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Atypical Presentation of Giant Cell Tumor in the Dorsal Spine and Management of Recurrence: A Case Report and Literature Review

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Abstract

Giant cell tumors (GCTs) of the bone are locally aggressive primary bone tumors with a benign nature. GCTs usually occur in long bones, but in the spine it is rare. In this report, we present a rare case of recurrent GCT with dorsal myelopathy managed through staged resection with neoadjuvant and adjuvant denosumab and radiation therapy. Here we present a case of a 37-year-old male patient who presented with sudden-onset back pain while playing football, paraplegia, and urinary incontinence for two days. MRI revealed a heterogeneous lesion involving the D11 vertebral body with cord compression. The patient underwent emergency posterior decompression separation surgery and fixation. Post Operative patient clinically improved and ambulatory after 1 month. On follow-up, the patient again developed partial weakness. A repeat MRI at the 16-month follow-up revealed tumor recurrence—subsequently tumor excision was done in a staged approach. At the 24-month follow-up, the patient was clinically stable with no deficit and no recurrence on follow-up MRI. The selection of an anterior, posterior, or combined approach should be guided by the tumor location and extent, aiming to achieve complete resection for effectively managing large recurrent GCTs of the spine. Denosumab therapy plays a crucial role in facilitating complete resection. Additionally, local radiation therapy reduces the likelihood of recurrence.

Keywords: Giant cell tumour, Dorsal spine, Recurrence, Denusumab

INTRODUCTION

Giant cell tumor (GCT) is a benign but locally aggressive primary bone tumor, composed of multinucleated osteoclast-like giant cells, proliferated mononuclear cells, and stromal spindle cells[1].

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GCT of the bone is relatively rare, accounting for only 10% of all primary bone neoplasms; of these, only 5% to 10% of GCTs are found in the spine, with the sacrum being the most common site of vertebral involvement. There is a discernible peak in incidence during the third decade of life, with a slight female predisposition.[2]

When a GCT is situated in a mobile spine, the preferred approach is en-bloc resection with wide margins to minimize the risk of local recurrence. Still, morbidity is high due to potential issues such as bleeding, infection, and neurological deficits. Surgical treatment for spinal GCTs is associated with a high recurrence rate, estimated at approximately 25–50%. Hence, the primary focus of treatment is to reduce the likelihood of recurrence.[3] Denosumab is a fully human monoclonal antibody targeting the receptor activator of nuclear factor-κB (RANK) and RANK ligand (RANKL). It is effective in as many as 86%–88% of patients with GCTs located in the extremities. Specifically, continuous denosumab injections led to pain relief, reduction in tumor size, and the development of bone within the tumor and/or at its periphery.[4] In this case report, we present a patient with a GCT at the D11 vertebral level who initially underwent decompression and separation surgery and commenced adjuvant denosumab therapy. However, the patient experienced a GCT recurrence, necessitating resection through a staged anterior and posterior approach.

CASE REPORT

A 37-year-old male presented with sudden onset gradually progressive paraparesis with associated back pain for 15 days, initially noticed during a game of football. He experienced gradual bilateral lower limb weakness and urinary incontinence for 3 days. On clinical assessment, the patient exhibited Grade 0 power on MRC grading, sensory loss below D12 level, brisk deep tendon reflexes of the knee and ankle, and plantar upgoing. MRI revealed a heterogeneous lesion involving the D11 vertebral body, posterior elements, and adjacent left rib and paraspinal muscle musculature with significant compression of the lower dorsal cord and conus medullaris(Fig 1).

The patient underwent emergency surgical decompression of the cord through separation surgery and stabilization from D9 to L1 through a posterior approach(Fig 4). Postoperatively, his incontinence improved, the catheter was removed after 15 days, and lower limb power regained fully over 1 month with aggressive physiotherapy. Histopathology confirmed a GCT comprising neoplastic mononuclear stromal cells with uniformly distributed osteoclast-like giant cells. Monthly denosumab injections were administered for 6 months and he was kept under clinical and 6 monthly radiological observations.

He remained symptom-free for 16 months but then developed lower limb weakness and urinary urgency. A follow-up scan revealed tumor recurrence with advancing size(Fig 2,3). Staged transthoracic tumor resection, D11 corpectomy, and anterior column reconstruction with cage and plate fixation were done. Subsequently, he underwent posterior decompression with a posterolateral paraspinal tumor resection (Fig 5). Histopathology confirmed GCT. Postoperatively, local RT was administered and Denosumab continued for 6 months. He clinically improved during follow up regaining full power. At 24 months, scans showed no evidence of recurrence.

