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Case report

A rare sacral localization of giant cell tumor in a young adult female: a case report

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Abstract. We reported the case of a 42-years-old woman who suffered from a giant cell tumor of sacrum. Although the giant cell tumor primarily affects the long bones, especially those of the knee joint, it can rarely affect the axial skeleton and the sacrum. The onset of symptoms is generally insidious and may include locoregional pain and swelling as well as movement deficits if nerve roots are involved at this level. In this case report we discuss on the radiographic imaging, computed tomography and magnetic resonance imaging features of this type of tumor in an unusual location of the disease. (www.actabiomedica.it)

Key words: Giant cell tumor, musculoskeletal imaging, spine, computed tomography, magnetic resonance imaging

Introduction

Giant cell tumor (GCT) of bone, first described by Cooper and Travers in 1818 (1), is considered a locally aggressive, intermediate grade benign bone neoplasm (2).

In a small percentage of cases GCT undergoes malignant degeneration or may cause distant metastasis, mainly in the lungs (3).

The tumor is characterized histologically by multinucleated giant cells with a background of mononuclear stromal cells. The multinucleated giant cells appear similar to osteoclasts, which led to the older term "*osteoclastoma*" (4). GCT accounts for 5% of all primary bone tumors and 20% of benign skeletal tumors, and shows an increased prevalence among females, with 80% of cases occurring between 20 and 50 years of age (3).

It mainly affects long bones; the distal femur and proximal tibia represent common locations for occurrence of GCT (5). The axial skeleton is a secondary site of involvement; GCT of the spine and sacrum is rare and it is reported in less than 3% of cases (6).

Case Presentation

A 42-year-old woman was referred to our institution for lower back pain, which slowly arose months earlier, radiating poster-laterally from the lumbar spine into the bilateral thighs. The physical examination revealed moderate enlargement of the sacral region. Moreover, on palpation, an immovable and painful mass could be felt. Plain X-rays showed an image of osteolysis with relatively well-defined margins (Figure 1). Afterwards, more complementary

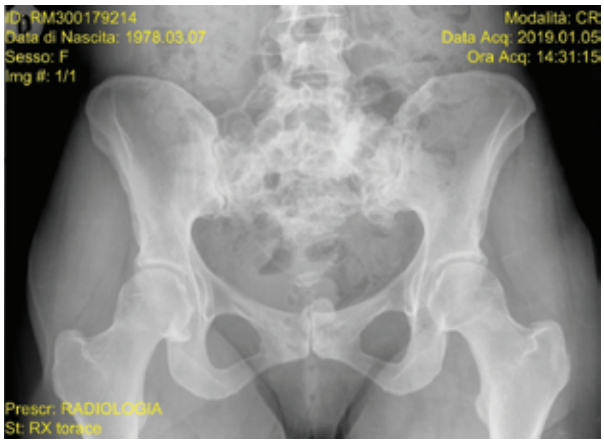

Figure 1. Plain Radiograph of the hip that demonstrate a lytic lesion of the sacrum.

investigations were recommended: total body computed tomography (CT) and contrast-enhanced magnetic resonance imaging (MRI) of the pelvis. The pelvic CT examination revealed an huge isodense soft-tissue mass that involved almost the entire sacrum up to the two sacro-iliac joints, with destruction of the cortical bone (Figure 2 A,B). The pulmonary CT did not reveal any suspected secondary lesions. MRI examination showed the sacral lesion with iso-intensity signal on T1 weighted images and inhomogeneous low signal intensity on T2-weighted images, with intense contrast enhancement in sequences acquired after gadolinium administration (Figure 3-4). Following these investigations, the incisional biopsy was performed, and tumoral tissue fragments were sent to the histopathological examination.

The final histopathological diagnosis was GCT.

The first-line therapeutic choice was surgical curettage with associated cryoablation with liquid nitrogen, a technique that consists in introducing liquid nitrogen into the tumor cavity producing osteonecrosis of the tumor bed. This technique reduces the risk of recurrence even if it can increase the risk of bone fractures. Furthermore, cryotherapy can be repeated to increase its effectiveness (7). After about 6 months from the treatment, the patient was re-evaluated by CT examination which showed a disease stability, without further volumetric expansion of the tumor lesion of the sacrum which remained almost identical to the previous evaluation. (Fig.5 A,B)

