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Indian Journal of Pathology and Oncology

Journal homepage: www.ijpo.co.in

Case Report

Secondary chondrosarcoma of humerus with osteosarcomatous de-differentiation: The importance of being on guard

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ARTICLE INFO

Article history:
Received 06-09-2021
Accepted 21-10-2021
Available online 14-02-2022

Keywords:
Chondrosarcoma
Dedifferentiation
Humerus
Osteosarcoma

ABSTRACT

Dedifferentiated chondrosarcoma is a highly aggressive but fortunately a rare variant of chondrosarcoma. It is characterized by juxtaposed elements of malignant cartilaginous tumor proliferation alongside mesenchymal tumor of the high malignancy. Careful radiological assessment coupled with sampling of different areas of the biopsy is important for planning treatment. We are reporting a case of secondary chondrosarcoma of humerus which underwent dedifferentiation to fibrosarcoma and osteosarcoma with a fatal outcome.

1. Introduction

Presence of dedifferentiated component in a chondrosarcoma is associated with high incidence of metastasis and a poor overall survival rate. [1–5] Characterized by a sharp juxtaposition of a cartilaginous component with a high grade sarcomatous area, accurate pre-operative diagnosis of dedifferentiated chondrosarcoma(DDCS) requires a careful radiological assessment followed by a multipoint biopsy. [6–9] We present a case of secondary chondrosarcoma of humerus in a middle aged man with dedifferentiation into fibrosarcoma and osteosarcoma which was surgically resected but had a fatal outcome. This case highlights the importance of being alert while working up these cases.

2. Case Report

A 50-year-old right hand dominant male, laborer by occupation, presented to the out-patient clinic with huge swelling on the left mid arm (Figure 1 a). There was a history of small bony swelling at the same site since many years which started to grow rapidly in size since last 5 months. This was accompanied by throbbing pain which was continuous in nature and significant loss of weight. There was also associated weakness of the forearm and numbness over the dorsum of forearm and hand.

On examination, an ovoid shaped solitary swelling was noted in the anterolateral aspect of left arm which was of variable consistency (firm to hard). The overlying skin was stretched out, shiny with dilated veins. The swelling had a bosselated surface with poorly defined margins and was found to be arising from the underlying bone. Also, the patient was found to have complete motor loss in the extensor muscles of forearm with complete sensory loss in dorsum of forearm and hand. There was no vascular deficit.

A clinical impression of soft tissue or bony neoplasm with involvement of radial nerve at the level of mid arm was made. Blood investigations showed hemoglobin of 10.1gm/dl, ESR of 54mm/hr and Alkaline phosphatase of 388 IU/L. Rest of the blood parameters were within normal range. Renal and Pulmonary function tests were normal.

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https://doi.org/10.18231/j.ijpo.2022.020

Radiograph of involved arm (Figure 1 b) showed multiple ill-defined radiolucent lesions involving the entire length of left humerus with endosteal scalloping and breach of cortex at few areas. The extensive soft tissue invasion, calcification and periosteal reaction can be noted at distal half of the humerus. Magnetic resonance imaging of left arm showed (Figure 1 c) focal intramedullary lesion involving nearly the entire length of left humerus with sparing of distal metaphysis and proximal subchondral bone. Presence of cortical breach in the middle third of humerus with extension of lesion into intermuscular compartment involving biceps, brachialis, brachioradialis muscles was noted. There was a partial encasement of the radial nerve.

Further investigations were carried out to stage the tumor. There were no any distant metastasis to lungs, intraabdominal organs and other bones.

A Needle Biopsy under general anesthesia was performed with sampling from heterogenous areas. On histopathological examination, a cartilaginous tumor tissue was seen exhibiting increased cellularity and presence of binucleate chondrocytes with mild to moderate nuclear atypia and conspicuous nucleoli. Also present were few fragments of tumor tissue with sweeping fascicles of spindle shaped cells exhibiting moderate nuclear atypia. No mitosis or necrosis were noted. The needle biopsy was reported as consistent with chondrosarcoma with a possibility of de-differentiation (to fibrosarcoma).