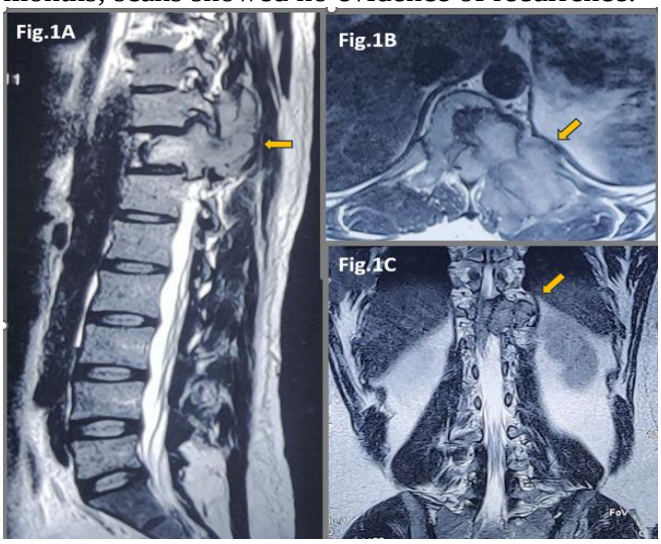

Fig. 1A, 1B & 1C: MRI at presentation reveals a heterogeneous lesion involving the D11 vertebral body, posterior elements, adjacent left rib, and paraspinal musculature. The lesion causes significant compression of the lower dorsal cord and conus medullaris.

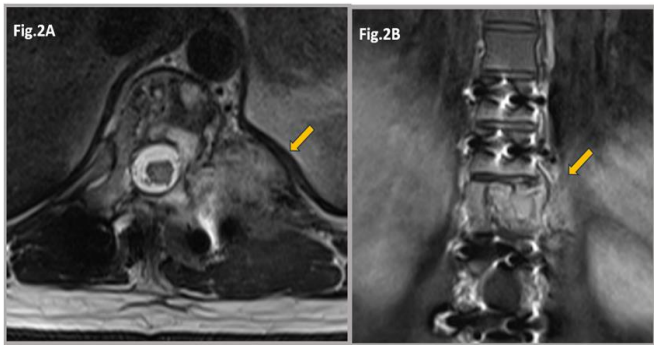

Fig. 2A & 2B: MRI at 12 months post intralesional curettage and fixation showing recurrent giant cell tumor (GCT).

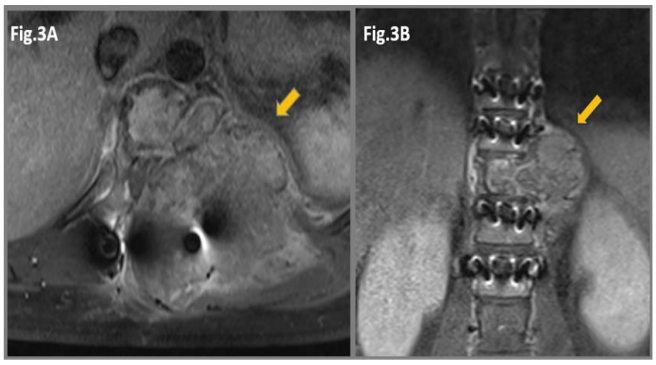

Fig. 3A & 3B: MRI at 15 months post intralesional curettage and fixation showing a significant increase in the size of the recurrent giant cell tumor (GCT).