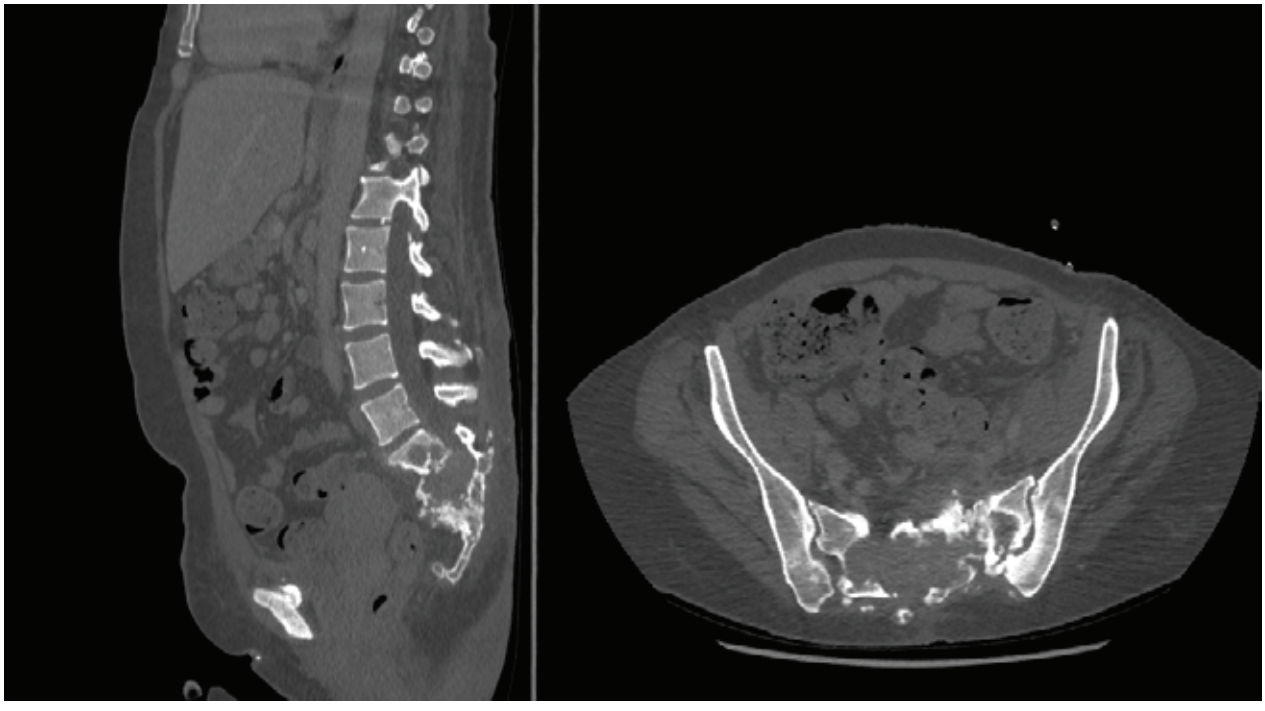

Figure 2. (A, B) The CT scan images show a large defect involving virtually the entire sacrum with a large soft tissue mass with destruction of cortical bone.

