International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2020): 7.803

Mistaken Sacral Giant Cell Osteosarcoma

Lekkala Naga Sandhya[1], Kishore Kumar Kota[2]

[1]Final Year PG, GSL Medical College & General Hospital, Rajahmundry, Andhra Pradesh, India

[2]DM Medical Oncology

[1]Corresponding Author Email ID: *sandhyalekkala19[at]gmail.com*

Abstract: ***A 29 - year - old female patient presented with complaints of low back pain radiating to bilateral lower limbs with parasthesia's and unable to sit since three months. The pain was insidious on onset, gradually progressive which did not relieve even on medication. The patient was advised X –ray LS spine AP & lateral view shows an ill - defined lytic area with adjacent soft tissue shadow & advised MRI for further evaluation. MRI LS spine was done which showed large expansile lytic lesion in sacrum, coccyx with extensive soft tissue component. To evaluate further we did biopsy & came as giant cell tumour of sacrum. Surgical resection of the tumour was done partly as the complete tumour resection was not possible due to pelvic adhesions. Resected tissue sample sent again for histopathology and reported as giant cell tumour. Patient came for follow up after three months with extensive tumour spread to pelvis & lung metastasis. Biopsy performed again & came as Giant cell rich Osteosarcoma.***

Keywords: Osteosarcoma, Giant cell rich osteosarcoma, Osteoclast, Spindle cell

1. Introduction

Osteosarcoma & Ewing's sarcoma are the most common primary malignant bone tumours in childhood & adolescence, most occurring during first two decades (1, 2). Osteosarcoma rarely involves the spine (1–3% of all osteosarcomas), and the sacrum is one of the most common spinal locations.

Giant cell - rich osteosarcoma (GCRO) is an extremely rare histologic variant, accounting for only 1%–3% of conventional osteosarcoma [3, 4, 5]. The unusual histopathological appearance and the rarity of the lesion poses a great diagnostic challenge. GCRO is an undifferentiated high - grade sarcoma with numerous osteoclast - like giant cells and variable amount of tumor osteoid [6]. The radiological and histopathological differentiation of GCROs from other benign and malignant giant cell tumors (GCTs) is highly challenging [6]. It is important to differentiate them from other aggressive GCTs as the prognosis and treatment differs between them [3]. To the best of our knowledge this is the first case, GCRO to involve the sacrum.

2. Case Report

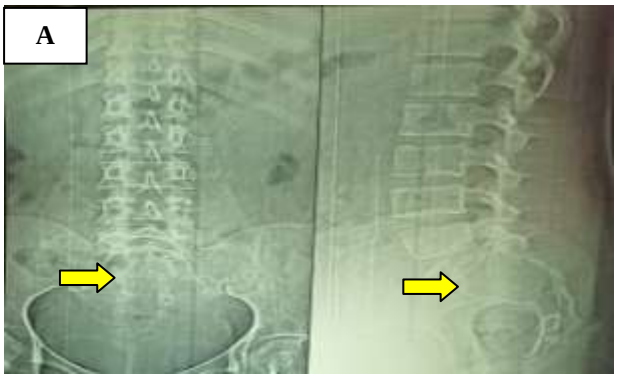
History: A 29 - year - old female patient farmer by occupation presented with chief complaints of low back pain radiating to bilateral lower limbs with parasthesia's and unable to sit since three months. The pain was insidious on onset, gradually progressive which did not relieve even on medication. She had no history of any previous co morbidities or past surgical history. The patient was advised X - ray LS spine, MRI spine & pelvis for further evaluation to rule out neoplasm.

Laboratory findings:
CBP: Normal
LDH=900
ALP: 1793

3. Radiological Investigations

3.1 X - ray LS Spine (AP & Lateral view)

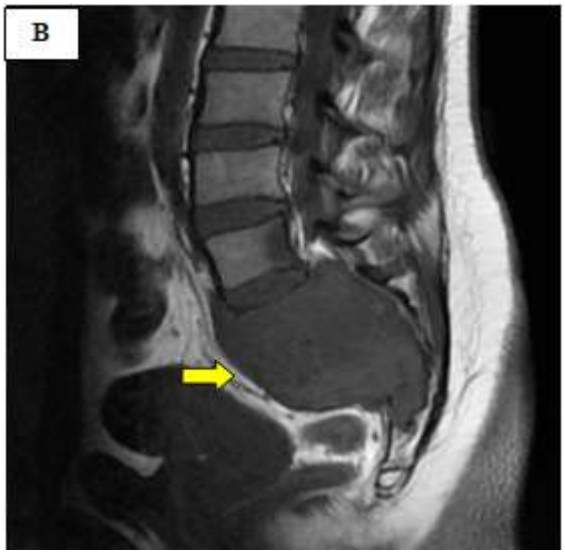
Showed ill defined lytic area in sacrum with adjacent altered soft tissue shadow (fig A).



