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CASE REPORT

Giant Cell Tumour of Bone as the Cause of Vertebra Plana in Young Patients: Two Case Reports

LY Lam, TWY Chin, KC Lai, MK Chan

Department of Diagnostic and Interventional Radiology, Queen Elizabeth Hospital, Hong Kong SAR, China

INTRODUCTION

Vertebra plana is a form of complete compression fracture of the vertebral body. Although Langerhans cell histiocytosis (LCH) is the most frequent associated entity,[1] there are other differential diagnoses such as giant cell tumour (GCT) of bone. We describe two cases of vertebra plana in young patients with GCT as the underlying cause.

CASE PRESENTATIONS

Case 1

A 30-year-old female presented to our orthopaedics department in September 2022 with back pain. She reported no history of trauma or constitutional symptoms. Physical examination revealed kyphosis at the upper thoracic spine and no focal neurological deficits. Initial radiograph demonstrated vertebra plana of the T5 vertebra with an ill-defined left pedicle (Figure 1). Positron emission tomography–computed tomography (PET/CT) showed a pathological compression fracture of the T5 vertebra (maximum standardised uptake value [SUV_{max}] = 4.6) and no lung masses or other hypermetabolic foci. Magnetic resonance imaging (MRI) revealed complete collapse of the T5 vertebra with involvement of the left transverse process, lamina and pedicle. The residual posterior portion of the T5 vertebra and an expanded left pedicle showed T1-weighted intermediate and T2-weighted heterogeneous signals with contrast enhancement (Figure 2). There was increased paravertebral soft tissue without intraspinal extension.

CT-guided biopsy of the T5 vertebra was performed in September 2022. Pathology revealed a giant cell-rich lesion with H3F3A G34W mutation, suggestive of GCT of bone. The patient had been treated with denosumab since November 2022. After the third dose of denosumab, follow-up CT showed increased sclerosis at the periphery and within the tumour (Figure 3), in keeping with denosumab effect. Follow-up MRI showed that the tumoural involvement was static. After nine doses of denosumab, total en bloc spondylectomy of the T5 vertebra was performed in May 2023. Final pathology of the T5 vertebra confirmed the diagnosis of GCT of bone.

Correspondence: *Dr LY Lam, Department of Diagnostic and Interventional Radiology, Queen Elizabeth Hospital, Hong Kong SAR, China*
Email: lly858@ha.org.hk

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Data Availability: All data generated or analysed during the present study are available from the corresponding author on reasonable request.

Ethics Approval: The patients were treated in accordance with the Declaration of Helsinki. Informed patient consent was obtained for Case 1 for the publication of the case report. Case 2 was lost to follow-up and its status was disclosed to the Kowloon Central Cluster and Kowloon East Cluster Research Ethics Committee of Hospital Authority, Hong Kong when approval was sought (Ref No.: IRB-2023-313). Potential identifiers have been removed to minimise the risk of identification.

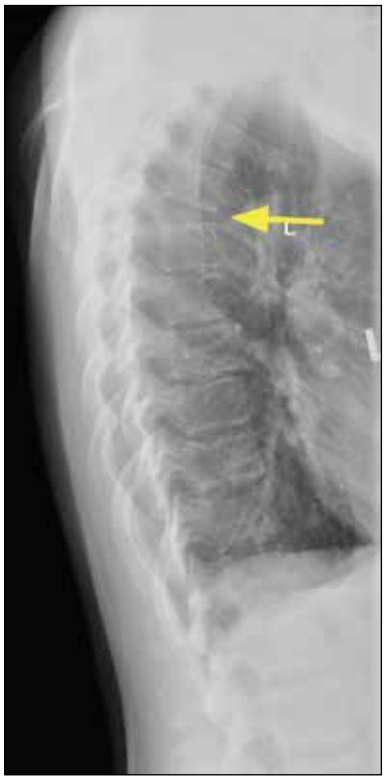

Figure 1. Case 1. Radiograph shows complete collapse of the T5 vertebral body (arrow), illustrating vertebra plana.

