Clinical Case Report

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Primary malignancy in giant cell tumor of thoracic vertebrae

A case report

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Abstract

Rationale: Primary malignancy in giant cell tumor of bone (PMGCTB) is extremely unusual. PMGCTB in the thoracic vertebrae is particularly rare.

Patients concerns: A 23-year-old man was admitted with a chief complaint of chest pain associated with cough for approximately 3 days. Physical examination revealed a palpable, immobile, tender, 7 cm mass in the right paravertebral area of the thoracolumbar spine.

Diagnosis: Computed tomography images revealed an osteolytic, expansive, and eccentric lesion on the vertebral bodies and right accessory processes with spinal cord compression in the thoracic vertebra, with right rib also having bone destruction. Magnetic resonance imaging revealed multiple fluid–fluid levels occupying more than one-third of the lesions. On the basis of the imaging and pathological findings, the final pathological diagnosis was PMGCTB with aneurysmal bone cyst.

Interventions: The patient underwent successful wide spondylectomy of T9/10 to remove the tumor, and adjuvant chemotherapy based on the protocol used for osteosarcoma.

Outcomes: After 4 years of follow-up, there is no clinical or radiological evidence of recurrence.

Lessons: PMGCTB is difficult to distinguish from giant cell tumor of bone. PMGCTB should be considered when lesions appear with multiple fluid–fluid levels and soft tissue mass.

Abbreviations: ABC = aneurysmal bone cyst, CT = computed tomography, GCRO = giant cell-rich osteosarcoma, GCTB = giant cell tumor of bone, MGCTB = malignant giant cell tumor of bone, MRI = magnetic resonance imaging, PMGCTB = primary malignant in giant cell tumor of bone, SMGCTB = secondary malignant in giant cell tumor of bone, TO = telangiectatic osteosarcoma.

Keywords: bone tumor, giant cell, malignancy

1. Introduction

Giant cell tumor of bone (GCTB) usually appears as a benign tumor with local aggressiveness.[1] Malignant giant cell tumor of bone (MGCTB) is rare and is currently described as either primary or secondary. Only a few cases have been described in the literature to date. Most published articles on primary malignancy in giant cell tumor of bone (PMGCTB) report a small number of cases.[2–5] PMGCTB is difficult to distinguish from GCTB, and most cases of PMGCTB have been single lesions, frequently located in the long bones around the knee joint. Herein, we report a case of multicentric PMGCTB, located in the thoracic vertebra and right rib.

2. Case presentation

A 23-year-old man was admitted with the chief complaint of chest pain associated with cough for approximately 3 days. He denied a history of tuberculosis and had no history of surgery or trauma. His medical history was unremarkable.

Physical examination showed an approximate 7 cm diameter mass that could be palpated in the right paravertebral area of the thoracolumbar spine; the mass was slightly hard, immobile, and obviously tender. The mass margin was not clear and was adherent to the adjacent tissue. No local superficial venous distention was observed around the thoracic vertebra. His vital signs were normal, with oxygen saturation 99%.

Computed tomography (CT) (Fig. 1–3) demonstrated an osteolytic, expansive, and eccentric lesion on the vertebral bodies and right accessory processes, with spinal cord compression at the T9/10 level, with right rib also having bone destruction. The bone destruction penetrated the local cortical bone with a large mass around it. The mass of density was inhomogeneous. The CT value was decreased from 40 to 20 Hu. A thin and discrete rim of bone was seen around the mass. In addition, the adjacent rib showed osteolytic and expansive destruction, and the cortical bone of the adjacent rib was thin. The contrast-enhanced CT showed obvious inhomogeneous enhancement of the lesions.

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The authors declare that they have no conflict of interest.

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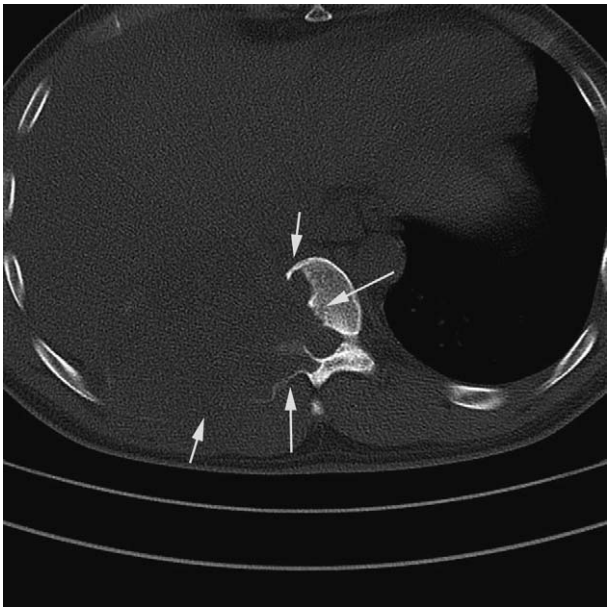

Figure 1. Computed tomography (CT) scan shows an osteolytic, expansive, and eccentric lesion on the vertebral and accessories (arrow heads). The lesion density is nonuniform.