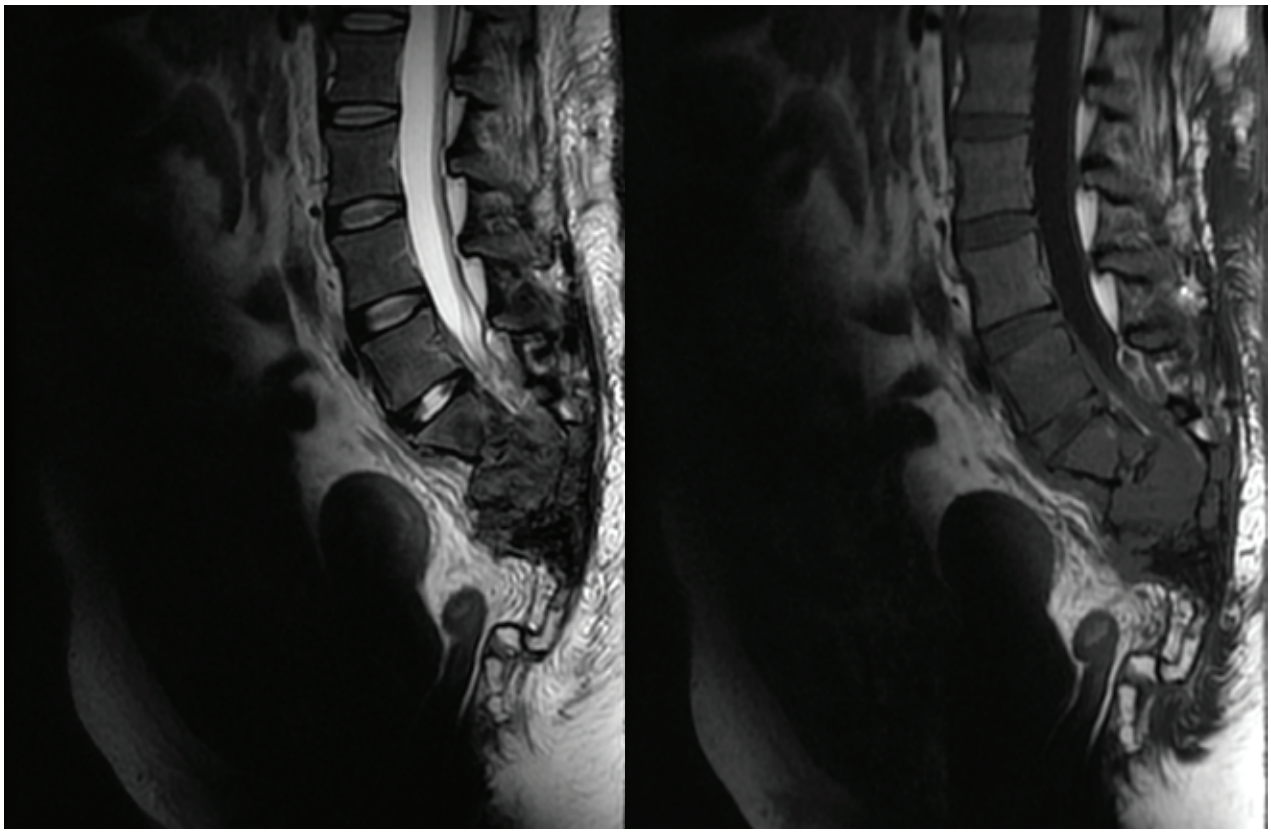

Figure 3. A) Sagittal T2-weighted fast spin-echo MR image shows sacral involvement of mass with anterior and posterior soft-tissue extension and heterogeneous signal intensity. B) Sagittal T1-weighted MR image shows the tumoral mass with homogeneous low signal intensity.

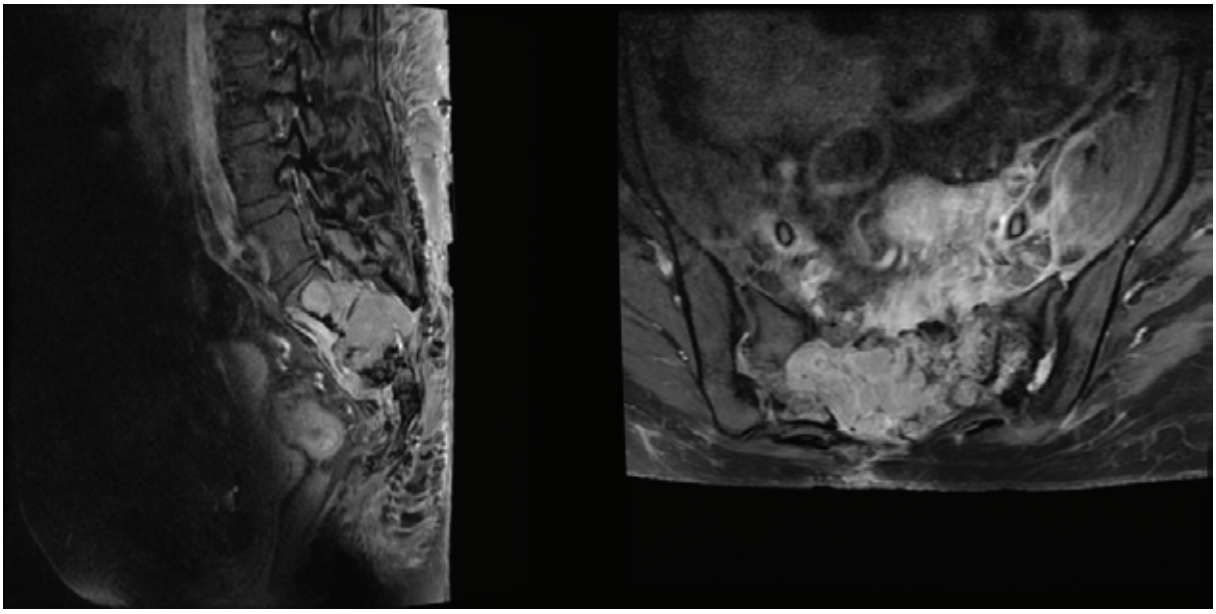

Figure 4. (A, B) Enhanced sagittal T1-weighted MR image shows marked and heterogeneous enhancement of the GCT.