TNM staging of 2B was established and institutional tumor board advise taken which suggested a forequarter amputation for the patient followed by chemoradiotherapy depending upon the excisional biopsy report.

Patient did not give consent for the above advised procedure instead agreed only for shoulder disarticulation. A tennis racquet incision was made at the axilla with posterosuperior aspect of shoulder as the base of the musculocutaneous flap. Deep tissues were dissected, neurovascular bundles were isolated carefully, tagged, ligated and severed. Joint capsule was opened up, adjoining ligaments were dissected and the glenohumeral joint was disarticulated subsequently. Ensuring homeostasis and clear margins of the stump along with musculocutaneous flap the wound was closed with drain in situ. The disarticulated limb was sent for histopathological examination.

Grossly, the specimen of left shoulder disarticulation showed a solid-cystic grey white, firm to hard tumor involving almost complete diaphysis and infiltrating into the surrounding soft tissue. The overlying skin showed hyperpigmentation. Few tumor areas were fleshy in appearance. The tumor measured 27.5x15x15 cm with cartilaginous areas and foci of calcification. Large areas of hemorrhage and necrosis were noted (Figure 1 d) Histopathology showed a heterogeneous tumor with chondroid component composed of atypical chondrocytes in sheets (Figure 2 a, b) The cells displayed nuclear pleomorphism and hyperchromatic nucleus. These areas were juxtaposed with low grade fibrosarcoma (Figure 2 c,d). The most striking feature was presence of pink lacy osteoid surrounding some of the malignant tumor cells (Figure 2 e, f). The tumor cells showed variable mitotic rate ranging from 1 to 3 per high power fields. The tumor showed infiltration in to the surrounding muscle and fat, reaching up to the skin infiltration. Based on the morphological findings, a diagnosis of dedifferentiated chondrosarcoma with components of osteosarcoma and fibrosarcoma was given. Postoperatively, wound was inspected on 3^{rd} and 7^{th} day and sutures were removed at 14^{th} day. The patient deferred chemotherapy and expired eleven months after the surgery.

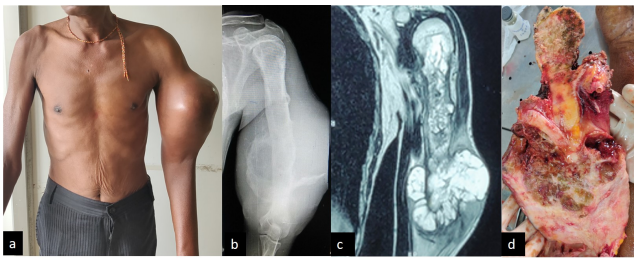

Fig. 1: a): Clinical picture of the left arm swelling, **b)**: X-ray showing involvement of humerus mid shaft with cortical breach and soft tissue mass with calcification, **c)**: MRI coronal image showing diffuse involvement of humerus with soft tissue extension, **d)**: Gross resection specimen

3. Discussion

Dedifferentiated chondrosarcoma is a rare highly aggressive tumor which manifests in 6^{th} to 7^{th} decade of life with no gender predilection.[1–3] However, when the chondrosarcoma is secondary to an enchondroma, the presentation is earlier, as seen in this case.[9] Pain, swelling, increase in size of an enchondroma or a recurrence after excision should alert the clinician to look out for a malignant transformation. In most of the cases, a chondrosarcoma arising from an enchondroma is of grade I.[4,9] The present case however had a high grade cartilaginous component along with dedifferentiation into fibrosarcoma and osteosarcoma. Other components which can be seen are malignant fibrous histiocytoma, leiomyosarcoma, angiosarcoma, rhabdomyosarcoma, lipoblastoma and very rarely an epithelioid sarcoma.[1,9]

The common sites involved by DDCS are femur and pelvis while humerus, ribs, spine tibia and upper extremities are rarer sites.[4]