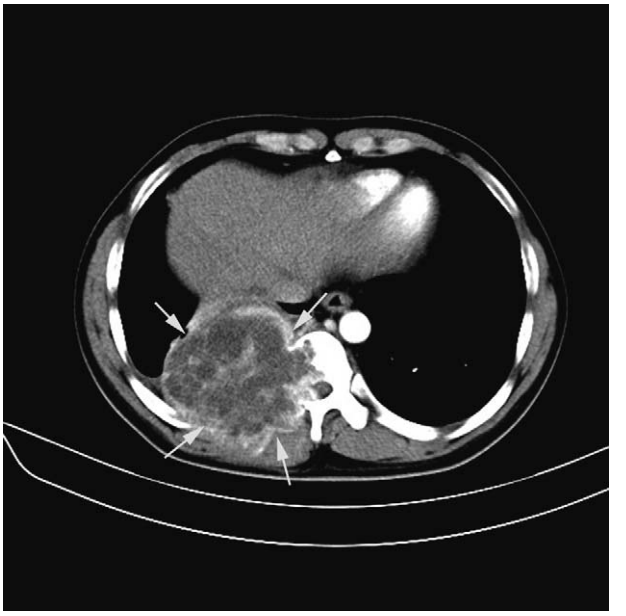

Figure 3. Contrast-enhanced CT scan shows an osteolytic, expansive, and eccentric lesion on the vertebral and accessories (arrow heads). The lesion density is nonuniform.

Magnetic resonance imaging (MRI) (Fig. 4–6) found that the lesion had inhomogeneous signals and low signal intensity in T1-weighted MR images, and relatively mix signal intensity in T2-weighted images. The margin between the lesion and the preserved bone was clear. Multiple fluid–fluid levels could also be seen; fluid–fluid levels were observed in more than one-third of the lesions. Some pleural effusion was seen in the right thoracic cavity.

Due to bone destruction on the vertebral bodies and right accessory processes and multiple cysts in the lesion, the diagnosis based on CT and MRI findings was malignant tumor with aneurysmal bone cyst (ABC). Wide spondylectomy of T9/10 was performed to resect the paravertebral tumor, with reconstruction using an endoprosthesis (Fig. 7). The surgical findings were as follows: The mass, which grew into the thoracic cavity, was located in the T9/10 thoracic vertebrae and paravertebral structures. It infringed on the 9th and 10th ribs and invaded the partial diaphragm. The mass invaded the spinal canal, resulting in cord compression.

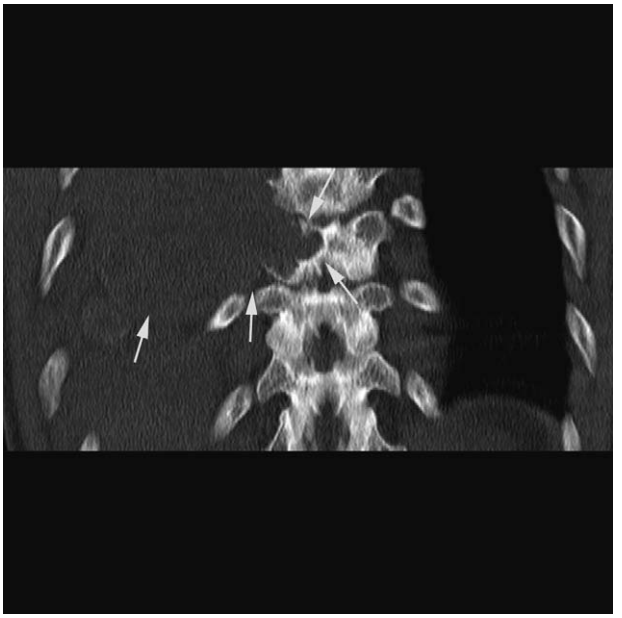

Figure 2. Computed tomography (CT) scan shows an osteolytic, expansive, and eccentric lesion on the vertebral and accessories (arrow heads). The lesion density is nonuniform.

