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Imaging — Pathological Correlation

Giant Cell Tumour of the Thoracic Spine

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ABSTRACT

Giant cell tumours of the spine above the sacrum are rare, and may pose diagnostic and treatment difficulties. A patient with thoracic vertebral giant cell tumour in which soft tissue extension and proximity to the spine complicated management is described. Magnetic resonance imaging is the imaging modality of choice for determining the extent of disease, assisting preoperative management, and monitoring for recurrent disease.

Key Words: *Giant cell tumour, Magnetic resonance imaging, Spine*

CLINICAL DETAILS

A 44-year-old woman presented to her doctor with right shoulder and scapular pain that improved with a 2-week course of ibuprofen. Six months later she presented again with continuing symptoms of right arm pain and paraesthesia involving the T1 and T2 nerve root distribution. Findings on physical examination included a 'rubbery', well-circumscribed, right supraclavicular fossa mass and right Horner's syndrome. Mild weakness and reduced coordination of the right arm and hand were evident.

Initial radiographic imaging of the thoracic spine (Figure 1) and the cervical spine (Figure 2) demonstrated a soft tissue lesion, with bony destruction at the right lung apex. Computed tomography (CT) scan of the spine showed an 8 cm mass arising from the T1 vertebral body (Figure 3). The mass extended into the adjacent C7, T1, and T2 foramina, with impingement on the C8 and T1 nerve roots, and extradural compression of the thecal sac. Magnetic resonance imaging (MRI) scan clearly showed the soft tissue extent of the lobulated mass, with involvement of the T1 and T2 vertebral bodies, spinal foramina, spinal canal, brachial plexus, and vessels (Figure 4). Image-guided core biopsies of the mass were obtained. Histology confirmed a giant cell tumour (GCT).

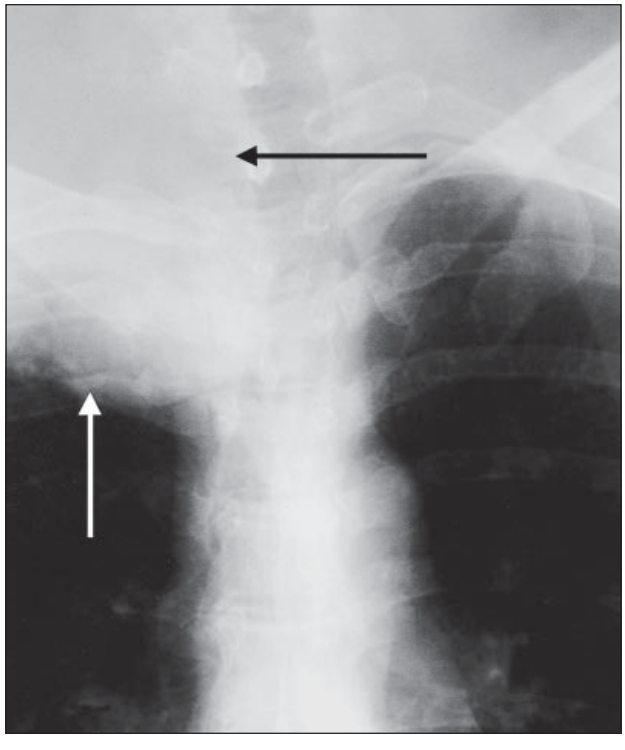

Figure 1. Frontal radiograph of the thoracic spine demonstrates a large soft tissue mass at the right lung apex (white arrow), with destruction of the right T1 transverse process, and right first rib posteriorly (black arrow), suggestive of a Pancoast tumour.

Prior to surgery, angiography and embolisation were performed to limit intraoperative bleeding (Figure 5).

