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Telangiectatic osteosarcoma in pelvis: A case report

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Abstract

Telangiectatic osteosarcoma is a rare subtype of osteosarcoma. We report an uncommon case of pelvic telangiectatic osteosarcoma in a young diabetic female who presented with a two-year history of a painless limp. The clinical, radiological, pathological features and management of telangiectatic osteosarcoma of the pelvis are described and discussed how uncommon symptoms can lead to diagnostic delay of the disease.

Keywords

Aneurysmal bone cyst; Osteosarcoma; Pelvis; Telangiectatic osteosarcoma.

Introduction

Telangiectatic osteosarcoma (TOS) is a rare subtype of osteosarcoma (OS) with similar radiographic and histological features to aneurysmal bone cyst (ABC) [1]. This may lead to a delay in presentation or misdiagnosis. We report a case of the TOS to illustrate the diagnostic pitfalls and challenges in the management of this rare and aggressive bone sarcoma.

Case Report

A 24-year-old woman presented with a 2-year history of a painless left limp requiring a walking aid and no analgesia. Her medical history included keratoconus diabetes mellitus treated by Metformin (850 mg), Vildagliptin/Metformin (50 mg/1000 mg), and Fluoxetine (20 mg), respectively. The patient denied any history of pain, loss of weight-appetite, fever, or family history of cancer.

A plain pelvis and left hip X-ray revealed a large expansile bone-forming tumour of the left hemipelvis. This involved the acetabulum extending to the left sacroiliac joint (Figure 1).

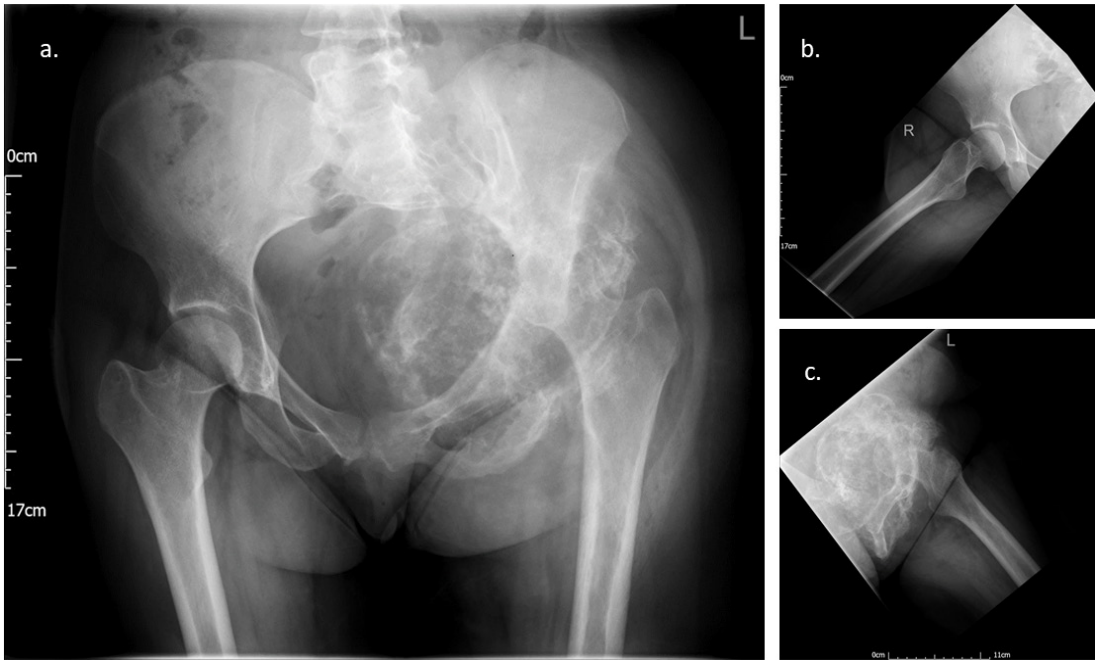

Figure 1: Telangiectatic osteosarcoma in a 24-year-old woman. Anteroposterior X-ray of the pelvis shows a large aggressive tumour of the left hemipelvis (anterior a., lateral right b., and lateral left c.).

