REVIEW

Imaging appearance of primary bony tumors and pseudo-tumors of the spine

Aspects en imagerie des tumeurs osseuses primitives et des pseudotumeurs du rachis

A.A.K. Abdel Razek\textsuperscript{a,}\textsuperscript{*}, M. Castillo\textsuperscript{b}

\textsuperscript{a} Diagnostic Radiology Department, Mansoura Faculty of Medicine, 62, ElNokrxi street·Meet Hadr, Mansoura, Egypt
\textsuperscript{b} Department of Radiology, University of North Carolina, Chapel Hill, North Carolina, USA

Available online 24 September 2009

KEYWORDS
Spine;
Tumor;
Bony;
Imaging

Summary We aim to review the imaging appearance of primary bony tumors of the spine and simulating lesions. Benign bone tumors commonly appear as well-circumscribed, slow-growing lesions with a calcified or sclerotic matrix. Malignancy is often aggressive permeative lesions with bone destruction, cortical invasion and associated soft-tissue mass. CT is an excellent imaging modality for characterization of the tumor matrix, exact location, extension and osseous changes, while MR imaging is superior for evaluation of the associated soft-tissue mass, bone marrow infiltration and intraspinal extension. There is a spectrum of pseudotumors that may also involve the spine. The imaging appearance of primary spinal bone tumor in conjunction with the patient’s age, gender and lesion location allows a high percentage of correct diagnosis. Imaging plays an important role in diagnosis, characterization and extension of bone tumors of the spine which will help guide therapy.

© 2009 Elsevier Masson SAS. All rights reserved.

Introduction

Primary bone tumors of the spine are uncommon, representing less than 5% of all bone tumors. Primary benign and malignant bony tumors can involve the spine and their clinical presentation is often nonspecific. Identification of bone tumors of the spine requires a high degree of clinical suspicion and imaging evaluation, with the resultant findings often being strongly suggestive of the diagnosis. Primary bony lesions of the spine may exhibit characteristic imaging features that can help the radiologist develop a differential diagnosis. CT and MR imaging are essential in characterizing bony spinal lesions and detecting their extension and associated neural compression [1–7]. Table 1 shows the World Health Organization (WHO) classification of primary benign and malignant bone tumors of the spine [8]. Tumor-simulating lesions represent non-neoplastic lesions with the appearance of benign and malignant bony tumors [3,6]. The aim of this work is to review the CT and MR imaging appearance of primary benign and malignant bony tumors of the spine and simulating lesions.

Table 1  World Health Organization (WHO) Classification of benign and malignant bone tumors of the spine.

<table>
<thead>
<tr>
<th></th>
<th>Benign</th>
<th>Malignant</th>
</tr>
</thead>
<tbody>
<tr>
<td>Osteogenic</td>
<td>Osteoid osteoma</td>
<td>Osteosarcoma</td>
</tr>
<tr>
<td>Osteoblastoma</td>
<td></td>
<td>Chondrosarcoma</td>
</tr>
<tr>
<td>Chondrogenic</td>
<td>Osteochondroma</td>
<td>Hemangioma</td>
</tr>
<tr>
<td>Osteochondroma</td>
<td>Chondroblastoma chondromyxoid fibroma</td>
<td>Lymphangioma</td>
</tr>
<tr>
<td>Vascular</td>
<td>Hemangioma</td>
<td>Hemangioendothelioma</td>
</tr>
<tr>
<td>Hematopoietic</td>
<td></td>
<td>PlasmacytomaLymphoma</td>
</tr>
<tr>
<td>Notochordal</td>
<td></td>
<td>Chordoma</td>
</tr>
<tr>
<td>Giant cell</td>
<td>Giant cell tumor</td>
<td>Ewing’s sarcoma, PNET</td>
</tr>
</tbody>
</table>

Figure 1  Osteoid osteoma. (A) Axial CT scan shows small lucent nidus (long narrow) with central calcification is seen in the right lamina that surrounded by reactive sclerosis (short arrow) in the right lamina and right transverse process. (B) Axial T1-weighted image shows concentric ring appearance: low signal intensity center (calcified nidus), surrounding intermediate signal intensity (uncalcified nidus) (large arrow) and outer rim of signal void (reactive sclerosis) (small arrow). (C) Sagittal T2-weighted image shows an area of abnormal high signal intensity representing bone marrow edema (white arrow) in the posterior vertebral body and neural arch with low signal intensity of the nidus (black arrow).

