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Primary bone tumours from the vertebral column

Introduction

Spinal tumours have been an issue in medicine for many years. Victor Horsley conducted the first successful operation to remove a spinal tumour in the 1890s (Sachs, 1955). Spinal tumours of primary origin are less common than those with secondary causes such as metastasis. However, because of the aggressiveness of these tumours, early detection and treatment is imperative. This article summarizes the spectrum of lesions, investigation and management of these conditions.

Spinal tumours can be classified from their origin: bone marrow, bone, cartilage, muscle, and other soft tissues. Some spinal tumours may have more than one origin, such as Ewing's sarcoma. Primary bony tumours from the vertebral column can be benign or malignant. Further subcategories can be made according to the aggressiveness of the tumour. The tumours mentioned are not necessarily restricted to the spine, and indeed can be more common in other regions of the skeletal system. Some tumours are more common in one particular area of the spine, and these will be alluded to if clinically relevant.

Spinal tumour diagnosis and management

Although there are great differences between the various types of spinal tumours, there is much overlap as well. Owing to the area of involvement the more aggressive spinal tumours usually present with pain, swelling and neurological symptoms caused by compression of the spinal root or cord. The pain is generally worst at night. Many of the primary bony tumours from the vertebral column are rare, and their causes unknown. This article will present each spinal tumour, and emphasize the key differences between them.

The investigation of all spinal tumours involves urine, blood tests, X-ray, computed tomography, magnetic resonance imaging and bone scans. X-ray is always the first port of call in terms of radiological investigations; it is simple and can highlight many of the various spinal tumours. Computed tomography gives a detailed view of bony involvement of the spinal tumour, specifically mineralization, sclerosis, expansile bone remodelling, and the thin membrane region surrounding some lesions. Magnetic resonance imaging is useful in that it can evaluate the extent of the spinal tumour into the soft tissue, specifically the spinal cord.

Benign primary spinal tumours

Osteoid osteoma

Osteoid osteomas (*Table 1*) of the spine are rare primary bone tumours consisting of osteoblasts that produce osteoid and woven bone (Kan and Schmidt, 2008). They make up 1% of bone tumours, and occur within the first three decades of life, with a male predominance (2:1). Osteoid osteoma is normally less than 2 cm in diameter, usually involving the posterior elements of the spine. It usually presents with pain (worst at night) and neuralgia.

Radiographs reveal an expansile lesion radiolucent nidus surrounded by fusiform cortical thickening. Bone scan shows an increased uptake at the site of the osteoid osteoma, with computed tomography (*Figure 1*) demonstrating the extent of the lesion (Shukla et al, 2009). Pain of the lesion can be controlled by salicylates. If this fails, further options include radiofrequency ablation and local resection.

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Osteoblastoma

Osteoblastoma is an uncommon osteoid tissue-forming primary neoplasm of the bone. Although similar to an osteoid osteoma (*Figures 2a* and *b*), it is larger, being between 2–6 cm. It is less painful than osteoid osteoma, usually not worst at night, and not relieved by salicylates. Lesions can be associated with scoliosis. Investigation and treatment of the lesion is similar to that of osteoid osteoma, but some osteoblastomas can be more aggressive and have a greater tendency for recurrence.

Enostosis

An enostosis or bone island represents a focus of mature compact bone within cancellous bone, from an increase of osteoblastic activity with an unknown cause (Greenspan, 1995). The lesion is present in 14% of cadavers (Resnick et al, 1983). It is usually benign, but on occasion it can expand causing pain. Plain radiographs reveal a homogenously dense, sclerotic focus in the cancellous bone with distinctive bone streaks, creating a feather-like border (*Figure 3*). Lack of activity can be seen in the area on the bone scan (Greenspan and Stadalnik, 1995). The treatment is normally conservative, but if on rescanning 6 months later the lesion has grown more than 25%, a biopsy should be organized.