Case 2

A male adolescent was referred to a local hospital in March 2019. He presented with a 1-month history of neck pain radiating to the arms, with no history of trauma or constitutional symptoms. Physical examination revealed reduced range of movement due to pain and no neurological deficits. Radiograph showed vertebra plana of the C6 vertebra (Figure 4). PET showed complete collapse of the C6 vertebral body with a rim of hypermetabolism (SUV_{max} = 9.7), with involvement of the left lamina (SUV_{max} = 7.4) and intraspinal extension (Figure 5). There were no lung masses or other hypermetabolic foci. CT revealed vertebra plana of the C6 vertebral body with an underlying lytic bone lesion involving bilateral pedicles and partes interarticulares (Figure 6). MRI showed the lesion with T1- and T2-weighted hypointensity, avid contrast enhancement, an extraosseous epidural soft tissue component and severe canal stenosis (Figure 7).

Pathology from the CT-guided biopsy of the C6 vertebra revealed a giant cell-rich lesion that showed strong

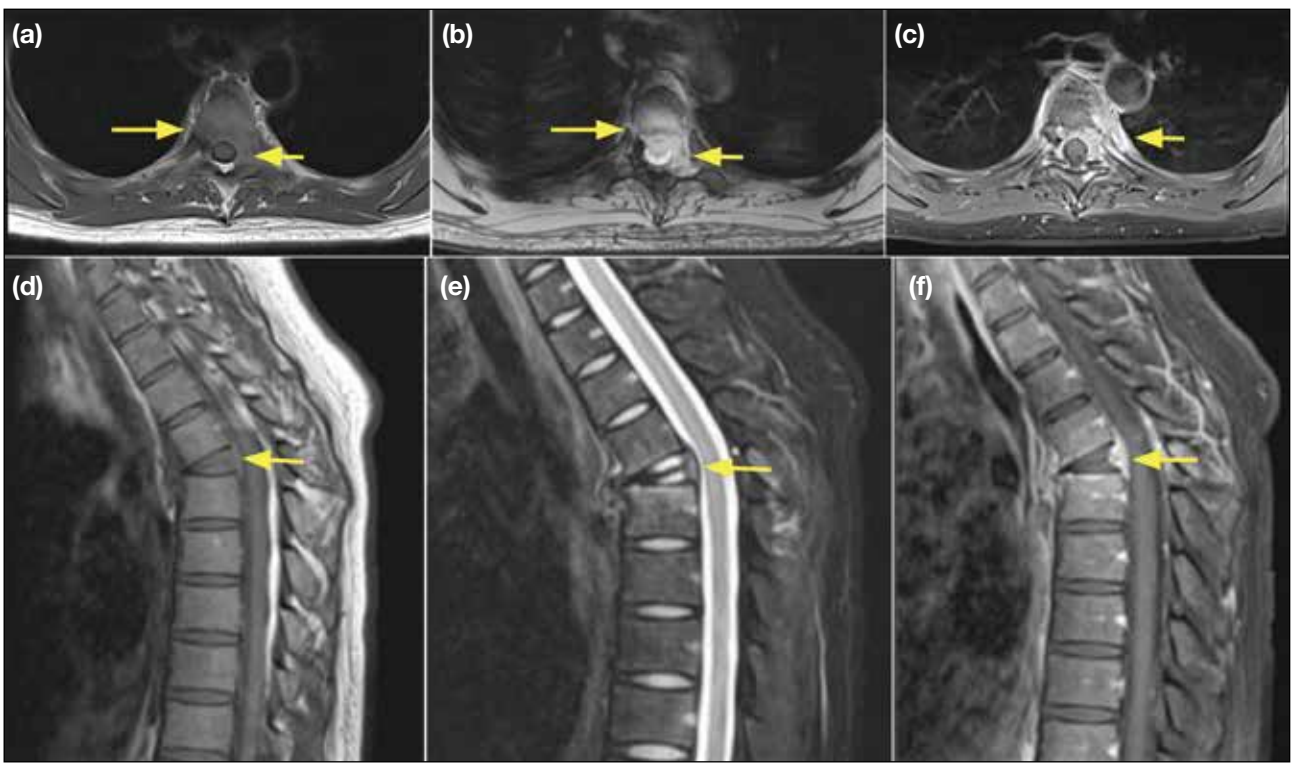