Benign tumors

Osteoid osteoma

Spinal ostoid osteoma accounts for 10% of an ostoid osteoma and 1% of spinal tumors [7]. They occur in the second decade of life with a 2:1 male predominance. The commonest location in the spine is the lumbar region (59%), followed by the cervical (27%), dorsal (12%) and sacrum (2%) spines. It is commonly located in the posterior elements (pedicles, facets, or laminae), (75%) may be in the transverse and spinous processes (18%) or rarely in the vertebral body (7%). The tumor is nidus less than 1.5 cm in diameter and is composed of well-organized trabecular bone with vascular fibrous connective tissue stroma surrounded by reactive cortical bone. The most common symptom is painful scoliosis with radicular pain. Pain is intense at night and relieved by salicylates; scoliosis occurs on the side of the lesion [1,2,7].

Radiographs show a round or oval radiolucent nidus, with surrounding sclerosis in the neural arch; the nidus may be masked by extensive sclerosis. CT is the imaging method of choice for accurate diagnosis and localization of the nidus. The nidus appears as a well-defined area of low attenuation with central mineralization and varying degrees of perinidal sclerosis. The nidus has low-to-intermediate signal intensity on T1-weighted images and variable signal intensity on T2-weighted images. Depending on the vascularity of the tumor and the presence of calcification, this lesion shows intense contrast enhancement. It may be associated with marrow edema that involves the pedicles and laminae and adjacent posterolateral parts of the vertebral body (Fig. 1). Bone scintigraphy shows marked increased uptake by the nidus with a characteristic “double density sign”. The small, central, high degree of radionuclide uptake, which corresponds to the nidus, surrounded by a less intense zone of tracer accumulation, which corresponds to the osseous reaction [1,7,9,10].

Osteoblastoma

Spinal osteoblastoma accounts for 30–40% of all osteoblastomas that occur in the second to third decades of life with a slight male predominance [1]. The lesion is equally distributed in the cervical, dorsal and lumbar vertebrae. Osteoblastoma most frequently involves the posterior...
vertebral elements (55%) with extension into the vertebral body (42%) and is rarely confined to the vertebral body (3%). Osteoblastoma shows interconnecting trabecular bone and fibrovascular stroma larger than 2 cm. Clinically, osteoblastoma is differentiated from osteoid osteoma by the dull, localized pain which is not relieved with salicylates, associated neurologic symptoms; scoliosis is less frequent and generally on the opposite side of the tumor [1,4,7].

Radiographs commonly show an expansile lytic lesion of more than 1.5 cm in diameter with matrix mineralization and may appear sclerotic. CT commonly shows a lesion larger than 1.5 cm in diameter with a well-defined margin, central calcification of the matrix and cortical expansion with demarcation by a thin bone shell. The lesion may show extensive matrix mineralization and appear sclerotic, or rarely, as an aggressive and destructive lesion. On MR imaging, the calcified portions of the tumor give rise to low T1 and T2 signal, and the uncalcified portion displays moderately high T2 signal with characteristic inhomogeneous contrast enhancement (Fig. 2). The lesion may be associated with characteristic peritumoral edema in bone and soft tissues (flare phenomenon). The edema in the bone marrow enhances homogeneously, more than the tumor itself. The tumor may be an aggressive one, leading to osseous expansion, bone destruction and infiltration of the surrounding tissues. On bone scanning there is usually marked increased uptake of radiotracer [2,11,12].

Osteochondroma

Spinal osteochondroma represents 1–4% of all solitary osteochondromas and 9% of all multiple osteochondromas. Solitary lesions are more commonly seen in males than in females (2:1) during the second decade of life, and multiple lesions have no sex predilection [1]. Solitary lesions commonly affect the cervical spine (50%) while multiple exostoses commonly involve the thoracolumbar spine. Most spinal lesions occur near the tip of the spinous or transverse processes, but may involve the vertebral body or pedicles. They are believed to originate from a separate fragment of the epiphyseal growth plate cartilage. Multiple osteochondroma may be associated with hereditary multiple exostoses. Osteochondromas associated with radiation therapy are seen in 10–15% of previously treated patients. The lesion is composed of normal bone, with a cartilage cap from which growth occurs. Malignant transformation occurs in 1% of all solitary lesions and in 5–5% of patients with hereditary multiple exostoses. The usual symptoms are pain or swelling, or it may be completely asymptomatic [1,6,7].