Osteochondroma

Also known as exostosis, this is a benign tumour that consists of cartilage and bone. It is a benign capped-cartilage outgrowth, connected to bone by a stalk. It is caused by a deregulation of the hedgehog signal-

Table 1. Primary bony tumours of the vertebral column

Name	Definition	Clinical presentation	Investigation	Treatment
Enostosis	Bone island that represents a focus of mature compact bone within cancellous bone, caused by increased osteoblastic activity	Usually benign, but on occasion can expand causing pain	Plain radiographs reveal a homogenously dense, sclerotic focus in the cancellous bone with distinctive bone streaks, creating a feather-like border	Biopsy if serial X-rays show expansion otherwise manage conservatively
Osteoid osteoma	A tumours consisting of osteoblasts that produce osteoid and woven bone	Presents usually in males, in the first three decades of life. Patients complain of pain (worse at night) with neuralgia	Radiographs reveal an expansile lesion radiolucent nidus surrounded by fusiform cortical thickening	Wide local resection
Aneurysmal bone cyst	Locally destructive lesion of the bone characterized by the presence of spongy or multi-loculated cystic tissue filled with blood	Presents in patients in the first two decades of life. It may cause pain, swelling and deformity. On palpation it can be warm, and possibly have a bruit	X-rays show an expansile cystic lesion having possibly a thin periosteal rim of bone with bony remodelling	Intralesional curettage
Osteochondroma	Benign capped-cartilage outgrowth, connected to bone by a stalk	Painless bump, but can appear with neurological deficit and myelopathy	X-ray shows ossification in the peritendinous tissues or an outgrowth of cartilage on a bony stalk	Removed surgically if causing irritation
Giant cell tumour	Characterized by the presence of multinucleated giant cells. Although benign it has a tendency for significant bone destruction, local recurrence, and occasionally metastasis	It usually presents with slow progressive pain, with or without a mass	X-rays demonstrate an expansile lytic lesion that has a radiolucent centre with increasing density towards the cortex. It is usually present in the epiphyseal region Classical 'soap bubble' appearance	Resection, with other adjuvant treatment including radiotherapy

Figure 1. Osteoid osteoma. Axial computed tomography study showing an osteoid osteoma nidus (arrow) in the right lamina of T7 with reactive sclerosis of the pedicle (arrowhead).

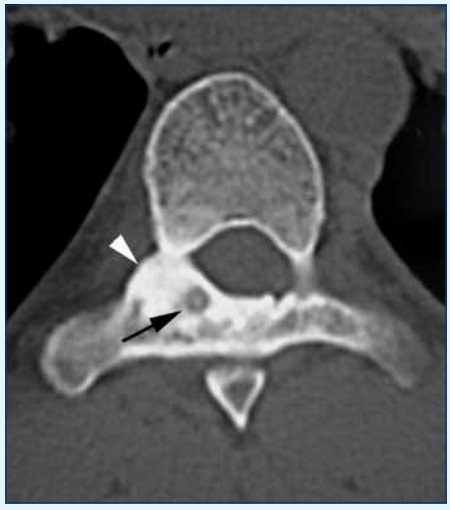
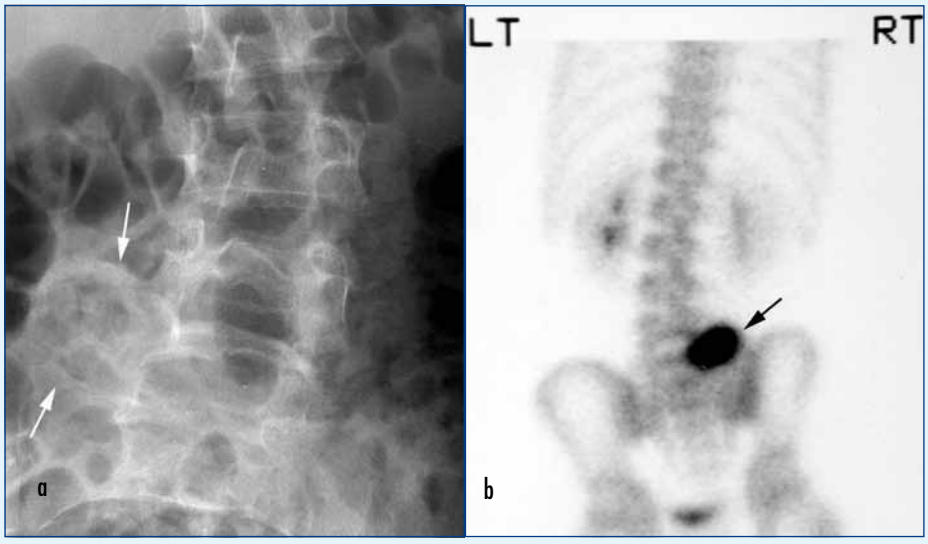


