Case Report

Unusual and aggressive course of giant cell-rich osteosarcoma of femur: A case report

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ABSTRACT

Primary tumors of bones are rare and composed of around 0.2% of the total load of tumors in humans. Among primary tumors, osteosarcoma is the most common primary non-hematopoietic malignancy. Giant cell-rich osteosarcoma (GCRO) is a rarest histologic variant and comprised of only 1–3% of the total burden of conventional osteosarcoma. Here, we present the case of a 16-year-old female patient with complaints of pain in the left distal thigh for 3 weeks and swelling for 1 week. The X-ray showed the lytic lesions, with a wide zone of transition over the meta-diaphyseal distal femur with a pathological fracture in the supracondylar area and Codman's triangle. Further investigation revealed the diagnosis of GCRO with the proximal tibia and lung metastasis. The prognosis was explained and radical amputation was planned in the form of above-knee amputation. However, in the post-operative period and before the commencement of the adjuvant therapy, the patient became dyspneic and her Glasgow coma scale started deteriorated and unfortunately, she could not be revived.

Key words: Femur, Giant cell, Osteosarcoma

Osteoblastic, chondroblastic, and fibroblastic variants are usual sub-types of osteosarcoma and characterized by their cellular atypia and extracellular matrix secreted by the tumor cells [1]. On the contrary, giant cell-rich osteosarcoma (GCRO) is a rarest histologic variant and comprised of only 1-3% of the total burden of conventional osteosarcoma [2]. There is no pathognomonic clinical or radiological feature of GCRO, which may aid in its identification. It presents as an exceptionally rare clinical entity that disguises to giant cell tumor. A meta-analysis of Gore found that theses malignancies tend to be aggressive and 33% of cases had distant metastasis and 5-year survival rate is <50% [3].

In a few literatures, the survival rate of GCRO has been reported only for months in some cases [4]. In our case, the survival of the patient was even <1 month. Here, we describe a case report of the patient with a history of pain and swelling in the thigh for a few weeks. Our experience with its rare and aggressive course necessitates bringing it to the horizon of knowledge.

CASE REPORT

A 16-year-old female patient visited the hospital in the out-patient department with chief complaints of pain in the left distal thigh for 3 weeks, swelling for 1 week, and now the patient is unable to use her left lower limb for 2 days. History revealed that initially, the pain was continuous, dull-aching, and mild, but now, it is moderate to severe in nature and even not relieving by taking painkillers. Initially, the swelling was noticed over the inner side of the distal thigh, but now, it has progressed and occupied the whole circumference of the distal part of the thigh.

On general examination, vitals were normal. The patient had an anxious look with pallor and flexed attitude of the left lower limb. There was a gross diffuse bony swelling (15 cm × 20 cm) around the distal thigh with an indistinct margin. The skin over the swelling was tensed, shiny with bluish discoloration of the vein (Fig. 1). It was warm and tender on palpation.

The X-ray of the left knee with thigh and leg showed the lytic lesions (usually the osteosarcoma is a blastic type), with a wide zone of transition over the meta-diaphyseal distal femur with a fracture in the supracondylar area and Codman's triangle (Fig. 2). There was also soft tissue impression, mainly over the posteromedial part of the thigh, but there were no periosteal reaction and sunburst appearance. The clinico-radiological features were favoring for malignant lesion, so the differential diagnoses of osteosarcoma and Ewing' sarcoma was conceded.

Magnetic resonance imaging (MRI) of the left thigh and high-resolution computed tomography (HRCT) of the chest scan was done and the biopsy was planned. The MRI showed the T1

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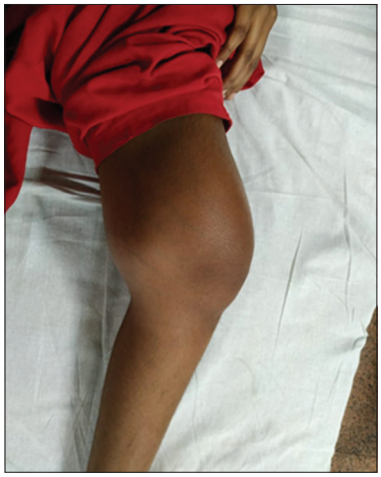