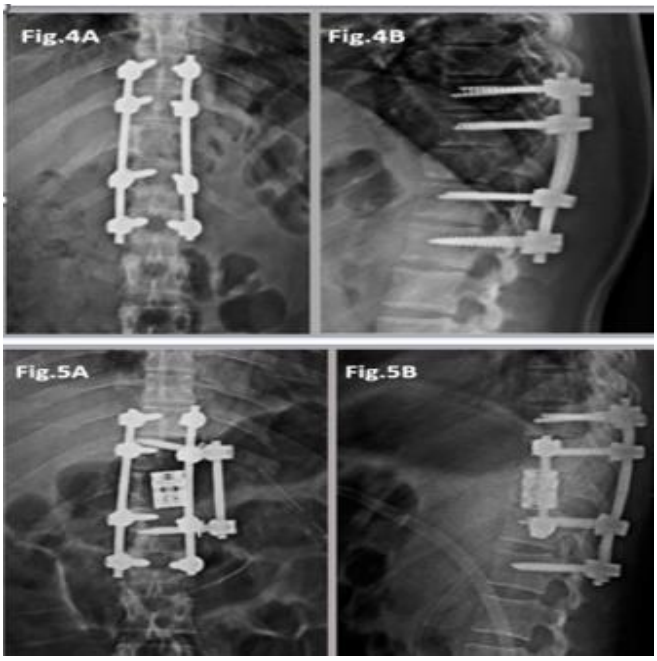

Fig. 4A & 4B: AP and lateral X-rays post separation surgery, intralesional curettage, and fixation.
Fig. 5A & 5B: AP and lateral X-rays post D11 vertebrectomy.

DISCUSSION:-

In 1845, Lebert documented the first microscopic examination revealing a bone tumor with multinucleated giant cells. Since then, significant progress has been achieved in the clinical, radiological, and histological aspects concerning the diagnosis and management of giant cell tumors. Giant cell tumors of the bone are rare, comprising only 10% of primary bone neoplasms. Within this category, 5-10% occur in the spine, with the sacrum being the most common site. The incidence peaks in the third decade, with a slight predisposition towards females. Pain is the primary presenting symptom, often detected after the evolution of neurological deficits. These deficits typically result from spinal instability, vertebral collapse, and extra-compartmental intraspinal tumor spread

affecting the spinal cord or nerve root. Tumor-induced cortical breach is a key factor leading to pathological vertebral collapse.[5]

Surgery, including curettage, intralesional excision, and en bloc resection, is the primary treatment option for bone GCT in clinical practice. If technically feasible to achieve tumor-free margins, en bloc resection should always be prioritized as the primary treatment for GCT of the mobile spine, as it reduces the risk of local recurrence and enhances survival outcomes. In 2009, the Spine Oncology Study Group's review found that total en bloc resection of spinal giant cell tumors (GCT) was technically feasible, but the recommendation relied on limited-quality evidence.[6] Total en bloc resection may not be universally applicable to all spinal giant cell tumors. This limitation is due to the proximity of critical anatomical structures adjacent to the vertebrae or the invasion of the tumor into the pedicle and vertebral accessory, rendering en bloc resection an exceedingly dangerous and technically demanding procedure in these scenarios. In this case, due to the atypical presentation and involvement of the spinal canal, separation surgery was considered which is associated with lower functional morbidity and complications.

Denosumab is a potentially effective treatment for spinal giant cell tumors (SGCT), used as an adjuvant or standalone therapy. The dosing is 120 mg subcutaneously every 28 days, with loading doses on days 8 and 15 in the first month, based on a Phase II principal study.[4] In 86% of patients, tumor response was confirmed with histopathological examination showing elimination of over 90% of giant cells, and there was no radiographical progression of the lesion. While there is no consensus on the duration of denusumab usage, in this case postoperatively denusumab was given for 6 months and stopped. But at 1 year follow up MRI showed local recurrence of GCT which is a major concern after stopping denosumab in case of subtotal resection of tumour. This implies that denosumab by itself does not lead to the complete elimination of the disease. Lau et al. found that denosumab does not cause apoptosis of stromal cells; rather, it simply inhibits their activity. In this case, owing to the large size of SGCT and extension into surrounding soft tissue Denusumab was administered to facilitate shrinkage and peripheral rim formation. Girolami et al. reported a transformation of the neoplastic

stromal cells to a fibrous matrix with a decrease in angiogenesis, which could explain the potential decreased vascularity of the denosumab-treated tumors.[7] In the present case, the recurrent GCT appeared to have a similar response, which aided in complete resection of tumor.

Radiation therapy has been shown to offer a favourable prognosis for GCT and may decrease the recurrence rate following surgery. Dunne C et al. reported that local recurrence was much lower in patients who received radiotherapy than in those who did not. Radiation therapy is advisable in cases of complete piecemeal gross excision with intralesional margins when surgical margins are compromised by substantial tumor extension into the pedicles, strong dural attachment, and/or extensive invasion of paraspinal soft tissue.[8]

COLCLUSION:-

In conclusion, this report emphasizes the unusual emergency presentation of a giant cell tumor in the thoracic spine and describes its recurrence managed through total corpectomy and neoadjuvant denosumab therapy. Total resection is crucial, aided by denosumab to reduce surgical morbidity. Adjuvant denosumab and radiotherapy help minimize postoperative recurrence. It also emphasises on need for further research on the duration of adjuvant therapy in GCT.

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