Telangiectatic osteosarcoma in pelvis: a case report

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Abstract

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, and pathological features of telangiectatic osteosarcoma of the pelvis and discuss how uncommon symptoms can lead to diagnostic delay of the disease.

Introduction

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

Case Report

A 24-year-old woman presented with a 2-year history of a painless limp on the left side requiring a walking stick to mobilise. Her medical history included keratoconus associated with diabetes mellitus and anxiety disorders, treated by Metformin (850 mg), Vildagliptin/Metformin (50 mg/1000mg), and Fluoxetine (20 mg), respectively. The patient denied any history of pain, loss of weight-appetite, fever, or family history of cancer.

Radiological examination was conducted with hip X-ray revealed a large aggressive tumour of the left hemipelvis. The tumour involved entirety of acetabulum and extended to the sacroiliac joint. (**Figure 1**).

Magnetic resonance imaging (MRI) showed an extensive bone forming tumour involving whole hemipelvis and eroding the hip (**Figure 2**). There were no lung metastases. The patient underwent an open biopsy through the ilioinguinal approach, and the diagnosis of a giant cell rich telangiectatic osteosarcoma was made by the specialist sarcoma pathology team.

The interdisciplinary sarcoma group recommended neoadjuvant chemotherapy; MAP (methotrexate, doxorubicin, cisplatin) regimen was given to the patient. The MAP regimen consisted of 120 mg/m2, doxorubicin 37·5 mg/m2 per day on days 1 and 2 (on weeks 1 and 6) followed 3 weeks later by high-dose methotrexate 12 g/m2 over 4 hours.

Sequential computed tomography (CT) indicated that the tumour did not respond to chemotherapy and the tumour had increase in size, involving the entire of the hemipelvis, the hip joint and crossed the sacroiliac joint (**Figure 3**). External hemipelvectomy (left hindquarter amputation) was performed in this case. Post-surgery, the patient received adjuvant chemotherapy MAP (methotrexate, doxorubicin, cisplatin) + Mifamurtide.

Histopathological 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**). The tumour surrounded the acetabulum, but did not breach the articular surface, there was no involvement of the hip joint or femur.

The tumour was relatively well circumscribed and mostly composed of large cyst-like blood filled spaces lined by markedly pleomorphic tumour cells and a small amount of bone and osteoid formation. Abundant osteoclast-like giant cells permeated the tumour and septa (**Figure 5**). The femur showed evidence of osteopenia.

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 a local pain, swelling, and tenderness in the affected bone area.[6] Although our patient had a limp for two years, the absence of pain delayed her presentation and the 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] 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 common 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 a need to investigate the underlying mechanism of TOS to understand pathophysiology and ontology of this rare sarcoma.

Declarations

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

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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.).

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.

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.

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.

Figure 5: Histopathology of the telangiectatic osteosarcoma hematoxylin and eosin (H&E) staining 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** .).