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Case Report

A case report of retroperitoneal dedifferentiated chondrosarcoma with femoral nerve involvement

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ABSTRACT

Chondrosarcomas, a type of malignant bone tumour, are relatively rare and originate from cartilage producing cells. Dedifferentiated chondrosarcoma (DDCS), which makes up about 10% of all chondrosarcomas, is a rare and extremely aggressive subtype with high risk of metastasis.

Case presentation- Here we report a case of 60-year-old lady with left lower abdomen lump of 10x10cm since 5months with normal gait, normal sensory, motor, spine and hip examination which shows no bony involvement in X-rays. On CECT and MRI there was large heterogenous enhancing soft tissue density mass lesion of size 11.7×10.9 cm with multiple coarse calcifications, without bone involvement. On Ultrasound guided core needle biopsy, it was found to be low grade chondrosarcoma. Patient was planned for wide local excision of tumor with adequate margins. Intraoperatively femoral nerve was seen passing inside the tumor which was resected and nerve repair done with sural nerve graft. In Post operative period patient had difficulty in extending left leg so knee gaiter was advised for support. After 6months follow up patient was seen healthy and walking without knee gaiter.

Discussion- Dedifferentiated chondrosarcoma (DDCS), which makes up about 10% of all chondrosarcomas, is a rare and extremely aggressive subtype. It is associated with high rates of metastasis and poor survival. Surgery is generally the standard of care for patients and adequate margins are recommended. Role of chemoradiotherapy is controversial. Surgery is the treatment of choice in dedifferentiated chondrosarcoma in patients with local disease with no metastasis.

Keywords: Dedifferentiated chondrosarcoma, Femoral nerve, Wide local excision, Knee gaiter, Sural nerve graft

INTRODUCTION

Chondrosarcomas, a type of malignant bone tumour, are relatively rare and originate from cartilage producing cells.[1] Chondrosarcomas predominantly manifest in the bony skeleton, though a minor proportion appear as primary soft tissue masses. These tumours have the potential to develop anywhere in the body, but they are most commonly found in the extremities (45%), followed by the axial skeleton (31%).[2] The most prevalent variant of chondrosarcomas is the conventional primary type, accounting for 85% of all cases. There are also fewer common subtypes, which include secondary chondrosarcomas that develop from benign precursors, as well as dedifferentiated, periosteal, mesenchymal, and clear cell variants.[3] De-differentiated chondrosarcoma in retroperitoneal region is rare, here we present a case of retroperitoneal de-differentiated chondrosarcoma arising from left iliac bone encasing the femoral nerve.

CASE REPORT

A 60-year-old lady with no known comorbidities presented to our tertiary care centre Lady Hardinge Medical College and associated Smt Sucheta Kriplani Hospital, New Delhi, India with chief complaints of lump in left lower abdomen since, 5 months which gradually increased in size from 3×3 cm to 7×7 cm and pain in lower abdomen which was dull aching, moderate in intensity and radiating to thigh. On examination patient was conscious

oriented with stable vitals, a lump of 10×10 cm present in left iliac fossa extending to left lumbar and suprapubic region, hard in consistency, non-mobile and non-tender. It had well defined upper and medial margins but lateral and inferior margins could not be palpated. Spine and hip examination were normal. Patient had normal gait, sensory and motor examination was normal. Bilateral lower limb peripheral pulses were palpable. All routine blood investigations like complete blood count, liver function tests, renal function test, coagulation profile was within normal range and tumour markers like CEA levels were normal

On radiological investigation, on X-ray hip and bilateral pelvis there was no bony involvement seen. On ultrasound whole abdomen there was well defined heteroechoic mass lesion in the left iliac fossa with minimal vascularity and internal foci of calcification, lesion was seen abutting left iliac bone. On CECT abdomen and pelvis there was large heterogenous enhancing soft tissue density mass lesion of size 11.7×10.9 cm with multiple coarse calcifications, lesion seen abutting left iliac blade and extending in left iliac fossa and displacing vessels medially, no bony erosion present. On MRI abdomen and pelvis there was large heterogenous mass lesion of size 11.1×11.1×8.5 cm noted in left iliac fossa abutting left iliac bone, left anterior abdominal wall, reaching upto left hip joint without obvious joint cavity involvement, femoral nerve cannot be seen. On T2W image it shows areas of both solid and cystic components. Ultrasound guided core needle biopsy was done which showed low grade chondrosarcoma.