Magnetic resonance imaging (MRI) confirmed an extensive bone forming tumour involving whole hemipelvis and left hip involvement (Figure 2). Chest X-ray confirmed no lung metastases. The patient had a planned underwent an open biopsy through the ilioinguinal approach with a diagnosis of a giant cell rich telangiectatic osteosarcoma confirmed at the Sarcoma MDT after Radiological and Pathological Review. Staging studies including CT chest revealed no distant or other metastatic disease.

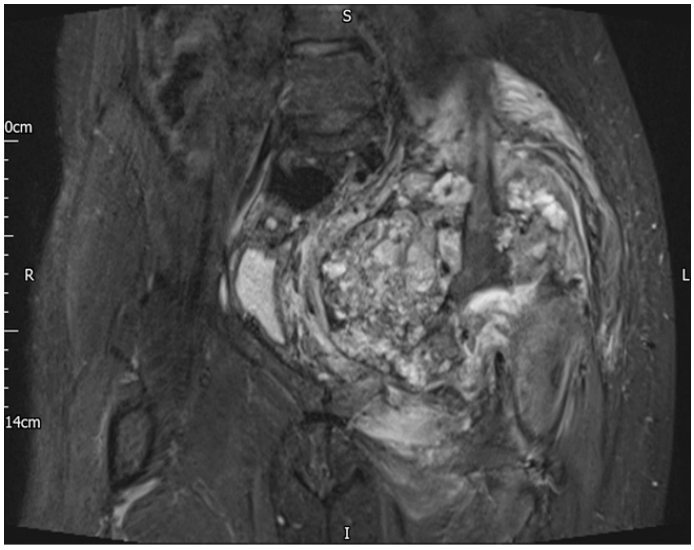

Figure 2: Magnetic resonance imaging shows an extensive bone forming. The tumour has multiple fluid-fluid levels and extended into the pelvis and eroded the hip.

The MDT recommended standard OS neoadjuvant chemotherapy, MAP (methotrexate, doxorubicin, cisplatin). The MAP regimen is consisted of 120 mg/m2, doxorubicin 37·5 mg/m^2 per day on days 1 and 2 (on weeks 1 and 6) followed 3 weeks later by high-dose methotrexate 12 g/m^2 over 4 hours.

Sequential computed tomography (CT) indicated that the tumour had no volumetric response to chemotherapy and minor increase in size, involving the whole hemipelvis and extension across the sacroiliac joint (Figure 3). An external hemipelvectomy (left hindquarter amputation) was undertaken as salvage surgery not achievable due to extent of disease. Post-surgery, the patient completed adjuvant chemotherapy MAP (methotrexate, doxorubicin, cisplatin) + Mifamurtide.

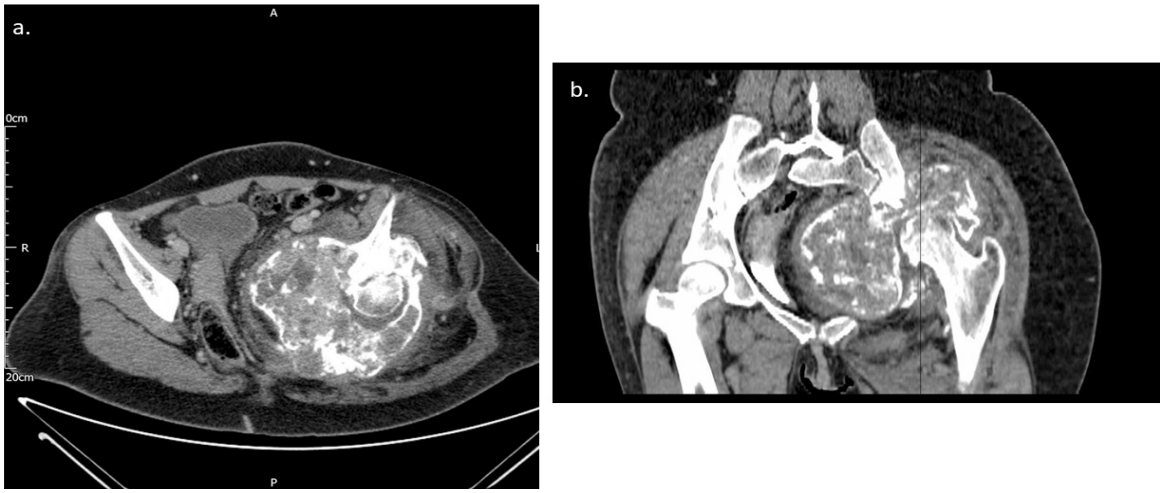

Figure 3: Computed tomography shows the tumour involves entirety of the hemipelvis, the hip joint and crossing the sacroiliac joint. The image also shows a mass lesion contains multiple fluid-fluid levels.