Figure 2. a. Osteoblastoma. a. Anteroposterior radiograph of the lumbar spine showing marked expansion of the right L5 transverse process (arrows). b. Posterior view bone scan showing intense increased tracer activity at the site of the tumour (arrow), consistent with its osteoblastic nature.



ing pathway that is involved in normal bone development (Bovee et al, 2010). This results in the peripheral portion of the physis herniating from the growth plate, causing an abnormal extension of metaplastic cartilage which responds to growth factors that stimulates the tumour, and therefore increases its size (D'Ambrosia and Ferguson, 1968). It is more common in the second decade of life and affects men more than women by a factor of 3 to 1. If the tumour is large enough, it will appear as ossification in the peritendinous tissues (*Figure 4*), although 15% of X-rays are normal. Magnetic resonance imaging will demonstrate a cartilage capped outgrowth that is continuous with bone marrow and the cortex of the bone. Unless causing any irritation to surrounding soft tissue, the tumour is monitored by periodic X-rays. It can be removed surgically.

Giant cell tumour

Giant cell tumour is characterized histologically by the presence of multinucleated giant cells. Although considered benign it has a tendency for significant bone destruction, local recurrence and occasionally metastasis (Simon and Springfield, 1998). The tumour comprises 5% of all primary bone tumours, and has a female predominance. It usually presents with slow progressive pain, with or without a mass. X-rays demonstrate an expansile lytic lesion usually in the neural arch or vertebral body that has a radiolucent centre with increasing density towards the cortex (*Figure 5*). This may appear as the classical 'soap bubble' appearance. Computed tomography of the chest should be performed to check for metastasis. Magnetic resonance imaging can be used to assess intramedullary and soft tissue (Murphey et al, 2001). Treatment is resection, with other adjuvant treatment including radiotherapy.

Aneurysmal bone cyst

Aneurysmal bone cyst is a benign but locally destructive lesion of the bone characterized by the presence of spongy or multi-loculated cystic tissue filled with blood (Singh et al, 2010). It appears to be caused primarily by microtrauma to vasculature, or secondarily by another neoplasm such as a giant cell tumour. It usually presents within the first two decades of life (Vergel De Dios et al, 1992), and makes up approximately 1% of bone tumours. An aneurysmal bone cyst may cause pain, swelling and deformity. X-rays show an expansile cystic lesion possibly having a thin periosteal rim of bone with bony remodelling (*Figure 6*). Magnetic resonance imaging will show a lesion with a multiple fluid interface. The normal treatment is intralesional curettage.

Malignant primary spinal tumour

Multiple myeloma

Multiple myeloma (*Table 2*) is a neoplastic proliferation of plasma cells involving the bone marrow (Raab et al, 2009). It can evolve from a solitary plasmacytoma, which differs from myeloma as it is in one location, and therefore more amenable to surgery (*Figure 7*). The plasma cells cause extensive skeletal damage, hypercalcaemia and anaemia. The plasma cells produce monoclonal antibodies which can damage

Figure 3. Enostosis. Anteroposterior radiograph of the lumbar spine showing a densely sclerotic bone island (arrows) in the inferior aspect of the L2 vertebral body.