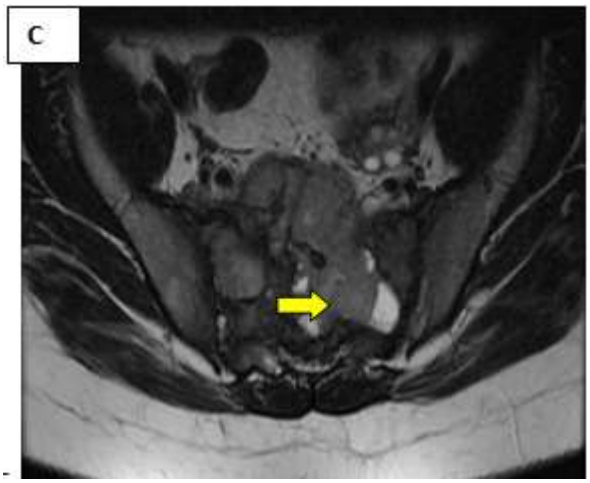
A. Plain radiograph of L - spine AP and lateral view revealed an ill defined lytic area (arrow pointing) in sacrum with surrounding altered soft tissue shadow.

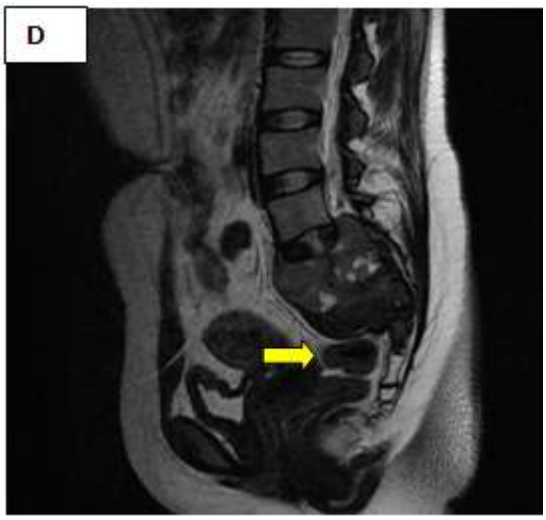
3.2 MRI LS Spine with both hip joints:

Showed large irregular expansile lesion with extensive soft tissue component arising from sacrum, coccyx & bilateral sacral alae. T1 W sagittal image showing large expansile hypointense lesion involving sacrum, coccyx, adjacent L5 vertebra with extensive soft tissue component (fig B). T2W axial and sagittal images showing heterogenous iso - intermediate signal intensity large expansile lesion involving sacrum, coccyx, L5 vertebral body with extensive soft tissue component and few cystic areas, fluid - fluid levels (fig C, D). The lesion extending into pelvis, SI joint and encasing the nerve roots. Possible DDs to be considered for the lesion given as Giant cell tumour, chordoma and advised biopsy from the lesion for further confirmation.



B: T1 W sagittal image showing large expansile hypo intense lytic lesion involving sacrum, coccyx & adjacent L5 vertebra with extensive soft tissue component.



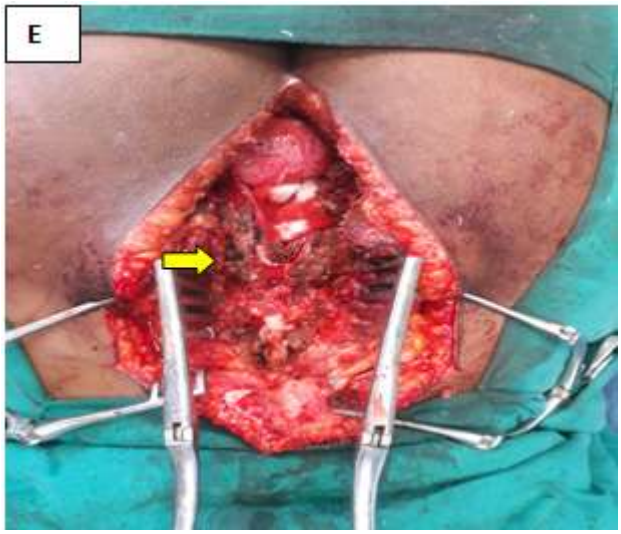


C, D: T2W axial and sagittal images showing heterogenous iso - intermediate signal intensity large expansile lytic lesion involving sacrum, coccyx with extensive soft tissue component and few cystic areas, fluid - fluid levels.

FNAB: Histopathological examination of the biopsy specimen was performed & showed multinucleated osteoclast giant cells, possibility of Giant cell tumour.