Figure 2. Case 1. Initial magnetic resonance imaging in axial cut (a-c) and sagittal cut (d-f). (a, d) T1-weighted images show intermediate signals involving the residual posterior portion of the T5 vertebral body and an expanded left pedicle (arrows). (b, e) T2-weighted images show heterogeneous signals involving the same parts (arrows). (c, f) Post-gadolinium images show avid enhancement of the involved parts. Contrast enhancement seen in the medial aspects of the bilateral posterior fifth ribs without corresponding abnormal marrow replacement signals in T1-weighted imaging is regarded as reactive change (arrows). An increase of paravertebral soft tissue without intraspinal extension is seen.

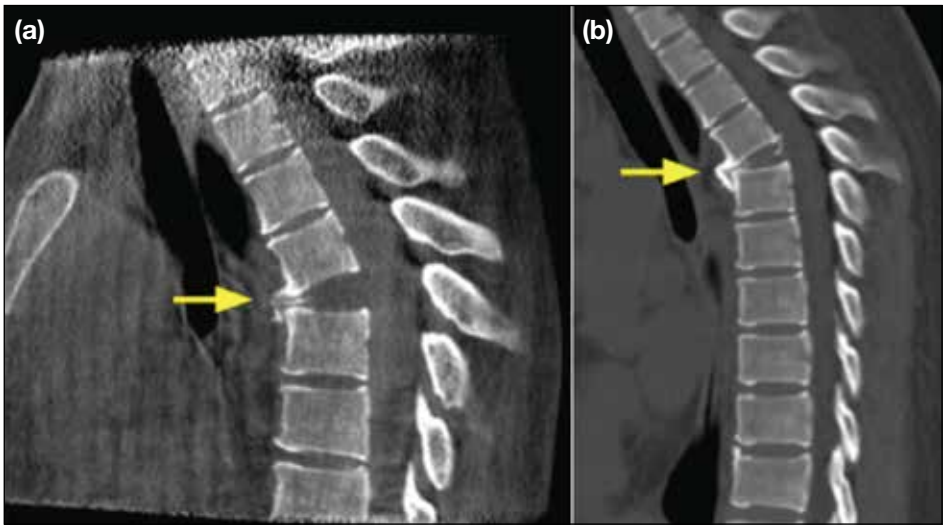

Figure 3. Case 1. Computed tomography (CT) sagittal images. (a) Initial CT image shows vertebra plana of the T5 vertebral body with normal posterior element and associated kyphotic deformity (arrow). (b) CT performed after the third dose of denosumab shows increased sclerosis at the periphery of the collapsed vertebral body (arrow).

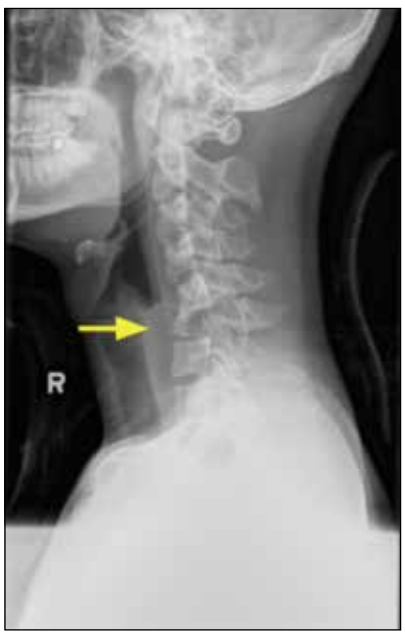

Figure 4. Case 2. Radiograph of the cervical spine shows vertebra plana of the C6 vertebral body (arrow).

