all image sequences\(^{(2,15)}\). However, absence of cortical destruction and perivertebral soft tissue mass and presence of preserved vertebral intramedullary fat in the low signal intensity areas on MR imaging eliminates any diagnosis with malignancy (Fig 3). There may rarely be marked osteopenia of the vertebra on imaging studies giving a “ghost” appearance on radiographs.

The main differential diagnostic considerations of Paget's disease may include trauma, aseptic necrosis, metastasis, lymphoma, myeloma, osteosarcoma, hemangioma, and infection. Mere imaging findings are generally sufficient for precise diagnosis.

Microscopically the lesion is composed of woven bone and irregular broad trabecula with prominent disorganized cement lines in a mosaic pattern. Presence of numerous large osteoclasts with multiple nuclei is a sign of profound bone resorption. In chronic phase, thick trabecula and thicker bone are seen. The intertrabecular spaces are filled with fine fibrosis (Fig 3e). Histopathological differential diagnosis includes chronic osteomyelitis, fibrous dysplasia and reactive bone adjacent to carcinoma\(^{(11)}\).

**Figure 2 a-e:** Eosinophilic granuloma of the C3 vertebra left pedicle in a 41 year-old male patient with neck pain and limited motion (arrows). (a) Sagittal reformatted CT bone window image of the cervical spine shows an ill defined osteolytic lesion with cortical destruction. (b) The lesion is hyperintense on axial T2-weighted MR image. (c) The lesion is hypointense on sagittal T1-weighted MR image. (d) Coronal contrast enhanced T1-weighted MR image shows densely enhanced mass lesion abutting the left vertebral artery. (e) Pathologic examinations shows Langerhans cells characterized by vesicular nuclei with indentations in mononucleated and multinucleated forms, admixed with prominent eosinophils and neutrophils (H&E ×400).
Plasmacytoma

Solitary plasmacytoma of the spine is a rare isolated hematologic malignancy without evidence of bone marrow plasma cell infiltration elsewhere. It is most commonly seen in the thoracic spine. Although it can present for many years as an isolated disease, multiple myeloma can develop. Direct invasion of adjacent soft tissues or any other vertebra does not constitute presence of multiple myeloma. It is mainly a lytic lesion and typically involves the vertebral body sometimes together with the posterior elements. The lesion may present as an expansile lesion with a soft tissue mass. Ivory vertebra may rarely be seen. The lesion may cause a pathological fracture of the spine. MR imaging studies usually show low signals on T1W images and high signals on T2W images, which are nonspecific. The disease shows marked enhancement on contrast-enhanced imaging studies. However, a mini-brain sign of plasmacytoma has been described in few cases (Fig 4). The radiological "mini-brain" appearance of spinal plasmacytoma appears very specific to this disease and should especially be looked for on axial CT scans when evaluating primary spinal neoplasms. Although a biopsy may still be needed to confirm the diagnosis, this radiologic sign may lead specific workup for plasmacytoma\(^9,14\). Metastatic disease and giant cell tumor must take place first in the differential diagnosis.
Microscopically, the tumor is composed of a pure population of plasma cells with small, round and eccentric nucleus, abundant basophilic cytoplasm and perinuclear halo. Binucleated or multinucleated tumor cells may be identified (Fig 4e). Immunohistochemical studies reveal light chain monoclonality and plasma cell differentiation. Multiple myeloma, reactive plasmacytosis, plasmablastic lymphoma and lymphoplasmacytic lymphoma should be considered in the histopathological differential diagnosis(13).

**Monostatic fibrous dysplasia**

Fibrous dysplasia (FD) is a tumor-like lesion that leads to deformed and weakened bones with medullary replacement by fibrous tissue due to developmental defect of osteoblastic differentiation and bone maturation of unknown origin. Spinal involvement occurs mostly in the polyostotic form of FD. Monostatic spinal involvement of the disease is a very rare entity in adults. The lumbar spine is the most common site of involvement and the lesion may involve anterior or posterior elements, or both of the vertebrae. The most common presenting symptom for patients with monostotic FD of the spine is pain. Some patients may show neurologic symptoms due to cord or nerve root compression from an expanding lesion or a pathologic fracture. The imaging features of FD may be characteristic, but not specific, and depend on the underlying histopathology of the lesion. The mixture of woven bone and fibrous components that replace the medullary space creates a characteristic “ground-glass” appearance. More radiolucent lesions are composed of predominantly fibrous elements, whereas more radioopaque lesions contain a greater proportion of woven bone. Amorphous or irregular calcification is often seen in the lesion on CT scans. On MR imaging, the signal intensity of the lesion varies from low to high on T2-weighted images and typically low on T1-weighted images. The lesion usually enhances on imaging studies. Fibrous dysplasia should be considered in the differential diagnosis when a lytic expansile lesion together with a marginal sclerosis of the spine is observed on CT and MR imaging studies (Fig 5). The differential diagnosis may include osteoblastoma, haemangioma, Paget's disease and plasmacytoma(10).

Microscopically the lesion is characterized by the presence of bland fibrous stroma admixed with trabecula of woven bone with a curvilinear branching appearance that lacks osteoblastic rimming. The lesion may contain foam cells and fatty change. Areas of chondroid metaplasia may occur in fibrous dysplasia (Fig 5e). Related with these changes and strong histologic resemblance to fibrous dysplasia, a number of fibrous and fibro-osseous lesions may enter in the differential diagnosis such as osteofibrous dysplasia, cementifying fibroma, nonossifying fibroma, desmoplastic fibroma, chondrosarcoma, parosteal osteosarcoma and low-grade intraosseous fibrous dysplasia like osteosarcoma(3).