Radiographs show large lesions protruding from the spinous processes where cortical and marrow continuity is evident. CT is the best diagnostic tool for demonstration of the continuity of the lesion with the marrow and cortex of the underlying bone (Fig. 3). At MR imaging, the lesion manifests a central area of high T1 signal intensity (fatty marrow) and a peripheral rim of low T and T2 signal intensity (cortical bone). A thin marginal hyperintense T2 cartilaginous cap may be present. A thick (> 1-cm) cartilaginous cap in an adult patient should raise suspicion of chondrosarcoma [1,2,13].
Chondroblastoma

Only 1.4% of all chondroblastomas originate in the spine [1]. Most patients present during the third decade of life, and it is more common in males than females (2:1). The tumor involves the vertebral body and posterior elements. Back pain is the most common symptom; however, neurological symptoms may occur when the spinal canal or foramina are invaded. The lesion is composed of sheets of chondroblasts with variable amounts of chondroid matrix with chicken wire-like calcification. The tumor commonly shows aggressive features at imaging, with bone destruction and a soft-tissue mass. CT may demonstrate a geographic lesion with sclerotic borders and areas of calcifications. Most lesions have hypointense areas on T2-weighted images. Low signal intensity on T2-weighted images represents immature chondroid matrix, calcifications, and hemosiderin [1,7,14].

Chondromyxoid fibroma

Only 5% of chondromyxoid fibromas occur in the vertebrae [15]. They occur during the third decade of life with a slight female predominance. They are commonly seen in the cervical region or in the sacrum but may occur in the dorsal or lumbar spine. They are commonly seen in the posterior element, especially the spinous processes. The histological features of the tumor include a lobulated growth pattern with mixed chondroid, myxoid and fibrous areas. The lesion causes neurological impairment varying from paraesthesia to cauda equina syndrome. Imaging shows extensive erosion of bony cortex into the surrounding soft-tissue or spinal canal which may be severe enough to suggest malignancy [2,15,16].

Hemangioma

Hemangioma is the commonest benign tumor in the vertebrae (11%) [2,7]. The lesion is usually solitary but may be multiple and is most commonly seen in the thoracic or lumbar vertebrae. Hemangiomas consist of thin-walled, blood-filled vessels and sinuses lined by endothelium interspersed among the longitudinally (vertically) oriented trabeculae of bones. Accumulation of lipid material is a common secondary phenomenon. The lesion is mostly asymptomatic and may become symptomatic as a result of vertebral body expansion, epidural extension, hemotoma formation or pathologic fracture [1,2,7].

On radiographs, vertebral hemangiomas assume the characteristic cordonuery appearance, with coarsened vertical trabeculae. CT typically demonstrates a characteristic “polka dot” pattern in which there are small foci of sclerosis, representing the enlarged vertically oriented trabeculae seen on with intervening regions of lower soft-tissue (abnormal vessels) and fat attenuation. The characteristic high signal intensity of hemangioma on both T1- and T2-weighted images is related to the amount of adipocytic and / or blood vessels and interstitial edema. The lesion shows contrast enhancement (Fig. 4). Active (symptomatic) hemangiomas show involvement of an entire vertebral body, extension into the neural arch, cortical expansion, thoracic location (T3–T9), irregular honeycomb pattern with thick, low-signal-intensity vertical struts, soft-tissue mass and avid contrast enhancement (Fig. 5). Bone scan may demonstrate no uptake, increased uptake, or decreased uptake. Selective spinal arteriography demonstrates an appearance ranging from normal vascularity to marked hypervascularity, with more aggressive lesions [1,3,5,17–19].

Lymphangioma

Primary lymphangioma involving bone is extremely rare with only a few cases reported. Pathologically, there are numerous dilated vessels within the vertebral body. Vascular channels have irregular, thin walls. CT shows coarse bony trabecular structure with the shape of the vertebral body preserved. MR imaging shows a coarse, inhomogeneously hyperintense structure in the spine on T2-weighted images and a soft-tissue mass with intense contrast enhancement. Lymphangiomatosis is a rare childhood disease characterized by abnormal lymph tissue at multiple sites. It is commonly believed to be a sporadic disease, but familial associations have been established [20].

Giant cell tumor

Only 7% of giant cell tumors occur in the spine [1]. They are commonly seen in women during the third to fifth decades of life. The commonest location is the sacrum followed in descending order by the thoracic, cervical, and lumbar regions. Spinal lesions are seen in the vertebral bodies and may extend into the posterior elements. Patients present with radicular pain. A dramatic increase in lesion size can be seen in pregnancy due to hormonal stimulation. The tumor consists of abundant osteoclastic giant cells intermixed throughout the spindle cell stroma. It may be associated with underlying aneurysmal bone cyst, osteoid osteoma, or osteoblastoma.
Imaging appearance of primary bony tumors and pseudo-tumors of the spine

Figure 5  Aggressive hemangioma. (A) Axial T1-weighted image shows multiple signal void structures with coarse vertical trabeculae (polka dots) involving the entire vertebral body with extra-osseous extension into epidural space. (B) Axial contrast T1-weighted image shows avid enhancement of the lesion with enhanced epidural soft-tissue that compresses the ventral aspect of the thecal sac.