3.3 Surgical Resection

High level sacrectomy with partial excision of sacropelvic lesion was done. Nerve encasement relieved. Lateral extent of the tumour is left behind due to adhesions. The tissue sample was sent for histopathological examination. The patient advised follow up after 3 months.



E: Operative findings show highly vascular tumour in sacrum & coccyx

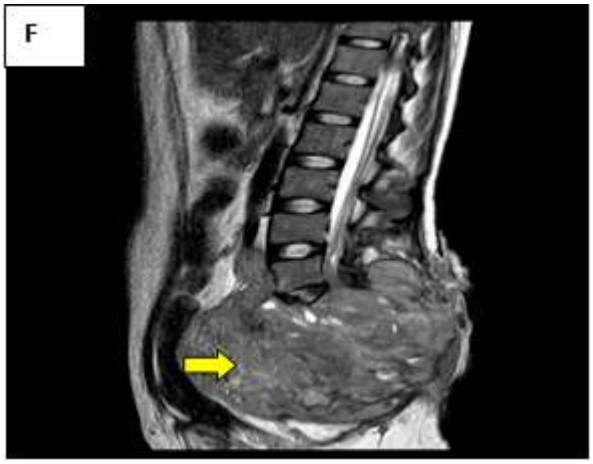
Histopathological examination of the excised specimen was performed showed multinucleated osteoclast giant cells, reported as giant cell tumour.

3.4 Follow Up

The patient visited hospital after 4 months with extensive pain in sacral region, loss of weight, shortness of breath.

CT chest: Shows multiple cannon ball metastasis.

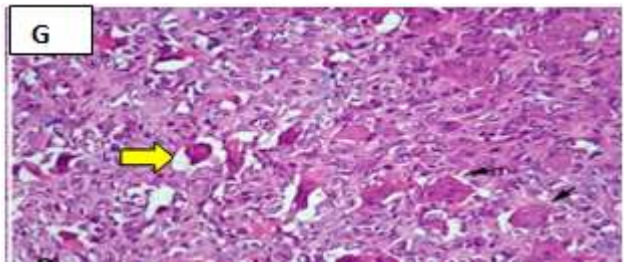
Follow up MRI was done showing large lobulated heterogenous soft tissue signal intensity destructive lesion in sacral region measuring 14 x 8 cm with extraosseous soft tissue component in pelvis & retroperitoneum. Superiorly the lesion infiltrating L5 - S1 IV disc, posteriorly extending into spinal canal, bilateral neural foramina, paraspinal muscles, through sciatic foramina into gluteal region & involving underlying soft tissue, skin (fig F) The lesion reported as recurrence & advised biopsy further.



F: sagittal T2W MRI shows large lobulated heterogenous soft tissue signal intensity destructive lesion in sacral region involving pelvis, spinal canal, neural foramina, adjacent skin & soft tissue.

3.5 Biopsy

Repeat biopsy was done from the sacral lesion showed extensive areas of necrosis, hemorrhage with multinucleated osteoclast like along with spindle tumour cells. Stroma shows myxoid changes, dense inflammatory cell infiltrate made up of neutrophils, lymphocytes. Focal area shows osteoid & new bone formation. Features suggestive of malignant mesenchymal tumour –**Possibly Giant cell rich osteosarcoma** (fig G).



G: HPE shows giant cells & spindle cells.

4. Discussion

Osteosarcoma is the most common, aggressive primary malignant tumor affecting the bone, in which the malignant mesenchymal cells produce tumor osteoid. [3]

Osteosarcoma was histologically classified into central, intramedullary and surface variants with a number of subtypes by the World Health Organization. The central osteosarcoma is further subdivided into the conventional osteosarcoma, telangiectatic osteosarcoma, small - cell osteosarcoma and low - grade osteosarcoma. GCRO is an uncommon histologic variant of conventional osteosarcoma comprising about 1%–3% of primary osteosarcoma. [3, 4]. Bathurst first described this variant in 1986 under the term "osteoclast - rich osteosarcoma. " [3, 5, 6]

Specifically, GCRO may show a mutation of the H3K27 me3 (the trimethylatedlysine residue at position 27 in the protein histone H3) mutation. [8]

Osteosarcoma in the long bones may develop in preexisting conditions such as Pagets disease, fibrous dysplasia, retinoblastoma or in patients having a history of previous irradiation or trauma.

Osteosarcoma in the initial phase manifests as a nondescript swelling, only to become overly aggressive in the later phase of life. [9] This was seen in the present case as well, where a small swelling became significantly large within a short span of time. The average duration of symptoms before it is diagnosed is reported to be 3 - 4 months.