Diagnosis was made of Retroperitoneal chondrosarcoma with suspected origin from soft tissue or bone. Patient was planned for wide local excision of tumour with adequate margins. In intraoperative findings there was 15×10 cm encapsulated mass present in left iliac fossa attached to the inner table of ileum which was abutting the internal oblique and transversus abdominis muscle with overlying thinning of iliacus muscle. Femoral nerve was seen passing inside the tumour. Left ureter visualised and found normal, femoral artery and vein were displaced by tumour and were normal. Wide local excision of tumour was done with adequate margins along with inner table of iliac blade. Femoral nerve which was passing inside the tumor was resected with 5 mm margins and femoral nerve repair was done with three strands of sural nerve graft with prolene 6-0 and fibrin glue. Left lower abdomen wall reconstruction done with inlay composite mesh repair. Histopathology shows de-differentiated tumour with all negative margins arising from left iliac bone. Proximal and distal end was found free from tumour. In post operative period patient had difficulty in walking and extending left leg, patient was advised knee gaiter for support to help in walking. Patient started walking with the help of knee gaiter and walker on POD8. Patient was discharged on POD10. After 6 months follow up patient was seen healthy and waking without the help of knee gaiter with the help of using only a cane as support.

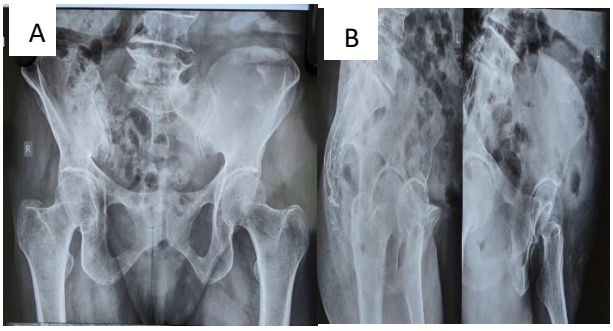

Figure 1: A) and B) showing X-ray of pelvis and hip joint with no bony lesions.

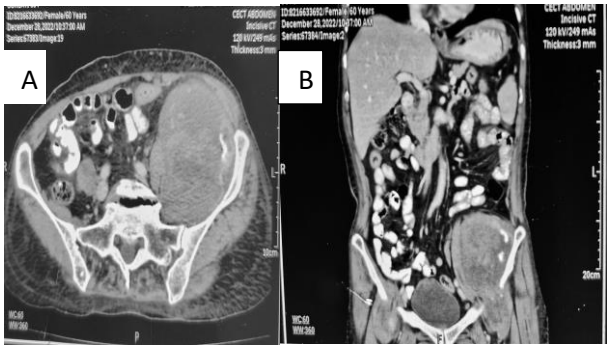

Figure 2: (A, B)- Cect abdomen with coronal and sagittal section showing the mass (blue arrow) abutting left anterior abdominal wall, left iliac bone, and displacing vessels (yellow arrow) medially with internal calcification.

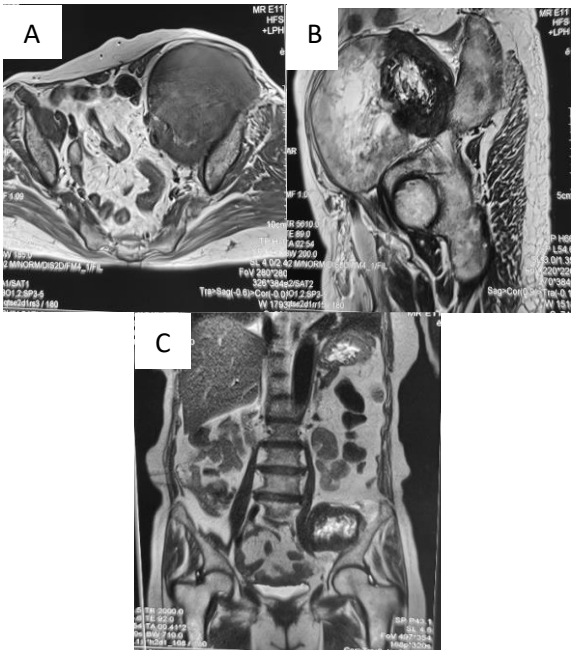

Figure 3: A) MRI abdomen with mass in left iliac fossa abutting left iliac blade and anterior abdominal wall, no bony involvement. B) Mass with solid and necrotic areas reaching upto left joint cavity without joint involvement. C) Sagittal section.