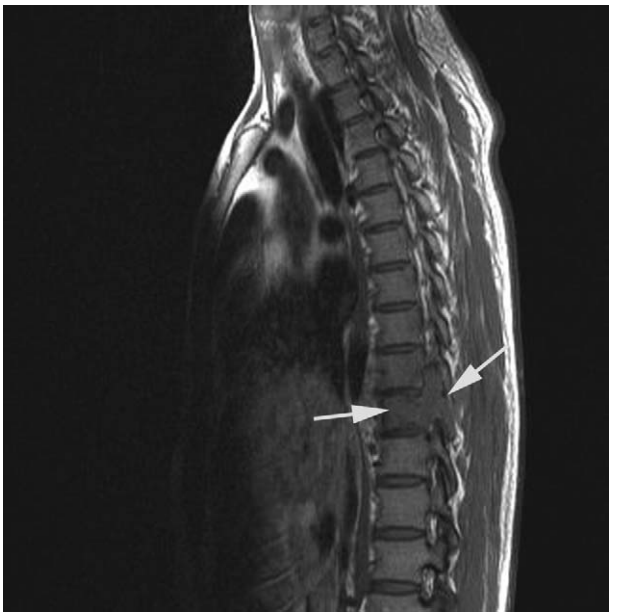

Figure 4. Sagittal T1-weighted image shows a large, irregular mass in the vertebral and accessories (arrow heads); the lesion contains multiple fluid–fluid levels.

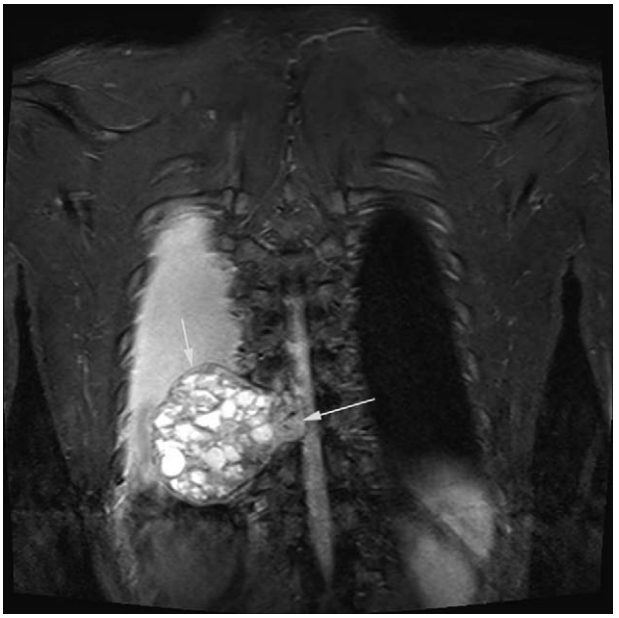

Figure 5. Coronal image shows a large, irregular mass in the vertebral and accessories (arrow heads); the lesion contains multiple fluid–fluid levels.

The gross pathology (Fig. 8) findings were as follows: The tumor with its complete capsule was resected integrally. It measured 9 cm × 8 cm × 6 cm. On the cut surface, the mass was grey-white. Some residual blood was seen in the cystic area. The solid mass was soft on palpation and grey-pink.

The microscopic findings (Fig. 9) were as follows: A large number of nuclear giant cells and powder stained bone matrix were observed. Immunohistochemical examination showed positive staining for CD163, CD68, SMA, and Ki-67, and negative staining for S-100 and P53. The presence of osteoid matrix in partial areas, cells with mild-to-moderate atypia, multiple nerve invasion, and vascular tumor thrombus were observed, suggesting malignant transformation. On the basis of the above findings, PMGCTB with ABC was the pathological diagnosis.

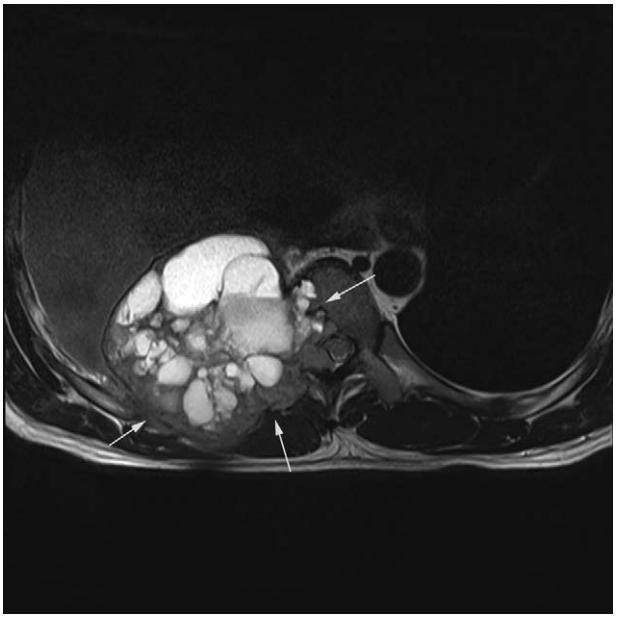

Figure 6. Axial T2-weighted image shows a large, irregular mass in the vertebral and accessories (arrow heads); the lesion contains multiple fluid–fluid levels.

