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A rare case report: Giant cell tumor of the presacral space and it's management

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Abstract

Background: Giant Cell Tumor (GCT) is a rare primary skeletal lesion accounting for approximately 5% of all primary bone tumors in adults. GCT has been described histologically as a benign neoplastic lesion. It clinically presents as painless, firm, well defined mass without any fixity to underling structures. Although it is benign in nature but rarely GCT may exhibit a much more aggressive phenotype, and the role of chemotherapy in metastatic GCT or residual tumor is not well defined in the literatures.

Case presentation: We present a case of 38-year-old female who presented at our setting with rapid growing mass of the presacral space managed by debulking surgery and chemotherapy.

Introduction

GCT of bone is a rare primary skeletal lesion accounting for approximately 5% of all primary bone tumors in adults. GCT has been described histologically as a benign neoplastic lesion [1-3]. The tumor is composed of an admixture of osteoclast-like multinucleated giant cells and mononuclear stromal cells [4]. Although giant cell tumors of soft parts are very uncommon 2 neoplasm, they have been described in numerous anatomic sites including extremities, trunk, superficial and deep fascia, tendon sheath, and skeletal muscle [5] but rarely described in the presacral space. The presacral space is an extra peritoneal potential space between the upper two thirds of the rectum and the sacrum.

The optimal treatment of GCT is by performing a surgical resection of the tumor aiming at achieving surgical free margin. This normally gives a good prognosis and a very low rate of tumor recurrence only if the lesion has been removed adequately [6].

Here, we report a rare case of GCT arising from the presacral space and description of the clinical pathological and radiological features as well as its response to surgery and adjuvant chemotherapy.

Case presentation

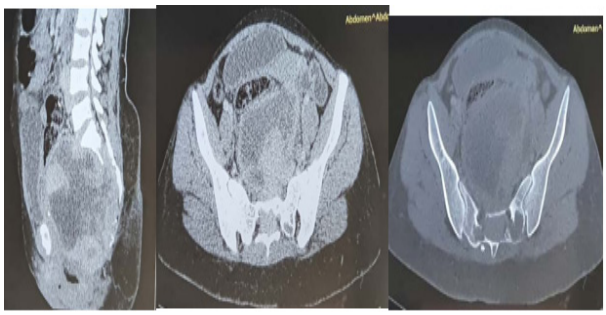
A case of 28-year-old female, Para 2 living 2, presented with pelvic pain, lower back pain, relative constipation and lower

limb neuropathic pain for six months. In first two months she was attended by neurologist for neuropathic pain and later referred to our gynecologic department due to worsening of pelvic pain.

On examination she was clinically stable, with Blood pressure 123/84 mmHg, Pulse rate 89 beats/minute, saturating 98% on room air, and her body temperature 36.7°C. She had normal muscle power and sensation on both lower limbs. Her cardiovascular and respiratory findings were uneventful. On abdominal examination, there was palpable suprapubic mass, mild tender, limited mobility, with soft margins. Pelvic examination, she had normal perineum, huge soft tender mass on posterior lower vaginal wall with menstrual bleeding accumulated in the upper vaginal canal. Rectal examination revealed soft tender mass on left anterolateral to rectal mucosa, suspected ovarian abscess or soft tissue tumor.

Radiology

Chest abdominal pelvic CT scan with contrast: Revealed presacral round heterogeneous round with areas of necrosis and calcifications, causing destruction of S2, S3, S4 sacral vertebrae, coccyx and bilateral adjacent part of iliac bones also compressing rectal sigmoid colon, measured about 14.35 cm x 10.9 cm x 14.35 cm. There was displacement of uterus and urinary bladder anteriorly other abdominal and pelvic structures were normal including lungs. CT scan images of the tumor are revealed below.



Radiology image

Her laboratory investigations were normal including the following, Serum creatinine 35 Umol/L, Serum urea 3.13 Umol/L, sodium 136.43 mmol/L, Potassium 3.84 mmol/L, CA 125 - 17.3 U/ml, Hemoglobin 13 g/dl and platelet count of 219 x 10^3/mm^3 other parameters of complete blood count were normal.