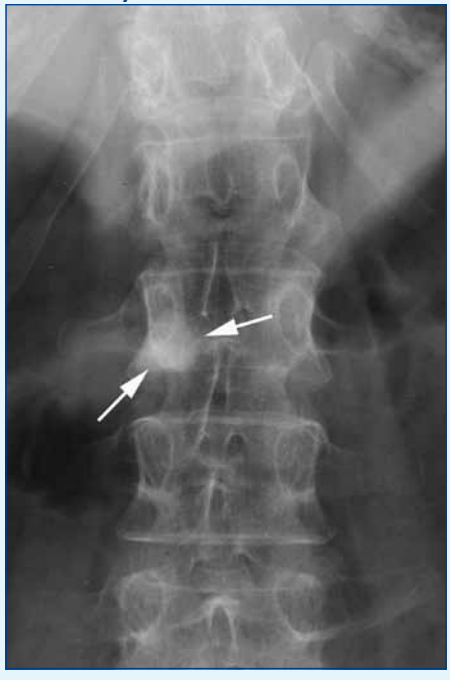


Figure 4. Osteochondroma. Oblique radiograph of the upper cervical spine showing a heavily mineralized mass (arrow) lying adjacent to the right C2 neural arch.

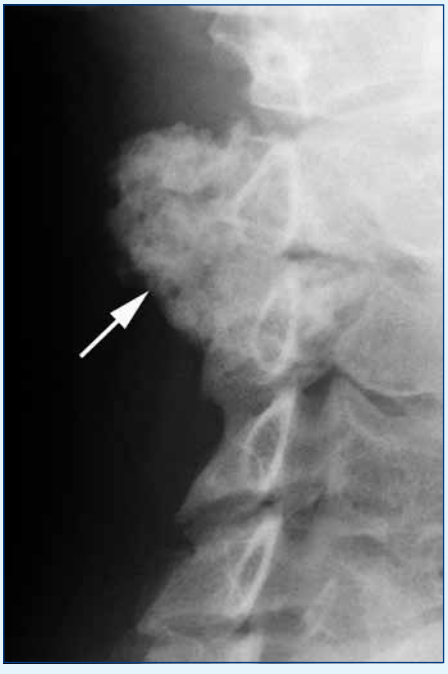
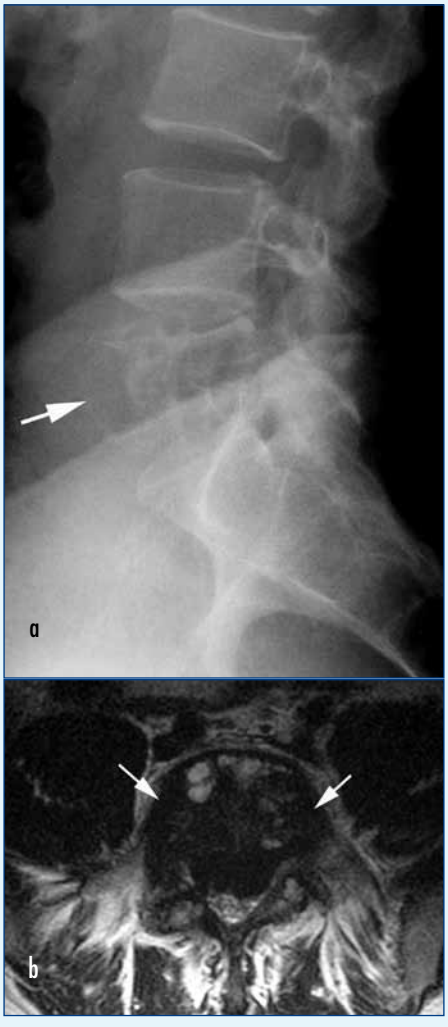


Figure 5. Giant cell tumour. a. Lateral radiograph of the lumbar spine showing lytic destruction of the L5 vertebra (arrow). b. Axial T2W magnetic resonance image showing markedly reduced signal intensity within the vertebral body (arrows), which is typical of giant cell tumour.



organs including the kidney. As the plasma cells are defective, the patient is more susceptible to infections.