Figure 1: Diffuse circumferential swelling (started from the posteromedial area) over the left distal thigh, with tense, and shiny skin

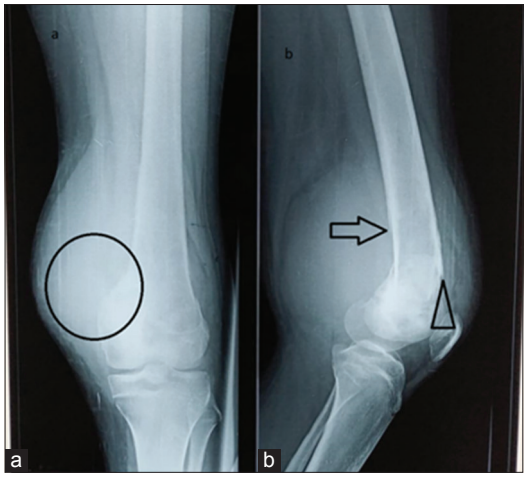

Figure 2: X-ray of (a) anteroposterior view showing soft tissue haziness (in circle) with lytic lesion arising from the medial aspect of metaphyseal area, lateral (b) view shows the mass arising from posterior aspect showing Codman' triangle (arrow) and fracture in the supracondylar part

hypointense and T2 hyperintense mass in the meta-diaphyseal area with cortical breaching and involving predominantly the posteromedial muscle plane and also involved the knee joint. There was also T1 hypointense and T2 hyperintensity changes in the proximal epi-metaphyseal part of the tibia suggested the metastasis (Fig. 3). HRCT revealed the bilateral multiple variable-sized soft tissue nodules involving the lung, and a few of them were pleural based suggestive of cannonball metastasis (Fig. 4). Histopathology showed the tumor cells (pleomorphic, bizarre nuclei, and abundant abnormal mitosis) were interspersed with a large number of osteoclastic giant cells with areas of necrosis, spindle cell and at a few places, laying down the lace-like pinkish osteoid appearance suggested the diagnosis of GCRO (Fig. 5).

Thus, now it was a case with GCRO of the left distal femur with metastasis to ipsilateral proximal tibial and bilateral lung for 3 weeks of duration. In our center, we have not come across the aggressive presentation of any appendicular osteosarcoma ever. Followed by proper prognosis, above-knee amputation was

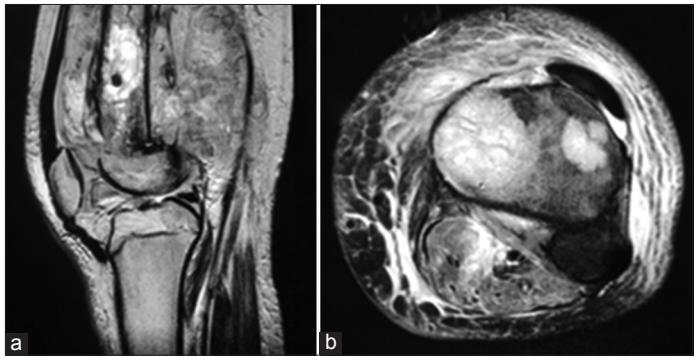

Figure 3: The magnetic resonance imaging showing the (a – T2 sag) hyperintense signal from mass arising from medullary cavity of metadiaphyseal area of the femur with anterior cortical breaching with permeation to suprapatellar area and (b) in PD A x F sat showed the intensity changes in proximal tibia which correlates to metastasis

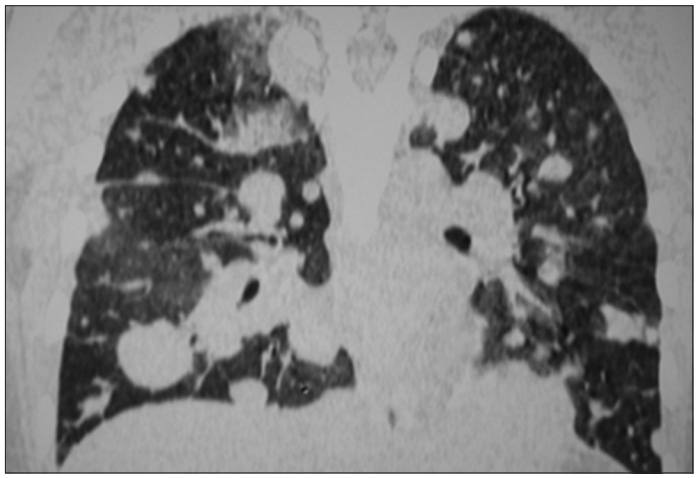

Figure 4: Image showing bilateral multiple soft tissue (variable-sized) nodule, giving a cannonball appearance