Imaging shows a well-defined expansile lytic lesion. CT demonstrates absence of mineralization and lack of a sclerotic rim at the margins of the tumor. Sacral lesions are usually large and commonly involve both sides of the midline with extension across the sacroiliac joint. Spinal lesions affect the vertebral bodies with extension into the posterior elements and paraspinal soft-tissues. Vertebral collapse may be seen. MR imaging shows characteristic heterogeneous signal intensity on all pulse sequences (Fig. 6). Low-to-intermediate intensity regions on T2-weighted images represent dense collagen matrix or hemosiderin. Evidence of hemorrhage may also appear as high signal intensity on both T1- and T2-weighted images, or as fluid-fluid levels. Low signal curvilinear areas on the T1- and T2-weighted images may be seen within the involved vertebral body, which presumably corresponds to a multicystic lesion with a thickened trabeculae, fibrous septae, or hemosiderin deposit [4,6,21,22].

Figure 6  Giant cell tumor. Axial gradient echo image shows a large expansile lesion is seen involving both sides of the sacrum.

Malignant tumors

Osteosarcoma

Osteosarcoma of the spine accounts for 4% of all osteosarcoma and 4–14% of all primary spinal malignant tumors [1]. Peak prevalence occurs during the fourth decade of life equally in both sexes. The thoracic and lumbar segments are involved with equal frequency, followed by the sacrum and the cervical column. The tumor arises in the posterior elements (79%) with partial vertebral body involvement and is confined to the vertebral body in 21% of patients. Secondary osteosarcoma occurs after radiation therapy and Paget disease. It is a high-grade malignant osteoblastic lesion with varying amounts of osteoid production. Patients may present with pain and signs of neurologic compression.

CT commonly shows a sclerotic lesion with matrix mineralization (80%) (Fig. 7). Ivory vertebra with marked mineralization in the vertebral body is seen in the rare sclerosing osteoblastic osteosarcoma, and purely lytic pattern is seen in telangiectatic osteosarcoma. On MR imaging, the sclerotic lesion shows very low signal on both T1 and T2 sequences with enhancement of the soft-tissue component. Fluid-fluid levels have been described in association with telangiectatic osteosarcoma and it can be differentiated from aneurysmal bone cyst by its thick solid parts around cystic lesions, matrix mineralization and aggressive course [1,23,24].

Chondrosarcoma

Spinal chondrosarcoma accounts for 7–12% of all chondrosarcomas and is the second most common primary malignant tumor of the spine in adults [7]. It commonly affects males (M:F = 4:1) with peak age prevalence between 30 and 70 years. The thoracic and lumbar spines are most frequently affected. It arises in the vertebral body (15%), posterior elements (40%), or both (45%). Secondary chondrosarcoma may occur when osteochondroma (solitary or multiple) undergoes malignant transformation. Presenting
symptoms include pain, palpable mass and neurological deficits.

Chondrosarcoma appears as a large, calcified mass with bone destruction (Fig. 8). Chondroid matrix mineralization is seen as rings and arcs on CT and as areas of signal void at MR imaging. The nonmineralized portion of the tumor has low attenuation on CT, low-to-intermediate signal intensity on T1-weighted MR images, and very high signal intensity on T2-weighted images due to the high water content of hyaline cartilage. Contrast enhancement may be lobular, septal, nodular or diffuse. Involvement of adjacent vertebral levels by extension through the disk is seen in 35% of lesions, and adjacent ribs may also be affected in thoracic tumors. Chondrosarcoma arising from osteochondroma is seen as thickening at the peripheral cartilaginous cap [4,25,26].

Hemangio-endothelioma

Hemangio-endothelioma occurs from the second to eighth decade with a slight male predilection. It is prone to arise in the thoracic and lumbar spine. Most of the lesions involve the vertebral body but it rarely affects the posterior elements. The tumor is often large and aggressive. The tumor may be multifocal and metastatic lesions (20—30%) involve mainly the lungs, regional lymph nodes and liver. Radiograph shows multifocal lytic lesions (honeycomb appearance) and aggressive bony destruction with expansion. CT shows an expansile lytic lesion with large soft-tissue mass, or a sclerotic lesion with dense vertebrae (Fig. 9). MR imaging shows prominent serpentine signal void vessels with prominent heterogeneous contrast enhancement. Extra-osseous extension of the tumor through zones of cortical destruction can also be seen [27].