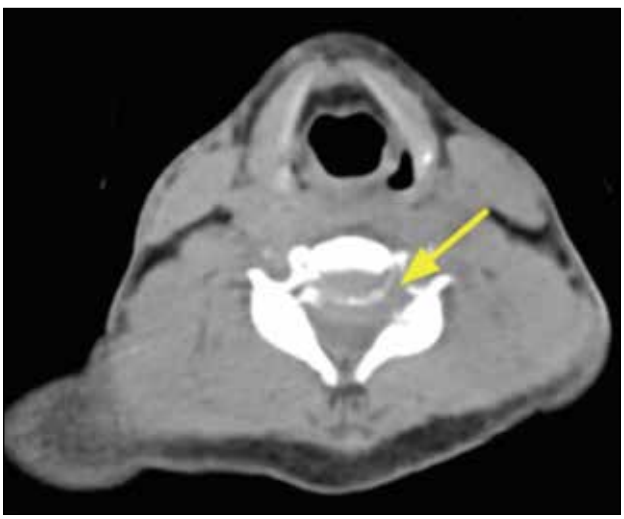

Figure 6. Case 2. Plain computed tomography axial image shows a lytic bone lesion in the severely collapsed C6 vertebral body with faintly seen extraosseous soft tissue component in the epidural space (arrow).

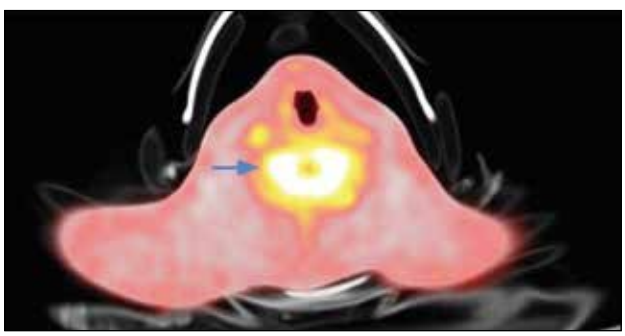

Figure 5. Case 2. Positron emission tomography image shows a rim of increased metabolism around the collapsed C6 vertebral body (arrow), with intraspinal extension of the hypermetabolism noted.

nuclear staining with H3.3 G34W mutant protein, suggestive of GCT of bone. The patient had been treated with denosumab since June 2019. After the fourth dose of denosumab, follow-up MRI showed marked shrinkage of the tumour with only a small residual enhancing area over the right pedicle (Figure 8). In view of the good response to denosumab, the patient and his family opted for non-operative treatment and continued denosumab therapy. The patient was lost to follow-up after January 2020.