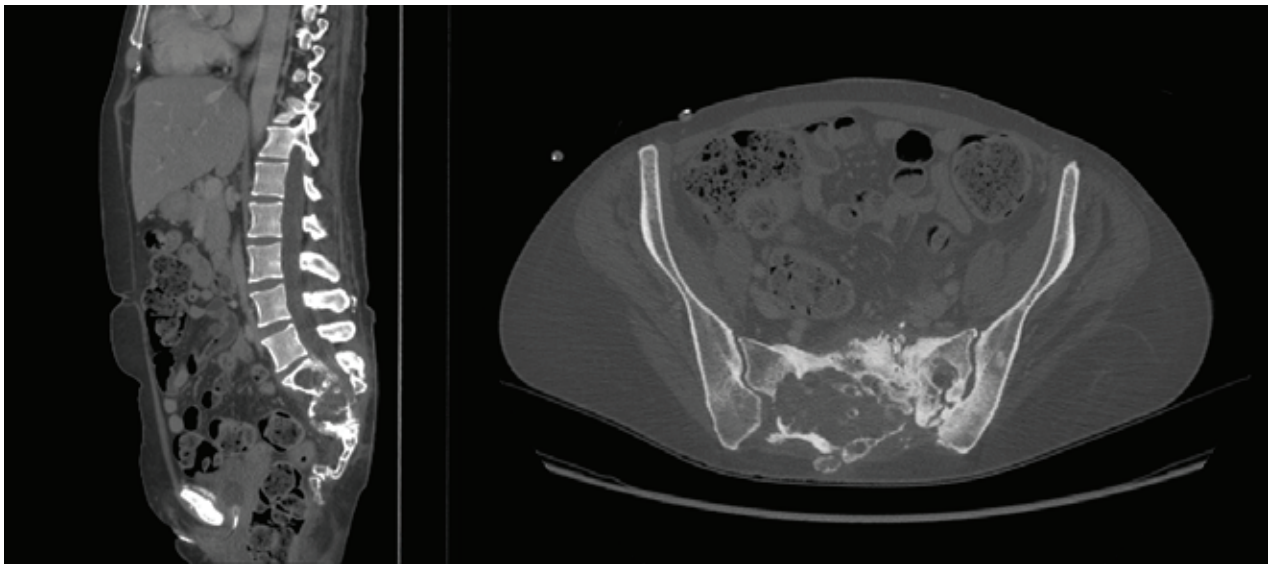

Figure 5. (A, B) The CT images acquired about 6 months after the cryoablation treatment show a picture of disease stability; in fact it is evident that the tumor mass has remained substantially unchanged in size and tomodensitometric characteristics since the last CT evaluation.

Discussion

Sacral GCTs tend to be clinically silent during their initial stages of development and cause few symptoms until they achieve an extremely large size.

Although histologically benign, these osteolytic expansive tumors are locally aggressive, and the local recurrence rate in the sacrum is higher than recurrence rates at other skeletal locations (8).

Plain X-rays, computed tomography and magnetic resonance imaging showed osteolytic destruction of the sacrococcygeal bones and a huge soft-tissue mass; in particular, in conventional radiographs GCT appears as a lytic lesion well-defined and with non-sclerotic margins, with extension to the subchondral bone. (9-11)

CT and MRI are also necessary for a complete assessment of tumor extension and its invasion into surrounding structures.

The magnetic resonance (MR) imaging findings are nonspecific and they usually consist of intermediate or decreased signal intensity on T1-weighted images, inhomogeneous signal intensity on T2-weighted images, and intense enhancement after intravenous administration of gadolinium contrast material.

There is no agreement regarding the therapeutic management of GCTs of rare localizations. In fact, these tumors are usually asymptomatic in the early stages and can become very large and vascular and can grow in the proximity of the nerve roots. Therefore, the chosen treatment should aim to preserve the nerve function and to cause minimal disability and complications to the patient without increasing the risk of tumor recurrence (12).

Although surgery (including curettage and excision) remains the first-line treatment for sacral tumors, their difficult location and huge size, as well as the possibility of life-threatening intraoperative bleeding, make surgery difficult (13).