The histopathology examination of the resected specimen showed a large malignant bone forming tumour, 20 cm x 25 cm x 16 cm, which partly destroyed the ilium and widely invaded adjacent soft tissues (Figure 4).

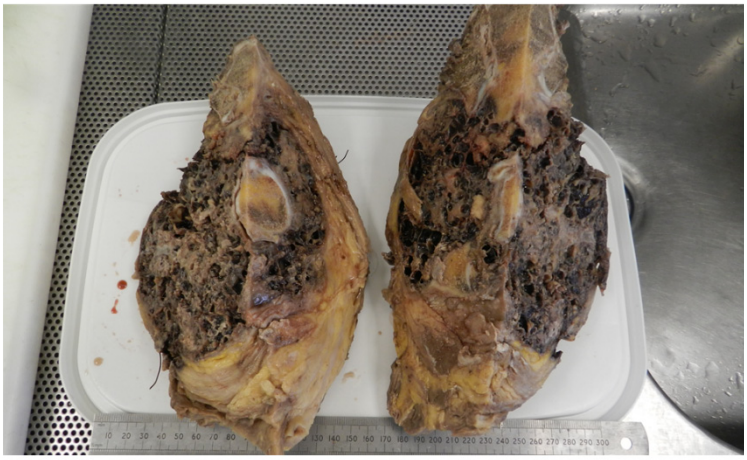

Figure 4: The gross specimen of telangiectatic osteosarcoma, 20 cm x 25 cm x 16 cm, partly destroyed the ilium and widely invaded adjacent soft tissues. The expansile mass contain numerous blood-filled cystic spaces and spongy areas separated by delicate fibrous septa.

The tumour surrounded the acetabulum, but without breaching the hip joint or femur. The tumour was relatively well circumscribed and composed of large cyst-like blood filled spaces lined by markedly pleomorphic tumour cells and a small amount of bone osteoid production. Abundant osteoclast-like giant cells permeated the tumour and septa (Figure 5). Interestingly, the femur showed evidence of osteopenia.