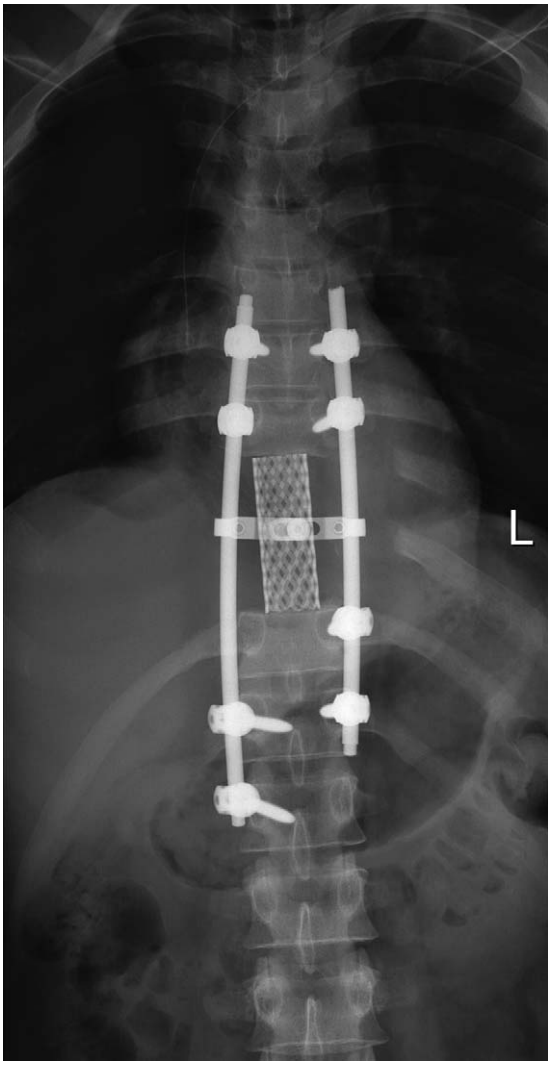

Figure 7. Radiograph showing wide resection of T9&10 with internal fixation.

After surgery, adjuvant chemotherapy based on the protocol used for osteosarcoma was administered. After 4 years of follow-up, the patient has no clinical or radiological evidence of recurrence.

The patient provided written informed consent. In this case, the patient accepted standard, proven diagnosis and therapy in the Clinical Department of Bone and Soft Tissue Tumor Department, so ethical approval was not necessary.

3. Discussion

PMGCTB typically occurs in the distal femur, proximal tibia, and distal tibia,[5] and its reports are extremely rare in the literature. We could not find any case reports of PMGCTB of the thoracic vertebrae. The radiologic features of the PMGCTB are similar to those of conventional GCTB: osteolytic lesions with well-circumscribed margins, cortical breakthrough, with soft tissue mass.[6]

Figure 8. Gross appearance shows complete resection of the tumor with a complete capsule, measuring 9 cm × 8 cm × 6 cm. The cut surface of the mass is grey-white. Some residual blood is seen in the cystic area.

MGCTB is an extremely rare entity with a very poor prognosis and only accounts for 2% to 9% of GCTB.[2–4] This disease was initially called "giant cell tumor with various degrees of anaplasia." Later, the diagnostic criteria of MGCTB were further elaborated by Dahlin et al[4] and Hutter et al,[5] and the disease was subdivided into PMGCTB and secondary malignant giant cell tumor (SMGCTB). PMGCTB is a high-grade sarcoma that arises side-by-side with GCTB, whereas SMGCTB is a high-grade sarcoma that occurs as a recurrent lesion at the site of GCTB previously treated by either radiotherapy or surgery. MGCTB is very uncommon, whereas SMGCT is relatively common.

SMGCTB can be osteosarcoma, fibrosarcoma, malignant fibrous histiosarcoma, or undifferentiated sarcoma. In the literature, more SMGCTB are fibrosarcomas than other sarcomas, whereas more PMGCTB are osteosarcomas than other sarcomas.[2,3] For the patient in the present case study, the PMGCTB was osteosarcoma.