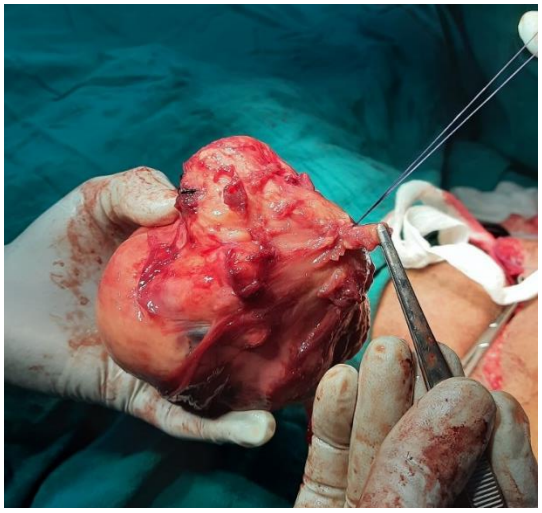

Figure 4: Intraoperative findings of tumor specimen with femoral nerve (yellow arrow) passing inside tumor.

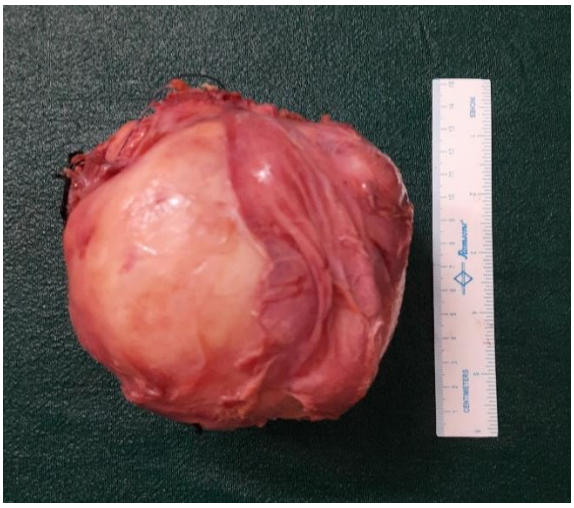

Figure 5: Well encapsulated tumor specimen of size 12×10 cm.

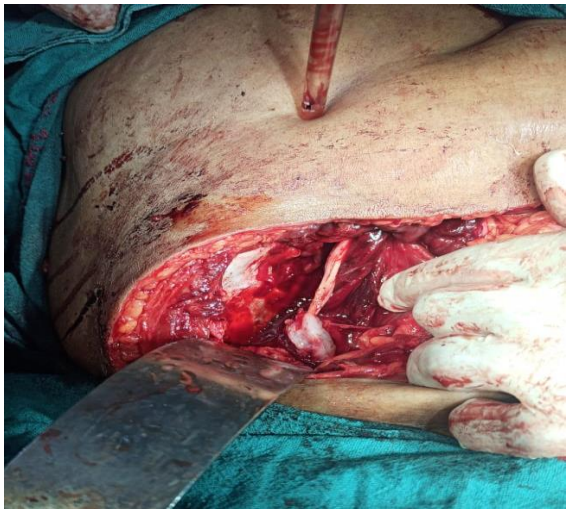

Figure 6: Femoral nerve repair with sural nerve graft.

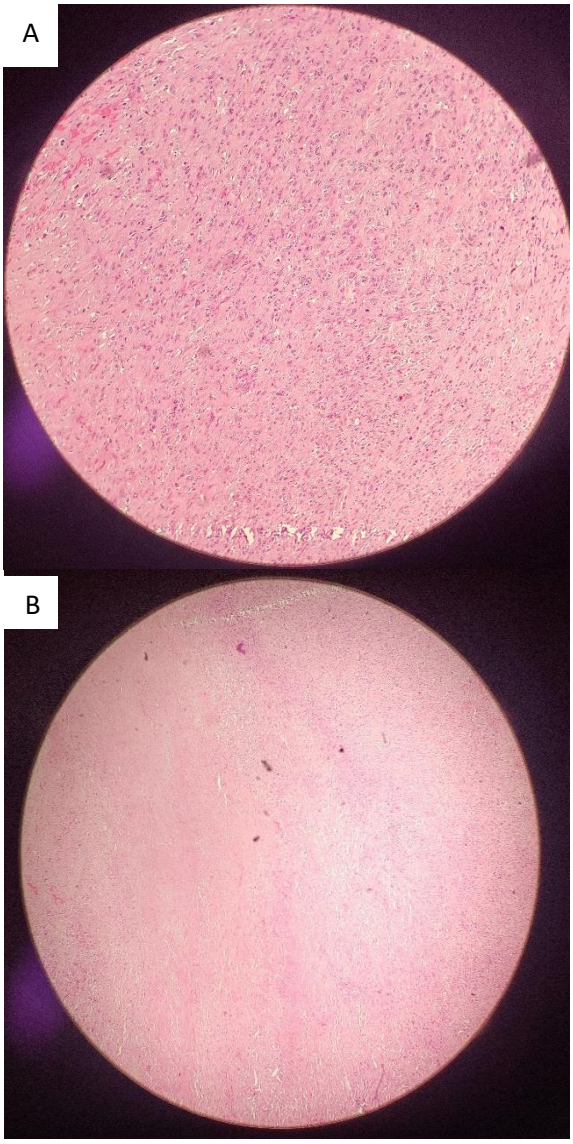

Figure 7: (A, B) Histopathology image.