Patient agreed for debulking surgery after consultation with provisional diagnosis of presacral tumor with differentials of soft tissue sarcoma, osteosarcoma and chordoma due to CT scan findings. Intraoperatively, there was presacral tumor about 10 x 7 cm in size pushing rectum, uterus, and bladder superiorly and anteriorly, normal uterus, ovaries, fallopian tubes, urinary bladder, rectum and bowel. Tumor debulking was achieved by 90% through opening of left pararectal space. Excessive bleeding encountered which necessitated gauze packing and re- 4 laparotomy on the following day for pack removal and hemorrhage was controlled. Her total estimated blood loss was 1500 mls and she received two units of blood transfusion each consists of 450 mls.

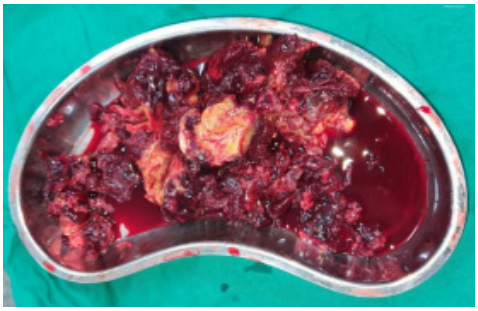


Image 1: Shows part of the tumor.

She had excellent recovery and discharged on day four post relaparotomy with relieved pressure symptoms. Biopsy result revealed Giant Cell Tumor (GCT) with extensive necrosis.

Later patient was discussed during tumor board meeting after biopsy results and agreed on adjuvant chemotherapy for remaining 10% residual tumor. She is currently on seventh months of Denosumab 120 mg monthly with no evidence of disease progression or resumption of pressure symptoms.

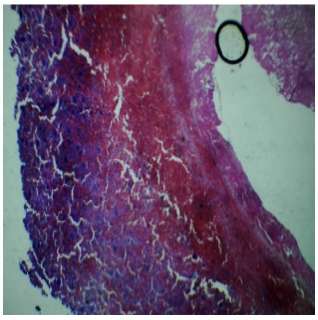


Figure 1A: H&E stained tissue showing extensive necrosis, hemorrhage and multiple osteoclastic giant cells, no evidence of osteoid formation x40.

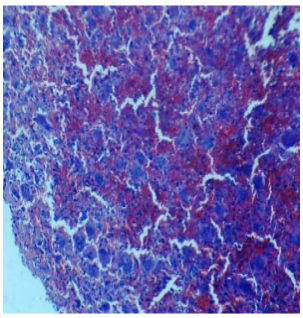


Figure 1B: H&E stained tissue showing numerous osteoclastic giant cells and hemorrhage at x100.

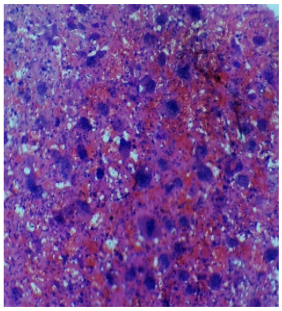


Figure 1C: H&E stained tissue showing numerous osteoblastic giant cells and hemorrhage at x200.

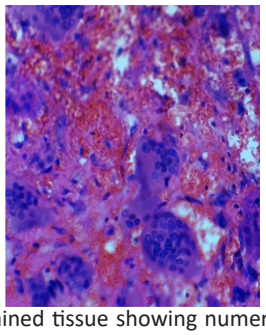


Figure 1D: H&E stained tissue showing numerous osteoclastic giant cells at x400.

Discussion

Presacral tumors are a rare group of heterogeneous lesions located in a potential space referred to as the retro rectal or presacral space [7]. Giant-Cell Tumor (GCT) though it is very rare tumor it is the second most common sacral tumor (20%) after chordoma (40%). GCT has a female predilection (with a female to male ratio of 2:1) and generally presents between 15 and 50 years of age [8,9]. GCTS are generally considered histologically benign, however they may exhibit locally aggressive behavior like in this case and high rate of local recurrence (20-50%). It has 1- 5% rate of lung metastasis and may convert to fulminate malignant variant, which has poor prognosis [10], luckily this patient had only localized disease.