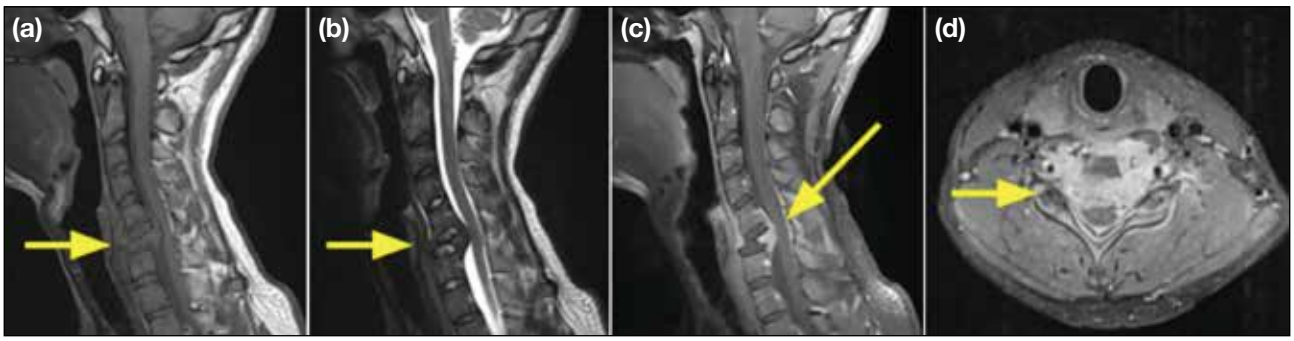

Figure 7. Case 2. Initial magnetic resonance imaging. (a) T1-weighted and (b) T2-weighted sagittal images show vertebra plana of the C6 vertebra with hypointense signals (arrows). (c) Post-gadolinium sagittal image and (d) axial image show avid enhancement of the lesion involving bilateral pedicles with associated extraosseous soft tissue component in the epidural space with severe canal stenosis (arrows).

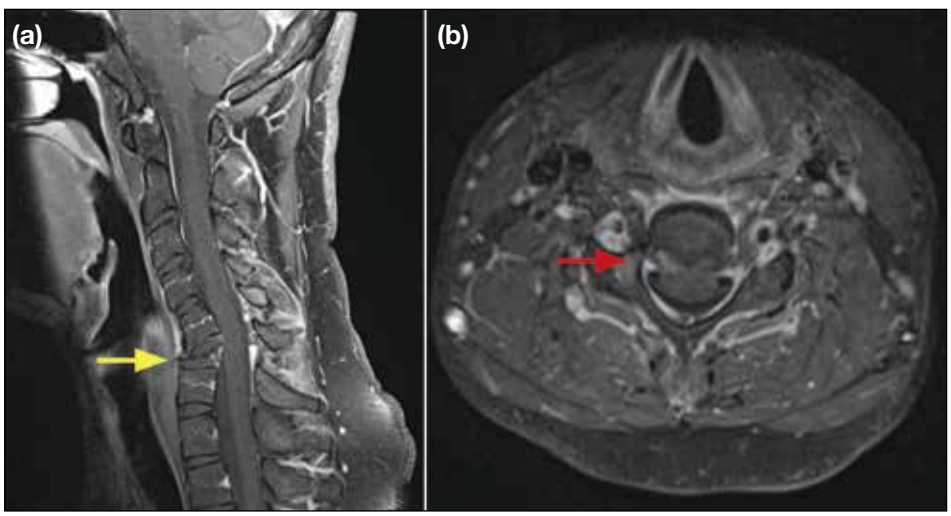

Figure 8. Case 2. Follow-up magnetic resonance imaging after four doses of denosumab. (a) Post-gadolinium sagittal image and (b) axial image show marked shrinkage of the enhancing tumour (yellow arrow) and improvement in spinal stenosis, with only a small residual enhancing area over the right pedicle seen (red arrow).

DISCUSSION

GCTs are benign bone tumours that can be locally aggressive and metastasise to the lungs.[2] It is one of the most common bone tumours and accounts for 4% to 9.5% of primary bone tumours and 18% to 23% of benign bone tumours.[2] Most patients with GCTs are in their third or fourth decade of life, with a slight predominance in females.[3] Our first case of a female in her third decade exemplifies a typical demographic profile of GCT.

The typical location of GCTs is the ends of long bones, particularly the distal femur and proximal tibia (up to 65%).[4] The spine is a relatively rare location for GCTs, accounting for only 7% of all cases.[5] Within the spine, GCTs most commonly occur at the sacrum, followed by the thoracic, cervical and lumbar spine in descending order of frequency.[6]

A large case series[6] showed that 85% of GCTs affecting the spine arose from the vertebral body with involvement of the vertebral arch, while others arose from the posterior elements. All cases in the series affected the vertebral bodies and pedicles.[6] This feature enables us to differentiate GCTs from other primary spinal bone tumours such as osteoid osteoma, osteoblastoma and aneurysmal bone cysts (ABCs), since these tumours preferentially affect the posterior elements.