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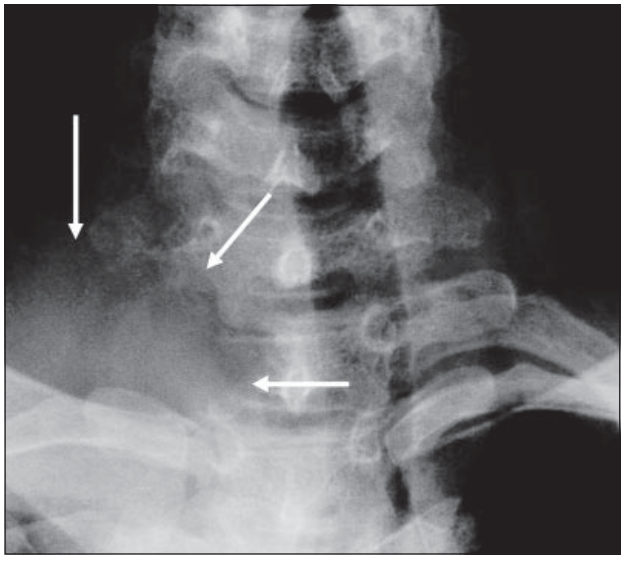

Figure 2. Frontal radiograph of the cervical spine demonstrates the extent of bony destruction of the right first rib and first thoracic vertebra.

A 2-stage operation was performed, with the initial stage involving a posterior approach. The definitive second stage required 20 consecutive operating hours and surgeons of different specialties. The tumour was removed with clear surgical margins, but required T1 vertebrectomy, C6 to T2 laminectomy, C5/6 to T1/2 discectomy, right first and second rib resections, and T2 nerve root resection. Spinal stabilisation was achieved using combined fibular and clavicular grafts, and spinal fixation hardware.

Postoperatively, the patient's pain resolved, with no analgesics required. Temporary C8/T1 nerve root impairment occurred, resolving within 6 months. The patient continues to have long-term follow-up due to the risk of recurrent disease.

DISCUSSION

Overall, GCTs account for approximately 5% of primary bone lesions[1] and 20% of benign skeletal tumours. Eighty five percent of bone tumours arise after closure of the epiphyses. They are usually solitary (85%), mildly expansile, and eccentric, and abut the subchondral plate of long bones, with a non-sclerotic margin. Soft tissue invasion is evident for approximately 25% of patients. The most common site is the knee (60%). Other common sites (in decreasing order of frequency) include the distal radius, sacrum, hands and feet, pelvis, and skull. Spinal GCTs above the level of the sacrum comprise approximately 1% to 3% of cases.[1,2,3,4] These lesions are usually centred in the vertebral body (55%), vertebral arch (17%), or within the body and arch (28%).

Patients with GCT may present with focal pain, tenderness, localised swelling (75%),[4] and pathologic fracture (5%). Pain is the most common presenting symptom for spinal GCT, with an average duration of 6 months. Nerve root symptoms (30%) or paraparesis (16%) due to local neurological involvement may also be presenting features.[3,5]