In a dedifferentiated chondrosarcoma, the cartilaginous component is usually seen within the bone and the dedifferentiated component exists outside in the soft tissue.[1]

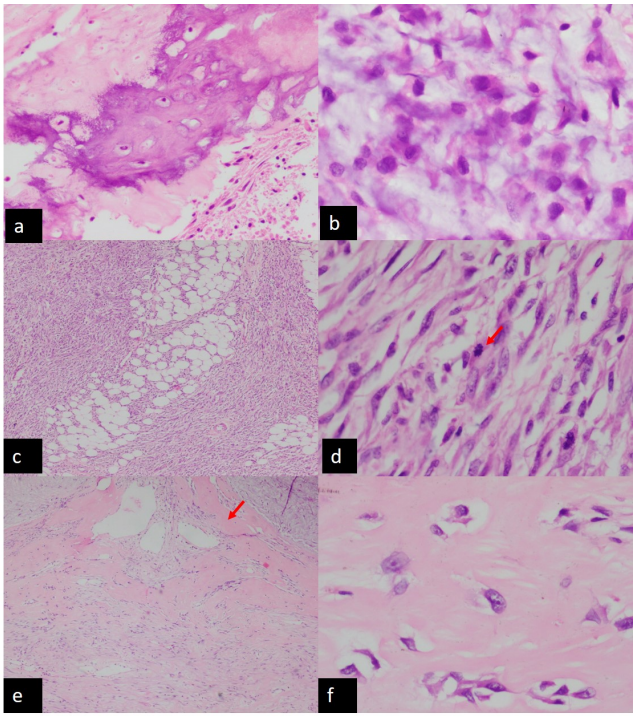

Fig. 2: a,b): High grade chondrosarcoma juxtaposed with fibrosarcoma and osteosarcoma, atypical mitosis and pink lacy osteoid marked with red arrows (**d,e**)

The appearance of these areas foretell a grave prognosis and hence are important to diagnose preoperatively. Careful radiological assessment is very important as juxtaposition of cartilaginous area and sarcomatous area with biphasic nature is a characteristic finding. Another clue is calcification within or adjacent to dominant osteolytic lesion. CT scan helps to appreciate fine calcification in the lesion, while MRI helps to assess the spread of tumor which is very essential to plan a surgery to get clean resection margins.[1,6] Hence both these modalities should be employed for the radiological work up. Preferably, the biopsy should be image guided and multipoint to ascertain that different tumor areas are sampled. It is equally essential that the biopsy be interpreted by an experienced pathologist so that no areas are missed. Variation in histologic assessment of cartilaginous tumors have been reported in the literature.[10] Being able to identify osteoid is important as osteosarcomatous component has a very high rate of metastasis and has the worst prognosis. Different variants of osteosarcoma can be found in the undifferentiated areas like telangiectatic osteosarcoma.[7] In the present case, osteoid was observed in the resected specimen and was of conventional type.

Worse outcome in DDCS is seen in association with a pathological fracture, larger tumor size, lymph node involvement and extra-osseous involvement. Whereas, an improved overall survivability is seen with surgical resection and chemotherapy which is the standard treatment modality for DDCS.[5,8] In this case, an amputation with shoulder disarticulation was done which gave a clean resection margin, but chemotherapy could not be given and postoperative assessment for metastasis could not be done as the patient could not survive the disease.

4. Conclusion

This case is reported to reiterate the importance of adequate tissue sampling in cases when a malignant transformation of a chondral lesion is suspected. After a clinical suspicion, radiological examination can guide the biopsy procedure which can give sufficient material for histopathological examination and thus the concept of triple diagnosis in orthopaedic oncology is always desirable.

5. Source of Funding

Nil.

6. Conflict of Interest

Nil.

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Cite this article: Gupta P, Pathak P, Goel S, Madegowda A. Secondary chondrosarcoma of humerus with osteosarcomatous de-differentiation: The importance of being on guard. *Indian J Pathol Oncol* 2022;9(1):84-87.