Plasmacytoma

Spinal involvement occurs in 25—60% of patients with plasmacytoma [29]. Seventy percent of patients are over 60 years of age. The thoracic spine is the commonest site involved, followed by lumbar spine, cervical spine and sacrum in descending order. The vertebral body is the most common site of involvement due to its rich red marrow content, but the tumor frequently extends to the pedicles. Plasmacytoma shows focal proliferation of malignant plasma cells that represent the early stages of multiple myeloma. Patients can be asymptomatic or may experience pain, nerve root irritation, and paraplegia.

Plasmacytoma usually presents as a purely lytic appearance with variable degrees of collapsed vertebra. It has low signal intensity on T1-weighted MR images, high signal intensity on T2-weighted images, and homogeneous marked contrast enhancement. The vertebral endplates may be partly destroyed but maybe also be partly sclerotic. Involvement of the intervertebral disk and adjacent vertebrae has been described. It may show a characteristic “mini brain”
Imaging appearance of primary bony tumors and pseudo-tumors of the spine

Figure 10  Plasmacytoma. (A) Sagittal T2-weighted image shows compressed D12 vertebra with high signal intensity. (B) Axial CT scan in another patient shows mini brain appearance with curvilinear thickened cortical struts and irregularity of the vertebral body cortex. The lesion extends into both pedicles.

appearance on axial images with curvilinear thickened cortical struts and irregularity of the vertebral body cortex. The thickened cortical struts are a result of a stress phenomenon from the lytic process involving the cancellous bone forcing the remainder of the bone to increase thickness (Fig. 10) [28,29].

Multiple myeloma
Multiple myeloma shows multiple punched out lesions (Fig. 11), expansile lesions with soft-tissue masses, and pathologic fractures on CT. Diffuse sclerosis of the axial skeleton is seen in 1–3% of patients and may be caused by a secondary myelofibrosis. MR imaging shows normal appearance of bone marrow (28%) when less than 20% marrow infiltration is present; focal infiltration (30%) appears as high signal on gradient echoes and T2-weighted images. Also, diffuse bone marrow involvement (39%) results in high signal intensity on T2-weighted images and low signal on T1-weighted images with moderate to high-grade disease and a variegated, or salt-and-pepper, appearance (3%) with patchy inhomogeneous bone marrow on T2-weighted images in patients with low-grade disease [1,3,30,31].

Lymphoma
Primary lymphoma of bone accounts for 1–3% of all lymphomas [1]. The spine constitutes the fourth most common site for primary lymphoma of bone where non-Hodgkin lymphoma predominates. Spinal lesions are more frequent in men (8:1) and affect patients in the fifth to seventh decades of life. Involvement of the spine can occur either due to a paraspinal tumor invading the vertebral body, or if the tumor originates in the vertebra. Spinal involvement may manifest as paraspinal, vertebral, and epidural involvement, either in isolation or combination. Spinal lymphoma is commonly manifested by pain and neurologic and systemic symptoms.

In contrast with bone metastasis, lymphoma may be well tolerated in some patients, even with epidural infiltration. It can be seen with other small round cell tumors such as Ewing sarcoma. Contiguous vertebral involvement has also been reported.

The radiographic appearance of the tumor is variable. The osteolytic pattern may be permeative, moth eaten, or rarely, geographic. Mixed osteolytic–osteosclerotic or purely osteosclerotic lesions are rare. The sclerotic (ivory vertebra) and mixed patterns are more common in Hodgkin disease. Additional findings may include pathologic fractures and soft-tissue masses. On MR imaging, lymphomatous infiltration appears as focal or diffuse areas of low signal intensity on T1-weighted images and variable signal intensity on T2-weighted images. Compressed vertebrae have been reported (Fig. 12). Diffuse enhancement of the lesion is usually seen. A focus of bone marrow replacement and a

Figure 11  Multiple myeloma. Sagittal CT scan shows multiple lytic lesions with mouth eaten appearance are seen involving the cervical vertebral bodies with lacking of C6 vertebral body.
surrounding soft-tissue mass without large areas of cortical bone destruction suggest lymphoma [1,6,32,33].

**Leukemia**

Spinal involvement is more frequently seen in children with acute lymphoblastic leukemia, with a slight male predilection. Radiographic changes include multiple radiolucent foci, or rarely, osteosclerotic areas. Compression fractures may be evident. On MR images, leukemic marrow demonstrates decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted images with diffuse contrast enhancement. Sclerotic foci can be seen as areas of low signal intensity on both T1- and T2-weighted images. It may be associated with soft-tissue extension and compromise of the spinal canal [6,34].