This is a rare case of presacral tumor presented with neuropathic pain and lower back pain, initially treated by neurologist for two months without response due to its vague presentation. Due its unspecific presentation and its rarity the possibility of delayed diagnosis is very high. Through literature review, the occurring symptoms are directly related to compression or infiltration of anatomical structures of the pelvis minor by the tumor and may manifest as lower back pain, pelvic pain or rectal pain or resemble various neurological defects [11]. However, often presacral tumors are asymptomatic; completely asymptomatic lesions occur in 26-50% of patients. Lack of characteristic symptomatology and difficult anatomical localization make the diagnostic process difficult and often delays the ultimate diagnosis [7]. Her severe pain in lower limb, lower back and adnominal were relieved after surgery confirms pain was caused by tumor pressure effects and not otherwise. Presence of intact muscle power and sensation in lower limbs also affirms nerve compression and not nerve destruction.

Unspecific clinical presentation of our patient includes vague abdominal and bowel symptoms, plus unclear abdominal pelvic examination findings lead to inconclusive presumptive diagnosis until CT scan proved to the contrary. Medical imaging plays a very important role in obtaining the comprehensive information of the tumor, including the location, size, internal condition, growth pattern, and the relationship with adjacent tissues, to achieve the diagnosis [10]. Our CT scan of chest, abdominal pelvic with contrast provided a clear tumor details, location and it's origin being in presacral area. Other added details include; relationship with adjacent structures, involvement of S4 sacral vertebrae, coccyx, bilateral adjacent part of iliac bones and compression of rectal sigmoid colon which corresponds with intraoperatively findings.

Immunohistochemistry can give added information regarding the tumor with positive stain for CD68, vimentin, tartrate resistant acid phosphates, cytokeratin, smooth muscle actin [2,12]. Unfortunately, these investigations could not be performed in our setting.

The treatment of GCT remains controversial because of the small number of reported cases. Radical surgery has been widely accepted as a mode of treatment for GCT. However, because of the possibility of local recurrence, clinical follow up with or without postoperative radiotherapy is advised after excision. Several reported cases advocate adjuvant radiotherapy as a treatment of GCT postoperatively, but it has not proven to be standard management for presacral tumors [4,7]. There are also few reported cases with excellent recovery with neoadjuvant chemotherapy [13]. Currently, there is no report in the literature about the choice of adjuvant chemotherapy and whether adjuvant chemotherapy can improve the outcome of patients with GCT of the presacral space but there are few reported cases of adjuvant chemotherapy with good recovery in other sites, and in most of the case reports. Few literature reviews have shown that patients responded well to Denosumab after failure of adjuvant radiotherapy, therefore Denosumab has demonstrated anti GCT-efficacy and has the potential to spare a young, vulnerable population from adverse long term effects of traditional adjuvant radiation therapy [14,15]. For 10% residual disease we preferred to give adjuvant chemotherapy (Denosumab) instead of radiation therapy because she was young and still in child bearing age.

Follow up of patient with GCT post treatment was found that local recurrence rate of 12% in the duration of 34 to 45 months with a very rare metastasis and death. Currently, there is no reliable clinical and pathologic risk factors that may predict recurrence and metastatic [4]. In one cohort study it was shown that, soft tissue extension and higher tumor grades are significantly associated 8 with development of recurrence in cases of bone GCT [16]. Other rare recurrences were shown after two months' post primary surgical treatment, and other types of chemotherapy [13]. We followed our patient post-surgery and adjuvant chemotherapy for seven months without any sign of metastatic disease or progression we are planning to performing next CT scan after one year of follow up.

Conclusion

We presented rare case report of local aggressive GCT of the presacral space, diagnosis challenges, surgical management and adjuvant treatment with Denosumab. Although the role of chemotherapy in GCT is not well-defined in literatures. There are current case reports of GCT in other sites responded well with Denosumab, for this reason we hope our patient is also benefiting from this regimen due to absence of signs of progression of disease in 7 months.

Declarations

Patient perspective: The care provided was timely with full explanation of the diagnosis and follow up plan.