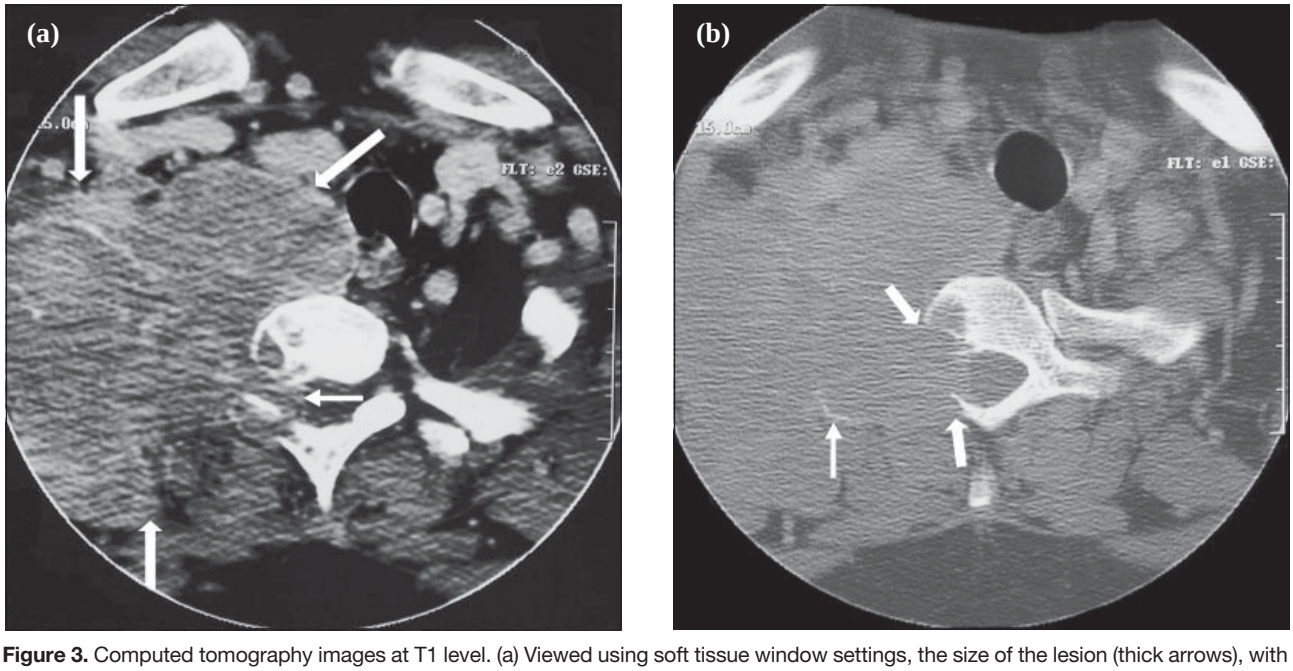

Figure 3. Computed tomography images at T1 level. (a) Viewed using soft tissue window settings, the size of the lesion (thick arrows), with extension into the intervertebral foramen and thecal sac compression (thin arrow) can be seen; (b) viewed using bone window setting shows vertebral body, right transverse process, pedicle, and lamina destruction (thick arrows). A thin shell of posterior calcification within the mass (thin arrow) is also shown.