The radiological feature of GCRO is different from that of the conventional osteosarcoma [5, 6]. According to Bathurst, "many of the classic radiological features of an osteosarcoma are absent in cases of GCRO and the radiological appearance involves an ill - defined margin surrounding a predominantly lytic lesion; a soft - tissue mass is usually not present. " [3, 6]

4.1 Treatment

In recent years, multidisciplinary approach and aggressive adjuvant/neoadjuvant chemotherapy have increased the oncologic outcome of patients with osteosarcoma.

Some authors suggested that patients with osteosarcoma of the spine should be treated with a combination of chemotherapy and at least marginal surgery [7]. Postoperative radiotherapy can also be applied in the treatment program and may be of benefit in selected patients.

Chemotherapy: Currently, chemotherapy is undoubtedly the method that is likely to cure the greatest proportion of patients with osteosarcoma. However the association with surgery is essential for the local control and the management program of all patients (10).

Doxorubicin, cisplatin, high - dose methotrexate, ifosfamide, and etoposide have antitumor activity in osteosarcoma and are frequently used with different protocols as the basis of treatment.

Sacrectomy: Patients whose tumors can be completely resected with adequate margins should be approached with curative intent. However, the surgical intervention is quite challenging given the magnitude of treatment, the significant compromise of neurological status, and the high risk of complications. To date, the question whether surgery is really beneficial to the affected patients is still debated.

Radiation therapy: In general, radiotherapy has a limited role in the management of osteosarcoma because of the relative radioresistance and the need for a large dose of radiation to achieve clinical response, but there are anatomical locations in which the possibility of complete surgical resection is unfeasible.

However, the effect of chemotherapy alone is usually temporary, and there is a need of intensive treatment for local control. In these cases, radiation therapy may be an option to try to extend the progression - free interval.

Another possible scenario is the use of adjunctive irradiation in patients who underwent intralesional or inadequate surgery, in which the overall survival was better compared with patients that not received further local treatments.

5. Conclusion

GCRO has no distinct clinical or radiological features that may aid in its recognition. It is a very rare clinical entity which poses a striking resemblance to that of a GCT.

The unusual histological appearance and the exceptional rarity of the lesion poses a great diagnostic challenge; thus, the clinical, radiological and histopathological findings should be integrated for its early diagnosis and proper management.

References

[1] Longhi A, Errani C, De Paolis M, Mercuri M, Bacci G. Primary bone osteosarcoma in the pediatric age: state of the art. Cancer Treat Rev.2006; 32: 423–36. CrossRefPubMed
[2] Unni KK. Dahlin's bone tumors general aspects and data on 11, 087 cases.5th ed. Philadelphia: Lippincott - Raven; 1996.
[3] Fu HH, Zhuang QW, He J, Wang LZ, He Y. Giant cell - rich osteosarcoma or giant cell reparative granuloma of the mandible? *J Craniofac Surg.2*011; 22: 1136–9. [PubMed] [Google Scholar]
[4] Sun LM, Zhang QF, Tang N, Mi XY, Qiu XS. Giant cell rich osteosarcoma of the mandible with abundant spindle cells and osteoclast - like giant cells mimicking malignancy in giant cell tumor. *Int J Clin Exp Pathol.2*015; 8: 9718–22. [PMC free article] [PubMed] [Google Scholar]
[5] Verma RK, Gupta G, Bal A, Yadav J. Primary giant cell rich osteosarcoma of maxilla: An unusual case report. *J Maxillofac Oral Surg.2*011; 10: 159–62. [PMC free article] [PubMed] [Google Scholar]
[6] Shetty SS, Dahake RN, Venkadasalapathi N, Radhakrishnan R. Giant cell rich osteosarcoma of the jaw - a rare entity and review of literature. *J Oral Maxillofac Surg Med Pathol.2*018; 30: 301–5. [Google Scholar]
[7] Vander Griend RA. Osteosarcoma and its variants. *Orthop Clin North Am.1*996; 27 (3): 575–81. [PubMed] [Google Scholar]
[8] Gore MR. Giant cell rich malignancies: A meta - analysis. *Ann Med Health Sci Res.2*018; 8: 225–32. [Google Scholar]
[9] Desai D, Pandith S, Jeergal PA, Arathi K, Saini R. Fibroblastic variant of osteosarcoma: A challenge in diagnosis and management. *Open Dent J.2*010; 4: 211–7. [PMC free article] [PubMed] [Google Scholar]
[10] Barwick KW, Huvos AG, Smith J. Primary osteogenic sarcoma of the vertebral column: a clinicopathologic correlation of ten patients. Cancer.1980; 46: 595–604. PubMed.

Author Profile

Lekkala Naga Sandhya is Final Year PG Scholar, GSL Medical College & General Hospital, Rajahmundry, Andhra Pradesh, India