Recently, the new chemotherapeutic drug denosumab has been used to treat GCT of bone,

a monoclonal antibody that targets the receptor activator of nuclear factor k-B *(RANK)* ligand *(RANK-L)* and stops the osteoclastic activity of cells in GCT of bone (3).

Finally, even though histologically benign, these osteolytic expansive tumors are locally aggressive, and the local recurrence rate in the sacrum is higher than recurrence rates at other skeletal locations (8).

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article

References

1. Cooper AS, Travers B. Surgical essays. London, England: Cox Longman & Co, 1818; 178–179.
2. Roitman PD, Jauk F, Farfalli GL, Albergo JI, Aponte-Tinao LA. Denosumab-treated giant cell tumor of bone. Its histologic spectrum and potential diagnostic pitfalls. Hum Pathol, 2017, 63:89–97.
3. Chakarun CJ, Forrester DM, Gottsegen CJ, Patel DB, White EA, Matcuk GR Jr. Giant cell tumor of bone: review, mimics, and new developments in treatment. Radiographics, 2013, 33(1): 197–211.
4. Balke M, Schremper L, Gebert C, et al. Giant cell tumor of bone: treatment and outcome of 214 cases. J Cancer Res Clin Oncol 2008;134(9):969–978.
5. Montgomery C, Couch C, Emory CL, Nicholas R. Giant Cell Tumor of Bone: Review of Current Literature, Evaluation, and Treatment Options. J Knee Surg. 2019;32(4): 331-336.
6. Kwon JW, Chung HW, Cho EY, et al. MRI findings of giant cell tumors of the spine. AJR Am J Roent- genol 2007;189(1):246–250.
7. Malawer MM, Bickels J, Meller I, Buch RG, Henshaw RM, Kollender Y: Cryosurgery in the treatment of giant cell tumor: A long-term followup study. Clin Orthop Relat Res 1999;(359):176-188.
8. Qin LF, Peng D, Qin LH, Xu M, Fang H, Zhang Q. Huge giant cell tumor of the sacrum: A case report. Oncol Lett. 2014;7(3):894-896. doi:10.3892/ol.2014.1812
9. Murphey MD, Nomikos GC, Flemming DJ, Gan- non FH, Temple HT, Kransdorf MJ. Imaging of giant cell tumor and giant cell reparative granuloma of bone: radiologic-pathologic correlation. Radio- Graphics 2001;21(5):1283–1309.
10. 5. Turcotte RE. Giant cell tumor of bone. Orthop Clin North Am 2006;37(1):35–51.
11. 6. Arnold RT, van Holsbeeck MT, Mayer TG, Mott MP, Koch SR. Necrotic giant cell tumor of bone manifesting with pathologic fracture. RadioGraphics 2011;31(1):93–98.
12. Ji T, Yang Y, Wang Y, Sun K, Guo W. Combining of serial embolization and denosumab for large sacropelvic giant cell tumor: case report of 3 cases. Medicine (Baltimore), 2017, 96(33):e7799.
13. Wuisman P, Lieshout O, Sugihara S and van Dijk M: Total sacrectomy and reconstruction: oncologic and functional outcome. Clin Orthop Relat Res 381: 192-203, 2000.
14. Poll A, Nimigean VR, Vîrlan MJR, et al. A rare tumor in a rare location: giant cell tumor of the sacrum and ilium - case report and current perspectives. Rom J Morphol Embryol. 2019;60(3):985-992.
15. Martin C, McCarthy EF. Giant cell tumor of the sacrum and spine: series of 23 cases and a review of the literature. Iowa Orthop J. 2010;30:69-75.
16. Randall RL. Giant cell tumor of the sacrum. Neurosurg Focus. 2003;15(2):E13. Published 2003 Aug 15. doi:10.3171/foc.2003.15.2.13
17. Raskin KA, Schwab JH, Mankin HJ, Springfield DS, Hornicek FJ. Giant cell tumor of bone. J Am Acad Orthop Surg. 2013;21(2):118-126. doi:10.5435/JAAOS-21-02-118
18. Mavrogenis AF, Igoumenou VG, Megaloikonomos PD, Panagopoulos GN, Papagelopoulos PJ, Soucacos PN. Giant cell tumor of bone revisited. SICOT J. 2017;3:54. doi:10.1051/sicotj/2017041
19. van der Heijden L, Dijkstra PDS, Blay JY, Gelderblom H. Giant cell tumour of bone in the denosumab era. Eur J Cancer. 2017;77:75-83. doi:10.1016/j.ejca.2017.02.021

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