The disease affects 4 in 100 000 people; it is more common in men and Afro-Caribbeans. Presentation may include back pain with neurological symptoms caused by spinal cord compression. Tiredness as a result of anaemia and recurrent infections

Table 2. Primary malignant bony tumours of the vertebral column

Name	Definition	Clinical presentation	Investigation	Treatment
Chondrosarcoma	A malignant mesenchymal tumour that produces cartilage matrix	Usually a male patient presenting with pain, a palpable mass, and possible neurological involvement. Mean survival is 5.9 years	Radiographs may show bone destruction and the chondroid matrix in the form of rings and arcs. Bone scan shows an intense uptake with a heterogenous appearance	Surgical resection
Ewing sarcoma	A malignant round-cell tumour that may originate from the bone or soft tissue	Usually pain in affected bone, but can present with mass, lethargy and fever	X-rays reveal bone lysis, sclerosis, osseous expansion, osteonecrosis	Radiation and chemotherapy
Osteosarcoma	Arises from primitive transformed cells of mesenchymal origin that exhibit osteoblastic differentiation and produce malignant osteoid	Presents in males in their 40s as pain and palpable mass	Densely mineralized matrix I, loss of vertebral height, but with sparing of adjacent disc	Patients with spinal involvement of osteosarcoma are usually dead within a year. Surgical resection with chemo- and radiotherapy
Chordoma	Tumour that arises from a notochord remnant	Subtle with gradual onset of pain, numbness, motor weakness, and constipation or incontinence	X-ray will reveal a destructive lesion with an associated tissue mass in the vertebral midline	Aggressive complete excision with wide margins if possible, give follow-up proton beam irradiation if available. If wide excision not possible, local recurrence is likely within a few years
Multiple myeloma	Malignant plasma cells, which produces abnormal quantities of immunoglobulins	Most common in middle-aged patients. Pain worst at night, anaemia, thrombocytopenia, erythrocyte sedimentation rate, increased immunoglobulins	Local areas of bone destruction. Computed tomography and magnetic resonance imaging to assess. Serum electrophoresis to check	Radio and chemotherapy. Surgery for stabilization
Solitary plasmacytoma	Descendent of plasma cell malignancies	Younger patients, can evolve to multiple myeloma		Surgical resection and stabilization 60% after 5 years

Figure 6. a. Aneurysmal bone cyst. Sagittal T2W magnetic resonance image showing the lesion in L3 containing multiple fluid levels, a hallmark of aneurysmal bone cyst. b. Anteroposterior radiograph of the lumbar spine showing lytic destruction of the left side of the L3 vertebral body.

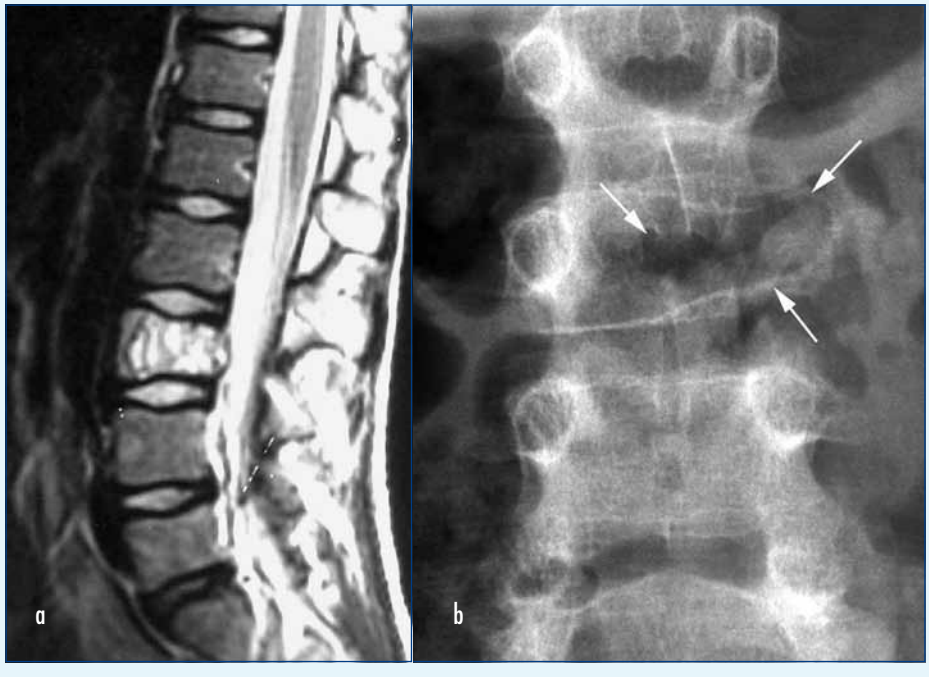
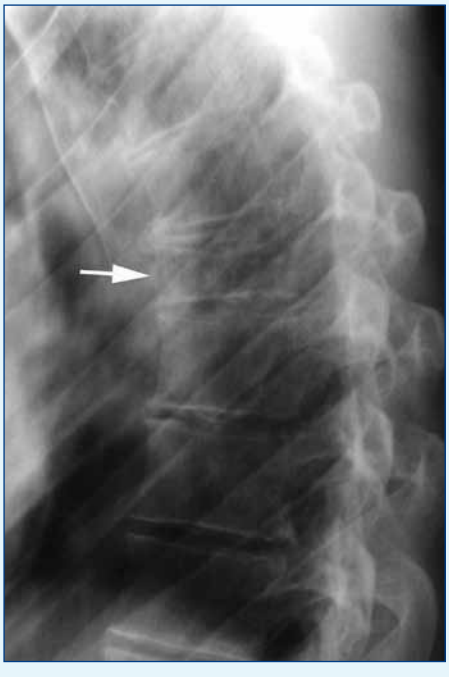