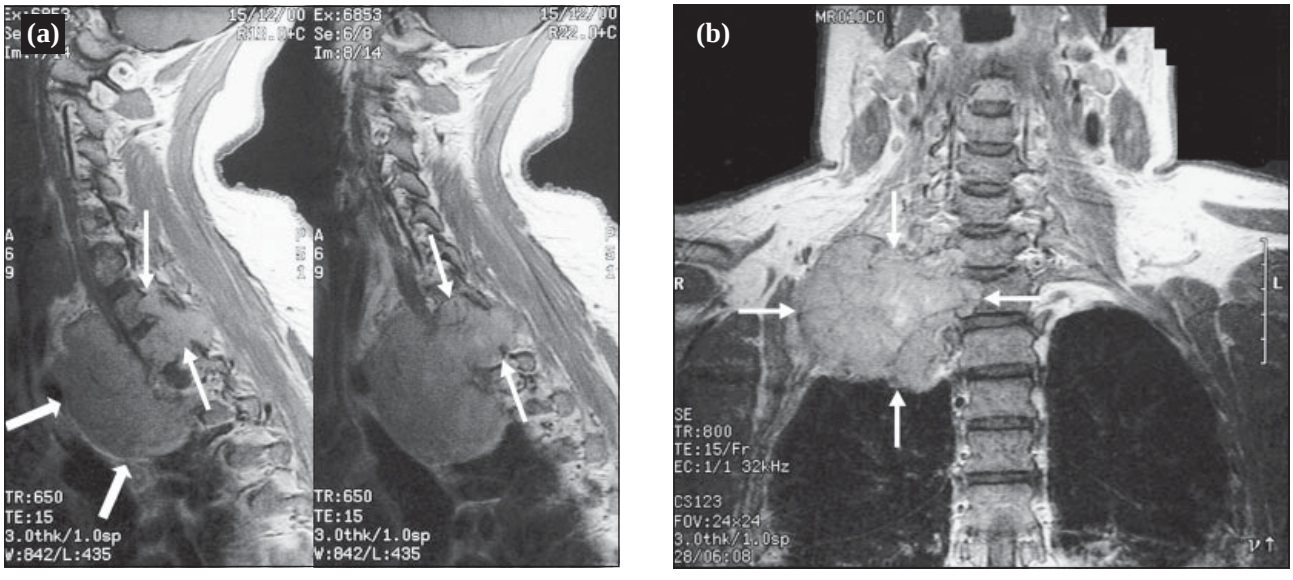

Figure 4. Magnetic resonance imaging scan of the lesion. (a) Sagittal T1-weighted image shows a lobulated soft tissue mass with foramen involvement (thin arrows) and a large anterior component (thick arrows); (b) coronal T1-weighted image showing a right apical soft tissue mass with bony involvement.

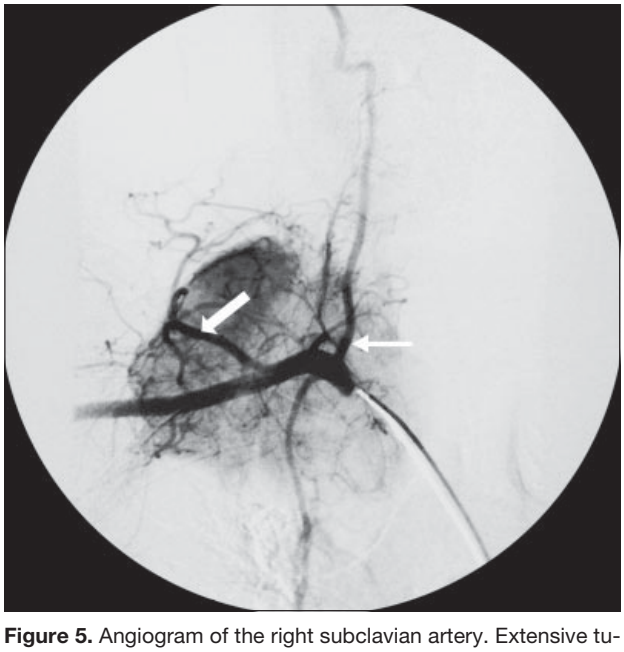

Figure 5. Angiogram of the right subclavian artery. Extensive tumour blush is evident, with predominant supply from the dorsal scapular artery (thick arrow). The tumour is also shown to encase the right vertebral artery (thin arrow)

Macroscopically, GCTs are typically haemorrhagic, dark red/tan-coloured soft masses. They expand the bone of origin and commonly demonstrate small cystic foci. A proportion of GCTs demonstrate vascular invasion. This feature is most commonly noted in tumours with extra-osseous extension. Microscopically, GCTs are composed of an admixture of plump, spindle-shaped, mononuclear cells and multinucleated, osteoclast-like giant cells (Figure 6). The nuclear morphology of the mononuclear cells and the individual nuclei of the

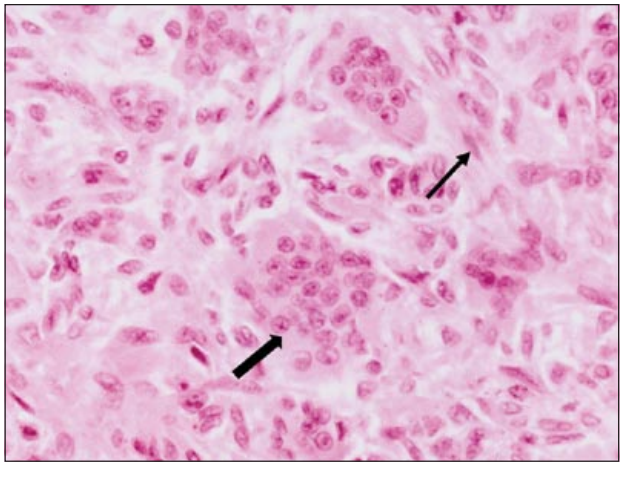

Figure 6. Histology slide showing multinucleated giant cells (thick arrow) and plump spindle shaped mononuclear cells (thin arrow).