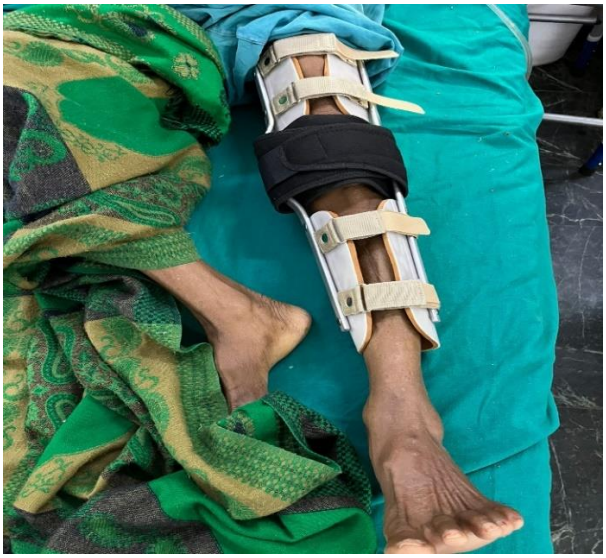

Figure 8: Patient wearing knee gaiter for support.

DISCUSSION

Cartilaginous tumours can be either benign or malignant. Chondrosarcoma, the malignant variant, is a diverse group of tumours that includes conventional, clear cell, mesenchymal, and dedifferentiated types. Dedifferentiated chondrosarcoma (DDCS), which makes up about 10% of all chondrosarcomas, is a rare and extremely aggressive subtype. It is associated with high rates of metastasis and poor survival.

Histologically, DDCS is characterized by a low-grade cartilage tumour that is adjacent to a non-chondroid, high-grade sarcoma. In radiographic images, DCS is seen as a destructive lesion, often accompanied by a soft tissue mass.[4] Dedifferentiated chondrosarcoma (DDCS) can indeed develop spontaneously. However, in about half of the cases, the high-grade component of DDCS emerges from a pre-existing low-grade chondrosarcoma. This suggests that DDCS can evolve from less aggressive forms of the disease.[5]

Dedifferentiated chondrosarcoma (DDCS) primarily occurs in the skeletal system, particularly in the appendicular skeleton, with a higher likelihood of appearing in the lower limb. The most common sites of occurrence are the femur (over 35% of cases), pelvis (up to 29% of cases), and humerus (16%). It can also appear in the scapula (6%), rib (6%), and tibia (5%). The symptoms of Dedifferentiated chondrosarcoma (DDCS) are often nonspecific and tend to appear as the disease progresses. Initially, there may be no visible signs of the malignancy. However, as the disease advances, symptoms such as pain, swelling, edema, and paraesthesia may occur in the vicinity of the tumour.[6]

While there isn't a specific treatment protocol for Dedifferentiated chondrosarcoma (DDCS), surgery is generally the standard of care for patients. This approach is primarily successful in patients whose tumours have not metastasized. However, due to the presence of the high-grade component in DDCS, there's a high risk of metastasis and local recurrence even after resection. Therefore, achieving a wide or radical surgical margin is necessary. The rate of recurrence, as reported in various studies, ranges from 18% to 45%.[7]

The role of radiotherapy in the treatment of Dedifferentiated chondrosarcoma (DDCS) is a subject of debate. Some researchers advocate for its use, while others question its effectiveness. This is largely because chondrosarcomas are known to be relatively resistant to radiotherapy. However, radiotherapy can be considered in two specific scenarios: it can be used after resection with the aim of achieving maximum local control, or it can be used as a palliative treatment when resection is not possible.[8]

Chondrosarcomas, including dedifferentiated chondrosarcoma (DDCS), are generally resistant to chemotherapy. While short-term local control can sometimes be achieved, it has not been proven to have a significant impact on the spread of the disease or overall survival. Therefore, the role of chemotherapy in the treatment of DDCS remains controversial, particularly in terms of its impact on overall survival.[9]

CONCLUSION

Dedifferentiated chondrosarcomas are a rare variety of chondrosarcomas. Dedifferentiated chondrosarcoma of pelvis has high recurrence rates when adequate margins are not taken due to the presence of the high-grade component in DDCS, there's a high risk of metastasis and local recurrence even after resection. Surgical resection offers the best prognosis. Pelvic chondrosarcomas are more challenging to excise due to its proximity to critical structures. Functional improvement can be achieved even after nerve resection with the help of nerve graft.

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