Figure 7. Solitary plasmacytoma. Lateral radiograph of the thoracic spine showing pathological collapse of a mid-thoracic vertebra (arrow).

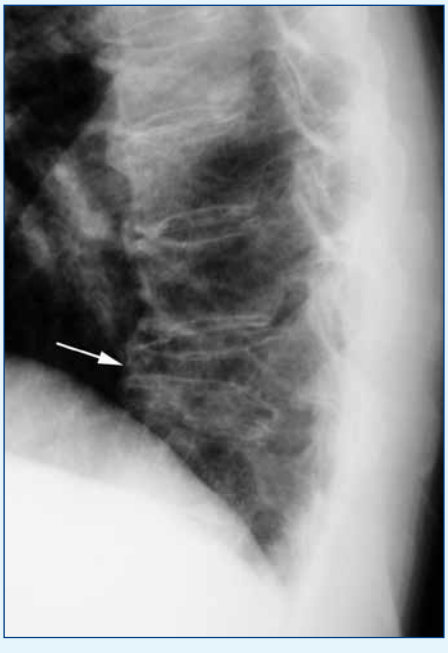


are some of the other symptoms. Blood tests include urine dipstick, renal function, full blood count, erythrocyte sedimentation rate, and protein electrophoresis (looking for Bence-Jones protein). X-rays may show lytic lesions (*Figure 8*). Magnetic resonance imaging will be more sensitive in identifying these lesions. At present multiple myeloma is incurable. Chemo- and radiotherapy are the predominant treatment to slow the progress of the condition, and surgery is only involved for stabilization of the bones. The prognosis of myeloma is poor with a survival rate of 20% at 5 years.

Osteosarcoma

Osteosarcoma is a cancer arising from primitive transformed cells of mesenchymal origin that exhibit osteoblastic differentiation and produce malignant osteoid (Buecker et al, 2005). It is the most common form of primary bone cancer, but is still rare in the spine. It occurs largely in male teenagers and in the 6th decade of life. Children with previous rare cancers such as retinoblastoma are at more risk of it (Ottaviani and Jaffe, 2010). The tumour is normally found in the vertebral body of the spine. X-rays may demonstrate an osteolytic or osteoblastic lesion (*Figure 9*).

Figure 8. Multiple myeloma. Lateral radiograph of the thoracic spine showing pathological collapse of a lower thoracic vertebra (arrow) with generalized osteopenia of the remaining vertebral bodies.



Extension of the tumour into the periosteum will give a sunburst appearance (Wenaden et al, 2005). A magnetic resonance imaging scan will show the extent of the tumour and where best to biopsy. Historically, the outcome of osteosarcoma of the spine is poor. However, a multidisciplinary approach, neo-adjuvant treatment and aggressive surgical treatment gives the best outcome.