**Chordoma**

Chordomas are the second most common primary malignant neoplasm of the spine in adults with their peak prevalence in the fifth to sixth decades [1,6]. Men are affected twice as often as women. Chordomas most commonly arise in the sacrococcygeal region (50%), followed by the sphenoccipital region (35%) and the vertebral bodies (15%). Spinal chordomas arise more frequently in the cervical spine than in the thoracic and lumbar regions. The most common site of involvement is the vertebral body but may also involve neighboring soft-tissues. It arises from the remnants of the primitive notochord with characteristic physaliphorous cells. The tumor shows gelatinous mucoid substance, hemorrhage, necrotic areas and may contain calcifications and sequestered bone fragments. Clinical manifestations are often insidious because chordomas are slow-growing lesions.

Chordoma appears as a destructive, often expansile lesion centered in the midline associated with soft-tissue mass with a "collar button" or "mushroom" appearance and a "dumbbell" shape, spanning several segments and sparing the disks. It contains areas of amorphous calcifications in the spine (40%) and sacrococcygeal lesions (90%). Additional findings include osteosclerosis (43–62%) and vertebral collapse. On T1-weighted images, chordomas are iso- or hypo-intense with focal hyperintense regions representing hemorrhage with myxoid and mucinous collections. On T2-weighted images, most chordomas have high signal intensity due to gelatinous substance with hypo-intense regions of fibrous sepatae and hemosiderin. The tumor shows heterogeneous or ring and arc enhancement [1,3,35,36] (Fig. 13).

**Giant notochordal rest**

Giant notochordal rest is differentiated from chordoma by lack of symptoms, absent pain and no progression or growth. It appears as normal bone with a variable degree of sclerosis.
that conforms to the shape of a vertebral body, whereas MR imaging shows a lesion with low T1 and high T2 signal intensity and no soft-tissue involvement. Bone scintigraphic findings are typically normal. If the lesion is found incidentally, periodic imaging follow-up is recommended [37].

Ewing’s sarcoma

Primary Ewing’s sarcoma of the spinal column comprises 3–10% of all Ewing sarcomas [6]. Metastatic foci of Ewing’s sarcoma involving the spine are more common than primary lesions. It is usually seen in the second decade of life with a slight male predilection (62%). The sacrum is the most frequently involved site (55%), followed by the lumbar spine (25%). More than one segment is involved in 8% of patients. The lesion commonly originates in the posterior elements with extension into the vertebral body. Ewing sarcoma is an undifferentiated high-grade proliferation of uniform small round cells.

The tumor is commonly lytic, aggressive, and may produce a vertebra plana (90%) and may have a pseudo-osteomangiomatous appearance. A purely sclerotic pattern (ivory vertebra) is rare and might correspond to necrotic or reactive bone formation. Invasion of the spinal canal is common (91%) and the paraspinous component is often larger than the intrasosseous lesion. Ewing’s sarcoma has low-to-intermediate signal on T1-weighted images and high signal intensity on T2-weighted images. The intramedullary portion of the tumor may be homogeneous or heterogeneous, depending on its histologic presentation. The extra-osseous component is often homogeneous in signal intensity because it is often devoid of calcification and new bone formation [6,38,39].

Extraskeletal Ewing sarcoma

Extraskeletal Ewing’s sarcoma is commonly seen in paravertebral and extradural locations. Rarely intradural and intramedullary instances have been reported. These tumors are highly malignant with an aggressive course, with a high recurrence rate and rapid progression with metastases most commonly to lung, bone, and bone marrow [40] (Fig. 14).

Primary neuroectodermal tumor (PNET)

Primary spinal osseous PNET is exceedingly rare. PNET is a malignant small round cell tumors exhibiting neuroectodermal differentiation. It is differentiated from Ewing sarcoma by immunohistochemical studies. The imaging appearance of vertebral PNET and ES are similar but PNET is more aggressive [1,6].

Pseudo-tumors

Aneurysmal bone cyst

Spinal aneurysmal bone cyst represents 3–20% of all aneurysmal bone cysts [1]. It usually occurs between the ages of 5–20 years with a slight female predilection. The cervical spine is affected in 22%, the thoracic spine in 34%, the lumbar spine in 31% and the sacrum in 13% of patients. It commonly involves the posterior elements (60%) and the vertebral bodies (40%). The three main hypotheses propose that the lesion is the result of either the improper repair of a traumatic subperiosteal hemorrhage, a vascular disturbance of bone, or hemorrhage into a preexisting lesion. ABC may arise within preexisting lesions such as chondrosarcoma and giant cell tumor. It is typically characterized by blood-filled cystic spaces separated by a spindle cell stroma with osteoclast-like giant cells. Patients usually present with local pain and radicular symptoms.