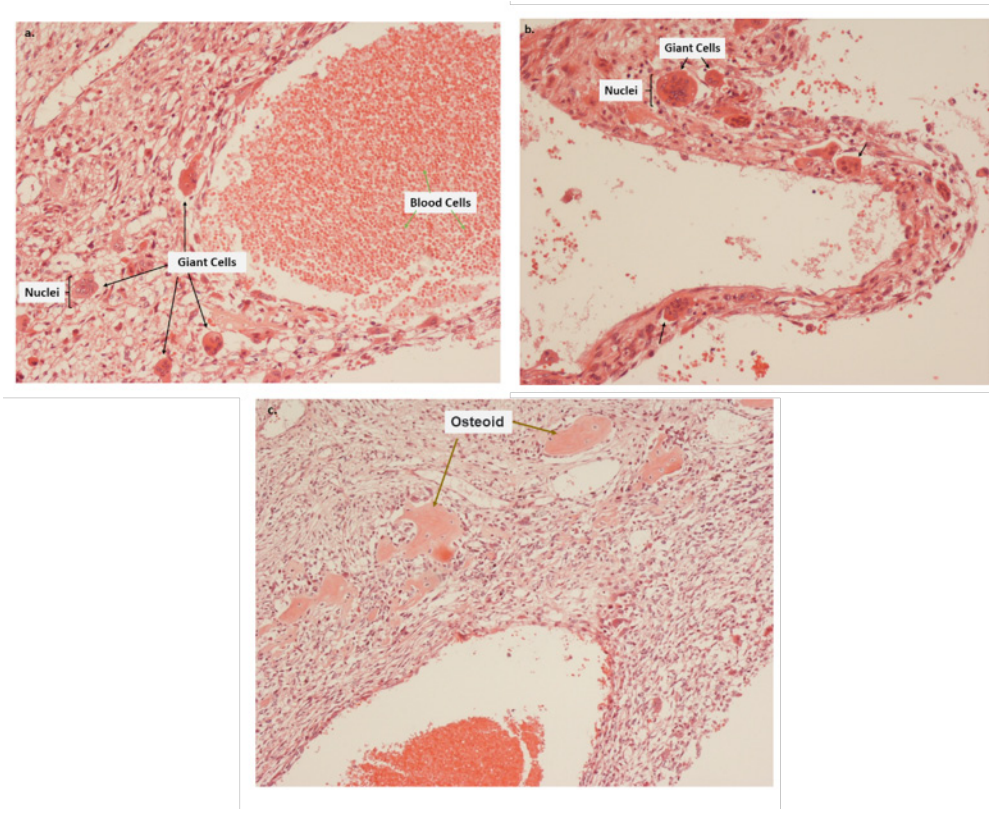

Figure 5: Histopathology of the telangiectatic osteosarcoma showed numerous cyst-like spaces, some of them contains red blood cells and resemble vascular spaces **(a)**. The fibrous septa contain tumour cells and of atypical, bizarre, malignant appearing spindle cells and multi-nuclei osteoclast giant cells with multiple nuclei **(b)**, and bony osteoid production within the tumour **(c).**

Discussion

OS has distinct subtypes including TOS, small cell osteosarcoma, low-grade central osteosarcoma, parosteal osteosarcoma, periosteal osteosarcoma, high-grade surface osteosarcoma, chondroblastic osteosarcoma, and pagetic osteosarcoma [2]. TOS is a rare subtype of OS that accounts for approximately 1% of OS [3] It often occurs in the metaphysal area of long bones including, the distal femur (41.6%), proximal tibia (16.9%) and proximal humerus (9.2%) [4]. The incidence of TOS in pelvis is rare, only 3.1% [5].

The most common symptoms of TOS included pain, swelling, and tenderness in the affected site [6] Although our patient had a limp for two years, the absence of pain delayed her presentation and diagnosis.

The characteristic radiological features of TOS are aggressive-asymmetric expansion, tumour ossification (fluffy, cloud-like) and osteolytic lesion with a permeative destruction with rapid growth tumour and visually minimal peripheral sclerosis [2]. A CT scan is an accurate tool to detect peripheral mineralization and septal enhancement within TOS. MRI is the investigation of choice to identify vascular spaces and fluid- fluid levels within the tumour [2]. Histologically, TOS is often composed of blood-filled or empty cystic spaces resembling ABC, and sometimes this similarity results in misdiagnosis. Blood-filled spaces mimic pseudo vascular spaces within the tumour. The septa are populated by a mixture of malignant osteoid producing OS cells and osteoclast like multinucleate giant cells [1].

TOS patients often present with multiple lung metastasis at the time of diagnosis, particularly when the diagnosis has been delayed for 2 years [7,8]. Interestingly, our patient showed no lung metastasis. She has diabetes, was taking daily Metformin. According to some studies Metformin inhibits metastasis and cell proliferation in MG63 and U2-OS OS cell lines [9,10]. However, this effect remains unproven in the clinical setting.

The standard treatment for TOS is a combination of neoadjuvant chemotherapy followed by limb-salvage surgery [11]. Most patients who received chemotherapy and post-surgery radiotherapy for contaminated margins developed multiple lung or liver metastases and died [8]. The prognosis of TOS was initially very poor, because of misdiagnosis or delayed diagnosis. The 10-year survival rate of people with TOS is approximately 60%, early diagnosis in TOS with an appropriate treatment is crucial for disease-free survival [7,8].

Normally, sarcoma patients present either with pain or a mass, but not in this instance, indicating that limited mobility can be an important presenting symptom. TOS remains an enigma, labelled an OS because of the osteoid producing malignant cells, the distinguishing features of vascular lakes and abundant multinucleate osteoclast -like giant cells characterise TOS histopathology. There is an urgent need to investigate the underlying mechanism of TOS to understand pathophysiology and ontology of this rare sarcoma.

Declarations

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Conflict of interest: The authors declare no conflict of interest.

Consent statement: The authors have no ethical conflicts to disclose. Informed consent was obtained from the patient.

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