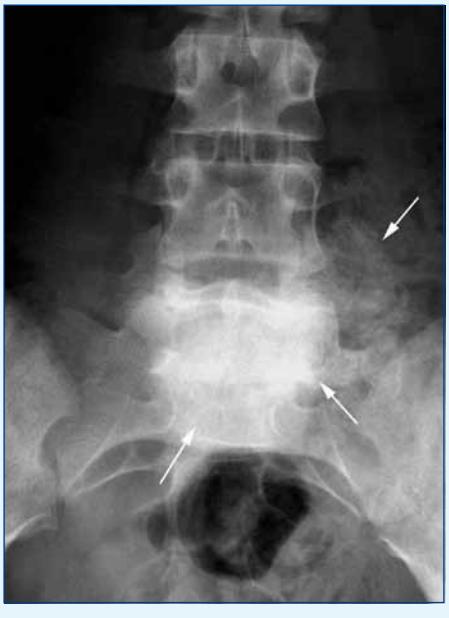
Ewing's sarcoma

This is a malignant round-cell tumour that may originate from the bone or soft tissue. It is the most common non-lymphoproliferative primary malignant tumour of the spine in children. It is caused by a translocation between chromosome 11 and 22. X-rays reveal bone lysis, sclerosis, osseous expansion and osteonecrosis (*Figure 10*). Because most patients present with metastatic disease, multidrug chemotherapy is used as well as local disease control with surgery or radiation.

Chondrosarcoma

Chondrosarcoma is a malignant mesenchymal tumour that produces cartilage matrix (Gelderblom et al, 2008). It predominantly affects men, with a mean age of 45 years, and comprises 7–12% of all spinal tumours (Shives et al, 1989). The clinical manifestation depends on the grade of the tumour. A high grade tumour presents with excruciating pain. Often the first symptom may be a broken bone as a result of mild trauma (Souhami et al, 2001). A low grade tumour presents with aching, pain and swelling.

Figure 9. Osteosarcoma. Anteroposterior radiograph of the lumbar spine showing a poorly defined densely osteoblastic tumour mass (arrows) arising from L5.



On X-ray a fusiform, lucent defect with scalloping of the inner cortex and periosteal reaction, with possible extension into soft tissue (*Figure 11*). Computed tomography will help define the integrity of the cortex and calcification. Magnetic resonance imaging demonstrates detailed soft tissue involvement of the tumour. Treatment is wide surgical resection with a limited role for chemo- and radiotherapy. Prognosis is dependent on the grade, size and stage of the tumour.

Figure 10. Ewing sarcoma. a. Axial computed tomography study showing a large soft tissue mass (arrows) anterior to the sacrum. b. Sagittal T1W magnetic resonance image showing marked tumour infiltration of S1 and S2 (arrows) with a large mass in the spinal canal (arrowhead).

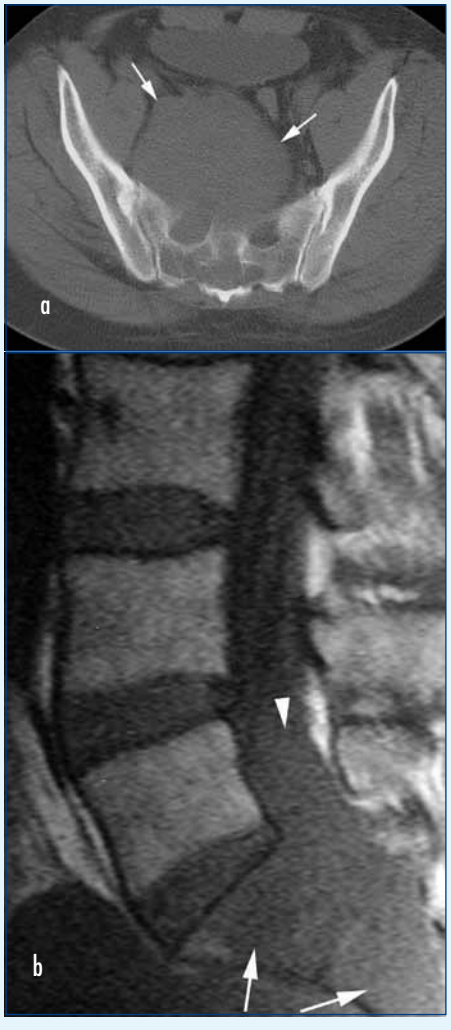


Figure 11. Chondrosarcoma. Anteroposterior radiograph of the lumbar spine showing marked expansion of the right L4 transverse process (arrows).