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Ethic approval, consent to participate and publish: Written informed consent was obtained from the patient for publication of this case report and related images. A copy of written consent is available for review of Editor-in –Chief of this journal. Additional consent was sought and ethical clearance was granted by the joint Catholic University of Health and Allied Sciences/ Bugando Medical Center Research and Ethical review.

Competing interests: Authors declare that they have no competing interests.

References

1. Inwards CY, Rose PS, Sim FH. Giant Cell Tumor of Bone Risk Factors for Recurrence. Clin Res. 2011; 469: 591–599.
2. Sen D, Jain A, Majumdar B, Sen S, Mishra P, Behra A, et al. Case Report Soft tissue giant cell tumour revisited – A case report. J Pakistan Assoc Dermatologists. 2015; 25: 332–334.
3. Rateb K, Ghozlen B, Leila A, Faten F, Med D, et al. Surgery Case Reports Giant cell tumor of soft tissues : Int J Surg Case Rep. 2017; 31: 245–249.
4. Sunumu O, Report C, Düşük B, Potansiyelli M, Hücreli D, Sunumu O, et al. Soft Tissue Giant Cell Tumor of Low Malignant Potential of the Neck : A Case Report and Review of the Literature. Olgu Sunumu/case Rep. 2013; 30: 73–77.
5. Hu Q, Peng J, Xia L. Recurrent primary mediastinal giant cell tumor of soft tissue with radiological findings : A rare case report and literature review. 1world J Surg Oncol. 2017; 15: 1–6.
6. Chand CK, Col L, Bhardwaj RK, Rappai MTJ. Study of 7 Cases of Giant Cell Tumor of Soft Tissue. Med J Armed Forces India [Internet]. 2009; 62: 138–140. Available from: http://dx.doi.org/10.1016/S0377-1237(06)80056-0
7. Dziki Ł, Włodarczyk M, Sobolewska Włodarczyk A, Saliński A, Salińska M, Tchórzewski M, et al. Clinical research Presacral tumors : Diagnosis and treatment – A challenge for a surgeon. Clin Res. 2019; 15: 722–729.
8. Nishant Patel, MD, MBA, Katherine E Maturen, MD, MS, Ravi K Kaza, MD, Girish Gandikota, MD, Mahmoud M Al-Hawary, MD and Ashish P Wasnik, MD Article R. 10 Imaging of presacral masses —A multidisciplinary approach. BJR. 2016; 89.
9. Wang J, Li D, Yang R, Tang X, Yan T, et al. Epidemiological characteristics of 1385 primary sacral tumors in one institution in. WORLD J Surg Oncol. 2020; 18: 2017: 1–12.
10. Services S, City SL. Giant cell tumor of the sacrum. Neurosurg Focus. 2003; 15: 13–16.
11. Ambrosio LD, Boccone P, Pellegrino P, Miano S, Bertotto I, Manca A, et al. Case 1 – Multidisciplinary treatment of a giant cell tumor of the bone affecting the sacrum of a young woman. cASE REPORTMEET Expert. 2016; 4: 36–41.
12. Franco V, Florena AM, Florena EO and GB. Giant cell tumor of the ovary. Immunohistochemical evidence of origin from stromal ovarian cells. Histo Histopathol. 1995; 10: 55–60.
13. Skubitz KM, Manivel JC. BMC Cancer Giant cell tumor of the uterus : Case report and response to chemotherapy. BMC Cancer. 2007; 7: 1–7.
14. Bardakhchyan S, Kager L, Danielyan S, Avagyan A, Karamyan N, et al. Denosumab treatment for progressive skull base giant cell tumor of bone in a 14 year old female – a case report and literature review. Ital Jouranal Pediatr. 2017; (43:32): 1–7.
15. Alani B, Alaqeeli A, Aldoori M, Farag I. Oral and Maxillofacial Surgery Cases Giant cell tumour of the infra temporal fossa treated with denosumab in an elderly patient : A case report. Oral Maxillofac Surg Cases. 2022; 8: 100247.
16. Hasan O, Ali M, Mustafa M, Ali A, Umer M. Treatment and recurrence of giant cell tumors of bone – A retrospective cohort from a developing country. Ann Med Surg. 2019; 48: 29–34.