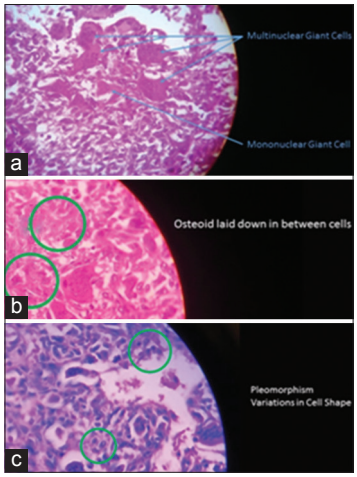

Figure 5: (a-c) Histopathology showing (H and E stained) osteoid laid down (upper slide, green circle) in between the cell, middle slide showing (green circle) pleomorphic variation in cell size of osteoclast, and in lowermost slide indicated cells are multinuclear giant cell confirming the giant cell-rich osteosarcoma

done. The proximal part of the thigh was decided for the level of amputation. Fish mouth incision was given, and superficial fascia and subcutaneous tissue were cut to the vertical to the skin edge. Deep dissection, neurovascular, and osseous dissection were done

in the desired fashion and after myodesis, wound closure was done under vacuum drain. However, in the post-operative period and before the commencement of the adjuvant therapy, the patient became dyspneic and her Glasgow coma scale (GCS) started deteriorating (GCS 15–GCS 5) and unfortunately, she could not be revived.

DISCUSSION

The first description of GCRO was given by Bathurst and Sanekin by the name of "osteoclast-rich osteosarcoma" [5]. The GCRO is comprised of an abnormally abundant number of giant cells, which almost swamps the sarcoma cells [6]. The general consensus of various literature shows that the predilection of GCRO is at 15–20 years and for the knee (metaphysis) as like its conventional form [7]. However, it is the uncustomary histology (more or less uniformly distributed giant cells, scanty osteoid, atypical spindloid cells, numerous and atypical mitotic activity, and necrosis) and radiology exhibited by the GCRO, which poses a marked diagnostic challenge due to close differential diagnosis like malignant giant cell tumor [8]. Some literature has suggested the female predominance of GRCO [9] which was in accordance with the present case report.

The fundamental structure of nucleosome present in a chromosome is histones protein. Mutation in the histone H3.3-protein, which codes for the H3F3A gene, is the key factor to cause the pathogenesis of giant cell tumors. Particularly, the mutation of the H3K27me3 (the trimethylated lysine residue at position 27 in the protein histone H3) attributes to the GCRO [3].

The treatment protocol of GCRO is in the same line as that of conventional osteosarcoma [6]. It is comprised of aggressive surgical resection, then adjuvant radiotherapy, and chemotherapy. The 5-year survival rate is up to 80%. Radiotherapy is reserved only for cases with positive surgical margins and before the chemotherapy. Our case was also planned in the same treatment protocol, but due to a highly aggressive nature, we could not even complete the due course of treatment.

Since the GCRO has conceded as high-grade conventional osteosarcoma as well, this variant also has the therapeutic and prognostic significance also, so this case seemed obvious to report it [10]. Although, the role of the presence of osteoclast-like giant cells is still not clear in the extent of prognosis. Hence, this case report should be seen in the context of that.

CONCLUSION

In this case, there was a predominance of abundant spindle cells and abundant osteoclast-like giant cells with few places of osteoid formation. Hence, it is an unusual histological variant that may pose a great diagnostic challenge. It has a short course of the presentation, but at the same time, the proximal tibial and lung metastasis makes it a rare and unusual and aggressive course of GCRO in comparison to conventional osteosarcoma. Hence, we want to emphasize that the prognostication of GCRO should also be seen in the wake of our experience.

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