Radiographs show a soap bubble appearance with eggshell-like calcification. CT shows a well-defined expansile lesion with internal septations that may extend into the adjacent vertebrae and the paravertebral soft-tissues. MR imaging reveals a multilocular expansile cyst with variable signal intensities. Fluid-fluid levels within ABCs are indicative of hemorrhage with sedimentation. On T1 and T2-weighted images, they may have increased signal intensity due to methemoglobin in either the dependent or nondependent component. The lesion margins show a low-signal-intensity rim on MR images that is thought to be caused by an intact, thickened periosteal membrane. Contrast studies show smooth enhancement of the internal septae within ABCs (Fig. 15). The presence of a solid component with diffuse contrast enhancement should raise suspicion for secondary ABC, although it may be encountered in the solid variant of ABC [1,4,6,41,42].

Bone island (Enostosis)

A bone island is a common spinal lesion (14% in cadavers) that is always discovered incidentally [1]. It is due to localized failure of bone resorption during skeletal maturation resulting in a mass of lamellar compact bone within cancellous bone (hamartoma). On CT, it appears as a circumferential osteoblastic lesion with an abrupt transition from normal to sclerotic bone and spicular border (Fig. 16). MRI demonstrates low signal intensity in all sequences. Bone scan findings are usually normal but lack of activity on bone scan is not always the rule, especially with big size lesions.
Enostosis is differentiated from osteoblastic metastases by normal adjacent bone, sharp margins, spicular border and no primary tumor. Most of them remain stable, but some may slowly increase in size. If an enostosis exhibits an increase in diameter greater than 25% in six months, a biopsy should be performed [1,2,43].

Schmorl’s node

A Schmorl’s node is most commonly found in the lower thoracic and upper lumbar spine. It usually involves the inferior endplate and is more common in men. It represents a vertical disk prolapse through an area of weakness in the vertebral endplate. The etiologies of Schmorl’s node can be idiopathic, degenerative, traumatic, malignant, infectious, or metabolic disease. Imaging shows irregularities of the vertebral contours or small radiolucent lesions of the vertebral bodies limited by reactive sclerosis and connected with the intervertebral disks. However, Schmorl’s nodes may appear as large cystic lesions (Fig. 17). Tunneling variants, with edematous nodes or nodes with surrounding marrow edema, have also been reported and may be acute and symptomatic [44,45].

Tuberculous spondylitis

Tuberculous spondylitis comprises 25–60% of bone and joint tuberculous infections. It commonly affects adults in their
Figure 18  Spinal tuberculosis. Axial contrast T1-weighted image shows an enhancing lesion of the disc associated with enhancing epidural mass and large paravertebral soft-tissue abscesses.

fourth and fifth decades of life. Lower thoracic and lumbar vertebrae are the most common sites, whereas the sacrum and cervical region are less common sites. More than one vertebral involvement is usual. Imaging appearances suggestive of tuberculous infection are enhancing intra-osseous abscesses, subligamentous spread, large paravertebral soft-tissue abscesses, and soft-tissue calcifications. The discs may be spared until later in the course of the disease (Fig. 18). Compression fracture, gibbus deformity and scoliosis also occur [46,47].

Hydatid disease

Hydatid cysts of the spine occur in 1% of cases of hydatidosis and 50% of skeletal hydatid cysts. It is common in males in their third to sixth decades. Spinal hydatid cysts are usually situated in the thoracic region. Signs of nerve compression including paraplegia are frequent. The imaging appearance is that of a multilocular cyst without reactive sclerosis, or an expansile lesion with a "blown-out" appearance. Hydatid cysts cause multiseptated lesions with minimal enhancement. While intra-osseous cysts show no calcification, extra-osseous cysts may calcify [1,46,47].

Paget disease

The spine is the second most commonly affected site (53%) in patients with Paget disease [48]. Vertebral involvement is seen in 35–50% of patients with polyostotic disease (66%). The lumbar spine is the most frequently involved site (58%), followed by thoracic (45%) and the cervical vertebrae. The vertebral body is almost always involved, as well as, the posterior elements. The disease is characterized by a disturbance in bone modeling and remodeling. Sarcomatous transformation is rare (<1%).

In the mixed phase, a combination of trabecular bone hypertrophy and thickening of the endplates with apposition / absorption on the periosteal / endosteal surfaces at the anterior and posterior vertebral borders leads to the "picture frame" sign. The trabecular hypertrophy and cortical thickening results in a variable degree of low signal on both T1- and T2-weighted images. Progression of the sclerotic phase in the spine leads to the "ivory vertebra" due to increased density of the vertebral body (Fig. 19) that shows low signal intensity on T1- and T2-weighted images. Rarely, a vertebral is lytic; there is marked osteopenia of the vertebra on radiographs, giving a "ghost" appearance. There may also be fatty transformation with high signal on both T1- and T2-weighted images. In the presence of osteolysis in a Pagetic vertebra, fat signal within the lesion can be seen. [48,49].