Imaging features of GCTs on radiographs include an expansile appearance and osteolysis. On CT, spinal GCTs appear as well-defined soft tissue density masses with a sclerotic rim and without internal mineralised matrix. They typically show avid contrast enhancement due to their hypervascular nature. On MRI, spinal GCTs exhibit low to intermediate T1-weighted signal intensities with gadolinium enhancement. On T2-weighted images, most spinal GCTs show heterogeneous high signals with areas of low signal intensities due to hemosiderin or fibrosis.[7]

Differential diagnoses of a spinal bone lesion showing T2-weighted hypointense signals are limited and include myelofibrosis, osteoblastic metastases and spinal GCT.[7] The first two do not match our cases. In myelofibrosis, the diseased bone shows homogeneous low signals, and myelofibrosis does not cause vertebra plana. Our patients' young age and lack of known primary malignancy do not suggest metastases.

A spinal GCT may result in collapse of the vertebral body, ranging from mild collapse to rarely, vertebra plana, as seen in our cases. Vertebra plana was first described by Calve in 1925 as 'extreme collapse of a vertebral body with sparing of the posterior elements and slightly widened adjacent disc spaces'.[8] Later, most reports described it as collapse of a single vertebral body with increased density, normal adjacent intervertebral discs and increased height of intervertebral space by at least one-third of normal.

Vertebra plana is often thought to be pathognomonic for LCH in young patients. This is a false assumption since many different conditions can cause vertebra plana, including ABCs, GCTs, lymphomas, and metastases.[9] As LCH is more commonly seen in paediatric patients,[5,10] it is an unlikely diagnosis based on our patients' demographics. In addition, LCH causes prominent oedema in the adjacent bone and soft tissues, which are absent in our cases. Spinal lymphoma can present as an osteolytic lesion and cause vertebra plana, though this is exceedingly rare and usually involves multiple levels.[11] Our cases also do not present with any B symptoms. An ABC typically affects the posterior elements with pure cystic components, making it unlikely in our cases. The age of our patients is not typical for plasmacytoma. Infection is also an important differential diagnosis but the intact endplates and intervertebral discs, absence of fluid collection, and the chronicity in our cases speak against infection.

The ideal modality of treatment for spinal GCTs is complete surgical resection. Some cases may present technical difficulties, especially if the tumour is located in the cervical spine with complicated surrounding anatomy, as seen in Case 2.

Denosumab has been suggested as an alternative therapy. It is a human monoclonal antibody that binds the receptor activator of nuclear factor kappa-beta ligand, preventing activation of its receptor on the surface of giant cells and osteoclasts. This inhibits osteoclast formation and reduces bone resorption in GCTs. A study reported that 96% of patients with surgically unsalvageable GCTs treated with denosumab showed no progression.[12] A report described complete regression of spinal GCTs following denosumab treatment without the need for surgical resection.[13]

Additionally, denosumab acts as a preoperative adjunct in cases of spinal or pelvic GCT. It reduces extraosseous soft tissue lesions, hardens the GCT, and therefore facilitates en bloc resection, as seen in Case 1. For GCTs in the extremities, denosumab treatment is not routinely recommended as curettage remains the mainstay of treatment and denosumab prior to curettage is known to increase the local recurrence rate,[14] possibly due to the effect of increased osteosclerosis, rendering it difficult to identify the tumoural involvement intraoperatively.

The optimal duration of denosumab treatment is unknown. Some authors recommend a neoadjuvant therapy duration of 3 to 4 months[15] but further studies are required to determine the optimal duration.

In summary, we report two cases of vertebra plana in young patients caused by GCT of bone. Although GCT remains a rare cause of vertebra plana, it should be considered as a differential diagnosis. Surgery is the mainstay treatment while denosumab therapy is helpful as a preoperative adjunct or as an alternative treatment in surgically challenging cases.

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