**Osteoblastoma**

An osteoblastoma is a rare benign primary neoplasm of the bone and accounts for 1% of all primary bone tumors. They occur in the spine in approximately 32% and 46% of cases. Affected patients are usually males aged 10 to 30. Main symptoms of the disease are pain, tenderness, limited motion, scoliosis and torticollis. Patients may also present with neurologic symptoms. These tumors most commonly originate in the neural arch and may extend into the vertebral body. The radiologic appearance of the neoplasm in the posterior elements of the spine usually allows a precise diagnosis. The lesion may resemble an osteoid osteoma, with the characteristic appearance of a nidus larger than 2 cm in diameter and a surrounding area of reactive bone. An expansile lytic lesion with multifocal matrix
mineralization surrounded by a peripheral sclerotic rim is the typical radiological appearance of the spinal osteoblastomas. Aggressive pattern of the tumor destructs the adjacent bone and infiltrates the soft tissues. CT scans easily demonstrate the matrix mineralization of the disease. Depending on the degree of tumor matrix mineralization, osteoblastomas show low-to-intermediate signal intensity on T1-weighted MR images and intermediate-to-high signal intensity on T2-weighted MR images. Flare phenomenon is a characteristic feature of the lesion and is defined as peritumoral edema in bone marrow edema and soft tissues reflecting inflammatory reaction. Soft-tissue invasion with cortical destruction must be differentiated from this severe inflammatory response. There is marked enhancement on contrast-enhanced MR images due to the vascular nature of the lesion (Fig 6)\(^4\).

Microscopically osteoblastoma is a demarcated tumor composed of osteoid and trabecula of woven bone rimmed by single layer of benign activated osteoblasts with pagetoid reversal lines. The intertrabecular spaces are filled with loose fibrovascular stroma. Numerous osteoclasts, hemorrhage, aneurysmal bone cyst-like changes, epithelioid osteoblasts, degenerative nuclear atypia and cartilage matrix may be seen (Fig 6e). Differential diagnosis includes osteoid osteoma, aneurysmal bone cyst, fibrous lesions and osteosarcoma\(^8\).

Figure 4a-e: Plasmacytoma of L2 vertebra in a 67-year-old female patient with cauda equina syndrome. (a) Axial CT scan shows a lytic lesion in the vertebral body with cortical destruction (thick arrow) and soft tissue components (thin arrows). There is characteristic "mini brain" appearance due to rest trabecular bone (thick arrow). The lesion shows low signals on sagittal T1-weighted MR image (b), high signals on sagittal T2-weighted MR image (c), and marked enhancement on sagittal contrast-enhanced MR image (d) (arrows). (e) Pathologic examination shows well-differentiated plasma cells have eccentric nuclei intermingled with pleomorphic plasma cells with large nuclei and prominent nucleoli (H&E \(\times 400\)).
**Figure 5a-e:** Monostatic fibrous dysplasia of the D1 vertebra in a 25-year-old female patient with dorsal back pain. (a) Axial CT scan shows a lytic expansile lesion with sclerotic margin, internal foci of calcification and characteristic ground glass appearance. (b) The lesion shows heterogeneous low signals on sagittal T1-weighted MR image. (c) The lesion shows heterogeneous high and low signals on sagittal T2-weighted MR image. (d) The lesion shows enhancement on sagittal contrast-enhanced T1-weighted MR image. (e) Pathologic examination shows trabecula of immature bone without osteoblastic rimming lie within a bland fibrous stroma (H&E ×400).

**Figure 6a-e:** Osteoblastoma of the C6 left posterior arc in a 22-year-old male patient with cervicobrachial neuralgia. (a) CT scan shows a hypodense expansile lesion with osteoid matrix and surrounding sclerosis (arrow). (b) Axial T2-weighted MR image shows a well-defined expansile lesion with low and high signal areas due to lytic areas, multifocal ossific matrix, and peripheral sclerotic rim. (c) The tumor shows heterogeneous low signals on sagittal T1-weighted MR image. (d) Axial contrast-enhanced fat-saturated T1-weighted MR image shows marked enhancement of the lesion (*) and severe inflammation of the surrounding soft tissues (arrows), indicating “flare phenomenon”. (e) Pathologic examination shows anastomosing trabeculae of woven bone rimmed by benign activated osteoblasts with dispersed osteoclasts in bland fibrovascular stroma between trabeculae (H&E, x200).
CONCLUSION

Despite their rarity, the radiologist must be familiar with the typical imaging findings of the solitary spinal bone lesions to provide most likely diagnosis for the guidance of patient management. CT examination is essential for the evaluation of lesion's margins and internal matrix. The cortex of the vertebra is also well characterized with CT. MR imaging evaluates the signal characteristics of the lesion and easily defines the involvement of marrow and soft tissue. Although the imaging appearances of primary spinal lesions can provide key diagnostic information for the definitive diagnosis in some cases such as Paget's disease and osteoblastoma, they do not usually allow precise diagnosis. However the accuracy of the diagnostic imaging can be improved by the addition of clinical data, the age of the patient and the lesion's site and matrix appearance. Pathological evaluation is necessary to establish final diagnosis.

Correspondence to:
Baran Yilmaz
E-mail: drbaranylmz2013@gmail.com

Received by: 12 July 2014
Revised by: 15 November 2014
Accepted: 17 November 2014

REFERENCES