Fibrous dysplasia

Fibrous dysplasia is a benign fibro-osseous lesion characterized by irregularly shaped trabeculae of woven bone and a fibrous component. It occurs in children and adults of both genders. Vertebral involvement is rare but occurs more frequently with polyostotic (25%) than monostotic (75%) disease. The lesion is frequently asymptomatic, but may be associated with pain and fractures. CT shows a mildly expansile lesion with a "blown-out" cortical shell, a lytic lesion with a sclerotic rim or the characteristic ground glass matrix. The latter exhibits intermediate-to-low signal intensity on T1-weighted images and variable signal intensity on T2-weighted images. It may be associated with pathological fracture or malignant transformation [50,51].

Langerhans cell histiocytosis

Vertebral involvement is seen in 8–25% of patients with Langerhans cell histiocytosis, with a peak prevalence between 5–10 years of age [52]. Patients usually have pain, which subsides rapidly after bed rest. Pathology reveals Langerhans cells variably admixed with inflammatory and plasma cells. The imaging appearance consists of complete
or incomplete collapse of a vertebral body, preservation of pedicles, posterior elements and adjacent disk spaces; absence of adjacent paravertebral soft-tissue shadow and increased opacity in the collapsed body [6,52,53].

Brown tumor

Brown tumors are found in 1.5—13% of patients with renal failure and rarely involve the spine. They commonly occur in females in the third decade of life. Brown tumor is caused by increased osteoclastic activity and fibroblastic proliferation in patients with hyperparathyroidism. On radiographs, a brown tumor is seen as an area of osteolysis with jagged sharp outlines and no sclerotic rim. CT shows an osteolytic tumor of uniform tissue density replacing the cancellous bone of the vertebral body and neural arch with a spared cortex. MR imaging findings include a hypo-intense mass causing expansion of the involved vertebra in both T1- and T2-weighted scans [54,55].

Amyloidosis

Amyloidosis is rarely found in the spine. It may be primary, familial or associated with chronic hemodialysis, multiple myeloma, and chronic infectious disease such as tuberculosis. There is a destructive spondyloarthropathy with well-defined discovevertebral erosions and large osteophytes (Fig. 20). Punctate calcifications have also been reported. MR signal characteristics include low-to-intermediate signal on T1-weighted images and intermediate-to-high signal on T2-weighted images. Enhancement of an amyloid lesion varies from none to marked [56,57].

SAPHO syndrome

Synovitis, acne, pustulosis, hyperostosis, osteitis (SAPHO) syndrome occurs in young and middle age adults in association with rheumatologic and cutaneous lesions (seronegative spondylarthropathy in the axial skeleton, 30%). Radiographs show sclerosis in combination with a variable amount of osteolysis and periostitis. Hyperostosis is seen in long-standing disease. CT shows vertebral body osteosclerosis, hyperostosis (osteosclerosis with increased bone size), paravertebral ossification, lesions at the discovevertebral junction, or vertebral collapse. At MR imaging, vertebral corner erosion is a diagnostic sign that may be associated with involvement of the endplate, and the anterior cortex of the vertebral body, adjacent disk space, adjacent vertebrae, and prevertebral tissues (thickening) [58,59].

Vertebral osteonecrosis

Vertebral osteonecrosis is an uncommon disease that occurs mostly in patients with a collapsed vertebral body after major trauma or repeated microtrabecular fractures in patients with osteoporosis or long-term ingestion of glucocorticoids. Imaging shows intravertebral air alone (40%), intravertebral fluid alone (40%) or both (20%). Intravertebral vacuum cleft sign is usually seen as an irregular lesion in the central area or adjacent to an endplate of a collapsed vertebral body that exhibits low signal intensity in all sequences. Fluid appears as a well-circumscribed area of low signal intensity on T1-weighted MR images, with high signal intensity on T2-weighted images in the vertebral body [60].

Summary and conclusion

Benign bone tumors appear as well-circumscribed lesions, often with specific tumor matrixes. Malignancy displays aggressive characteristics such as cortical erosion, bone destruction, and associated soft-tissue masses. Imaging plays an important role in diagnosis, characterization and extent of primary bone tumors of the spine as well as differentiating them from simulating lesions. CT is an excellent imaging modality in characterization of the tumor matrix, origin, extension and osseous changes, while MR is superior for evaluation of associated soft-tissue mass, bone marrow infiltration and intraspinal extension of the tumor. We concluded that the imaging appearance of bone tumor in the spine in conjunction with age, gender and location allow a high percentage of accurate diagnosis; however, biopsy is essential to reach the final diagnosis in many instances.

References

Imaging appearance of primary bony tumors and pseudo-tumors of the spine