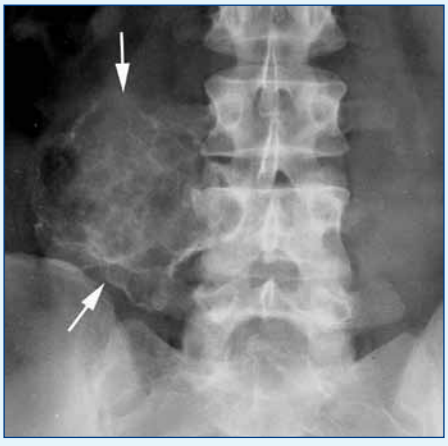
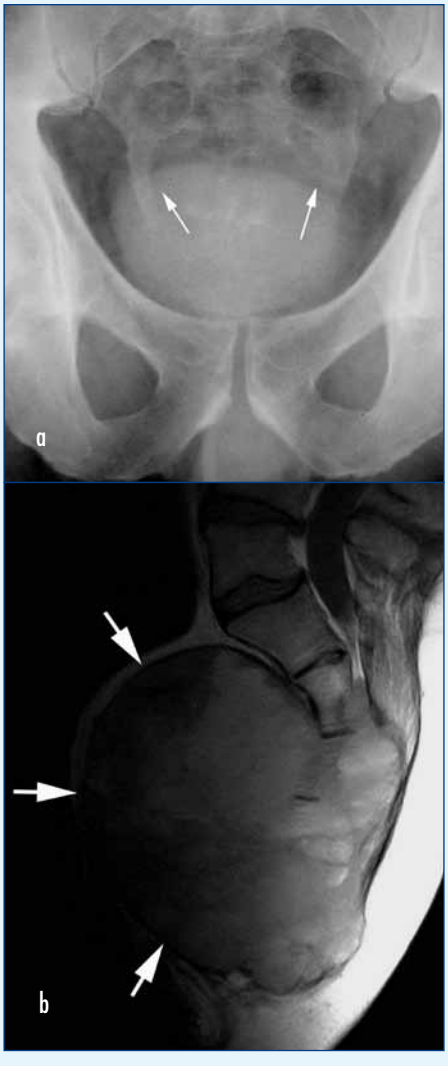


Figure 12. Chordoma. a. Anteroposterior radiograph of the sacrum showing marked destruction of the distal half of the sacrum (arrows). b. Sagittal T1W magnetic resonance image showing a huge tumour mass (arrows) arising from the S2 sacral segment.



Chordoma

Chordoma is a rare slow-growing tumour that arises from remnants of the notochord (Chugh et al, 2007). It represents 1–4% of all bone tumours between the ages of 55 and 65 years. Lesions in the sacral region may be asymptomatic, but eventually present with back pain, a feeling of fullness in the rectum and a change in bowel habits. X-rays shows areas of osteolysis, with possible expansion and osteosclerotic rimming (*Figures 12a* and *b*). Magnetic resonance imaging and computed tomography are useful to evaluate the extent of the tumour. Surgical resection is the rule with adjuvant radiotherapy treatment. Chordomas should be treated by aggressive complete excision with wide margins if possible, and given follow-up proton beam irradiation if available. If wide excision is not possible, then local recurrence is likely within a few years.

Conclusions

Primary bony tumours from the vertebral column comprise a large variety of important conditions. Early detection is associated with significant reduction in mortality and morbidity. Although the presentation may be subtle, an awareness and knowledge of these conditions will help in earlier detection. An understanding of these basic principles of spinal tumour investigation and treatment can be applied in all cases. A national database of patients with these conditions and outcome is required in the UK to predict future trends of these conditions. **BJHM**

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KEY POINTS

- Primary bony tumours of the vertebral column can be aggressive, so early detection and treatment is imperative.
- An awareness and knowledge of these conditions will help in earlier detection.
- Key investigations of primary bony tumours include X-ray, computed tomography, bone scan and magnetic resonance imaging.
- Early intervention including surgery, radiotherapy, and chemotherapy is key to reducing morbidity and mortality of the patient.