multinucleated cells are similar, and it is thought that the multinucleated cells result from fusion of mononuclear cells. Individual tumour cells demonstrate pale-staining nuclei, with visible nucleoli. Mitotic figures are usually readily seen. Necrosis may also be present, particularly in the presence of pathological fracture. Marked nuclear pleomorphism and atypical mitotic figures are not seen in conventional GCT of bone and, if present, strongly suggest the diagnosis of a malignant neoplasm, such as malignant GCT of bone, or osteosarcoma. Since the multinucleated giant cells that characterise GCT of bone are a non-specific finding in many benign and malignant bone tumours, careful radiological correlation is required when making the pathological diagnosis of a bone tumour. This is particularly the case when establishing the diagnosis

using a small volume of tissue such as with needle biopsy specimens (as for this patient).

Plain radiographs of vertebral GCTs can show bony destruction, but significantly underestimate the degree of bony and soft tissue involvement. CT can more accurately assess both bone and soft tissue components, in particular encroachment into the intervertebral foramen and spinal canal. Classically, a fluid-fluid level can be seen within GCTs due to internal haemorrhage. However, this is a non-specific feature, which can be seen in other lesions such as aneurysmal bone cysts, chondroblastoma, and telangiectatic osteosarcoma.[6] Linear high-density areas within the tumour (described as a 'soap bubble' appearance),[7] may be enhanced after intravenous contrast. Shell-like marginal calcification may be present.[8] CT images can be of limited value, particularly for lesions in the region of the cervicothoracic junction, as image degradation due to beam hardening from the shoulder girdle can occur.

Providing high tissue contrast and multiplanar views, MRI is ideal for determining the extent of disease, particularly for assessing tumour involvement of the brachial plexus, great vessels, vertebral artery, and spinal cord. MRI of vertebral GCT shows an intermediate to low signal mass on T1-weighted sequence and variable enhancement (from non-necrotic portions) with gadolinium, although up to 10% of GCTs are shown on angiography to be relatively avascular. An encompassing rim of low signal (T1) has been described.[9] It has been suggested that this relates to haemosiderin deposition or reactive bone formation, although there has been no definitive histological confirmation of this premise to date. On T2-weighted sequences, there is variable internal high signal, with surrounding bone and soft tissue oedema. Fluid-fluid levels can be demonstrated within the lesion, as seen on CT, but are non-specific findings.

Treatment of spinal GCT is usually wide margin excision of the tumour. Patients treated within 3 months of neurological symptom onset have a greater chance of neurological recovery.[10] Extensive surgery can cause significant functional or cosmetic impairment, however, and wide resection margins are often difficult to achieve due to potential problems of maintaining spinal stability and neurological integrity. Radiation therapy can be considered for patients with incomplete resection or local recurrence,[1,5] and is balanced against the risk of malignant transformation documented in approximately 5% of patients. If metastatic disease occurs (approximately 5% of patients), this usually involves the lung.[1]

Local recurrence occurs in approximately 50% of patients,[3] usually within 3 years (for 80% to 90%).[6] Some series have reported similar recurrence rates for vertebral body GCT and GCT occurring at other sites,[2,5] but most reports indicate a substantially lower recurrence rate (approximately 25% lower) for vertebral GCT.[3,10,11,12] This is somewhat surprising given the general difficulty in obtaining clear surgical margins. MRI is the recommended modality for assessing recurrent disease.[6]

Key MRI features[6] are:

- presentation within 12 months of surgery
- a persistent high signal in the surgical bed
- a rounded, mass-like appearance with rapid and/or eccentric growth.

This appearance is similar to that of other conditions, such as giant cell granuloma. Ultimately, image-guided core biopsies may be required to confirm the presence of recurrent disease.

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