The sites of MGCTB are similar to those of conventional GCTB, which preferentially involve invasion of the ends of long bones, including the distal femur, proximal tibia, and distal tibia. The sacrum can also be involved, but rarely the thoracic vertebra. This case showed multicenter lesions with invasion of multiple thoracic vertebrae and ribs. The rarest presentation of GCTB is multicentric GCTB, which has 2 or more separate GCTB lesions confirmed by histopathology. Multicentric GCTB is further subdivided into synchronous and metachronous. Synchronous GCTB presents with multiple GCTB lesions at once or with the second GCTB diagnosed within 6 months of the initial GCTB. Metachronous GCTB is a second GCTB diagnosis 6 months after the initial GCTB.

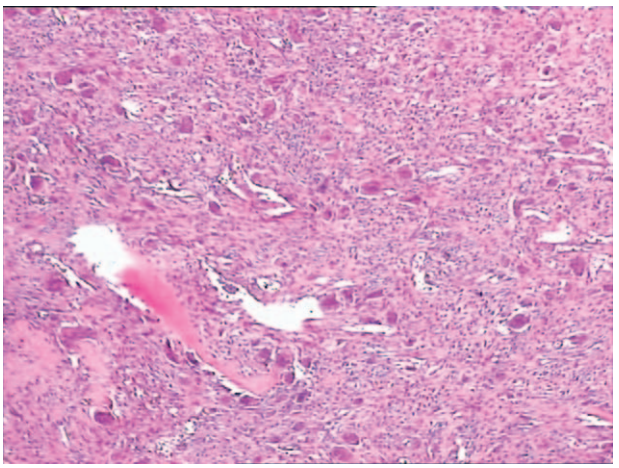

Figure 9. Photomicrograph of primary malignancy in giant cell tumor of the bone. The giant cell component of the tumor shows numerous mitoses (hematoxylin and eosin, ×40).

Although patients with MGCTB tend to present at an older age than those with conventional GCTB, the patient in the present case was young; we think that this may be related to the presence of multicentric lesions, because multicentric GCTB patients tend to present at an earlier age than those with GCT.[7]

The clinical manifestations of MGCTB are always nonspecific. The most common symptoms are pain and swelling, which may last for months. The radiologic features of MGCTB are similar to those of conventional GCTB: an osteolytic lesion with thinning or fading cortical bone, sometime breaking through the cortical bone to form a vast soft tissue mass. Occasionally, a periosteal reaction can be seen. However, in most cases, radiologic features are not diagnostic for malignancy, and the final decision mainly relies on pathology. In the present case, there was osteolytic bone destruction, with interrupted bone shell and bone crest, which is similar to conventional GCTB, but there was also a vast soft tissue mass around the bony lesion, and there were multiple small fluid–fluid levels in the lesion; these indirect signs may hint at malignancy.[8] Furthermore, when fluid–fluid levels are observed, it is important to consider that ABC can coexist with MGCTB, but the fluid–fluid levels in ABC are often large and occupy the entire volume of the lesion.[9] SMGCTB has a much more malignant appearance on imaging (e.g., soft tissue mass, periosteal reaction, etc), but some cases of SMGCTB are indistinguishable from benign recurrences of GCTB. The final diagnosis relies mainly on pathology.

MGCTB can very often be confused with ABC, which presents at a younger age and has more expanded lesions than MGCTB. In ABC, the lesion usually invades the vertebral appendix, it has more and larger fluid–fluid levels than MGCTB, and a soft tissue mass is not present. The differential diagnosis of MGCTB should also include telangiectatic osteosarcoma (TO) and giant cell-rich osteosarcoma (GCRO), both of which present at a younger patient age than MGCTB.[10,11] Currently, no published studies have described the imaging features of GCRO in the spine. GCRO in the long bone usually presents as expansile, eccentric, and osteolytic lesions, and sometimes a soft tissue mass and periosteal reaction are observed.[12] Because it is often difficult to distinguish MGCTB from other conditions, pathologic examination is usually needed. The imaging findings of TO are an osteolytic, destructive tumor with limited or no matrix mineralization or periosteal new bone formation, usually with multiple and small fluid–fluid levels.[10]

4. Conclusion

PMGCTB can exist as multicentric lesions that are located in multiple vertebrae and/or ribs, with cortical bone destruction and a soft tissue mass, and with multiple and small fluid–fluid levels. PMGCTB is difficult to distinguish from GCTB. When a lesion appears with multiple fluid–fluid levels and a soft tissue mass, PMGCTB should be considered in the differential diagnosis.

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Validation: jianling Cui.
Writing – original draft: hong Yu.
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