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A rare case of rib osteosarcoma: pathological features and review of literature

Saguem Ines.¹, Lobna Ayadi.¹, Moez Trigui.², Abdessatar Bahloul.³, Mariem Ben Amor.⁴, Salwa Makni.¹, Naourez Gouiaa.¹, Tahya Sellami Boudawara.¹

¹. Laboratoire d'anatomie et de cytologie pathologiques; CHU Habib Bourguiba Sfax

². Service d'orthopédie; CHU Habib Bourguiba Sfax

³. Chirurgien cardio-vasculaire de libre pratique ⁴. Carcinologue de libre pratique

RÉSUMÉ

L'ostéosarcome est une tumeur osseuse maligne courante qui produit une matrice ostéoïde. L'ostéosarcome télangiectasique (OST) est une forme rare d'ostéosarcome. Il touche les os longs, surtout l'extrémité inférieure du fémur et supérieure du tibia et de l'humérus, comme l'ostéosarcome commun. L'atteinte costale est exceptionnelle. Nous présentons un cas d'OST costal qui a causé des problèmes diagnostiques à cause de sa localisation inhabituelle et sa ressemblance à un kyste anévrysmal.

ABSTRACT

Osteosarcoma (OS) is a common primary malignant tumor of bones that produces osteoid matrix. Telangiectatic osteosarcoma (TOS) is an uncommon variant of OS. It affects the long bones especially the lower end of femur and the upper ends of tibia and humerus, a distribution similar to the conventional osteosarcoma. The rib involvement is very infrequent. We present a case of TOS of the rib that posed a diagnostic difficulty owing to its unusual location and to its resemblance to giant cell tumor and aneurysmal bone cyst.



I. INTRODUCTION

Osteosarcoma (OS) is the most common primary malignant bone tumor. It usually occurs in adolescents and young adults with a slight male predominance. It arises principally in the metaphyses of the long bones. Short bones, spine and flat bones such as the ribs, are less frequently involved^[1-3]. Several distinct clinico-pathological variants of OS have been recognized.

Telangiectatic osteosarcoma (TOS) is a rare and aggressive subtype that represents from 2% to 12% of all cases of OS^[4-6]. It has the same distribution as the conventional OS. Bones such as sternum, scapula and ribs are uncommonly involved^[7-9].

TOS is characterized by blood-filled cystic formations, resulting in an appearance similar to that of aneurysmal bone cyst radiographically and pathologically. The lesion is identified as osteosarcoma through the detection of malignant stroma in the septa that separate the bloody cysts^[4-7]. We report a TOS in an unusual location, which is the rib. We briefly present its principal characteristics especially the radiological and pathological features that allow us to distinguish it from aneurysmal bone cyst (ABC) and giant cell tumor (GCT).

II. CASE REPORT

In December 2013, a 26-year-old woman presented with a pain and a left-sided chest wall swelling. A chest x-ray showed opacity in the left lung field. A computed tomography (CT) scan revealed a mass measuring 8.1 x 6.3 x 5.5 cm arising from the left chest wall with lytic destruction of the sixth rib (Fig.1).

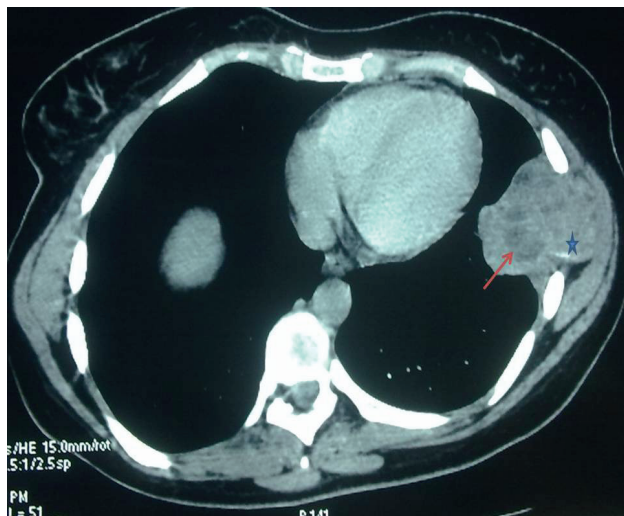


Fig.1: A CT examination of the chest showing a heterogenous solid mass arising from the skeletal chest wall with lysis of the rib (*). This mass has a multiloculated aspect (→)

This mass presented a heterogeneous density and enhancement with necrotic areas. Biopsy of the tumor was performed. At histological examination, many osteoclastic giant cells were scattered on a background of mononuclear cells with round regular nuclei (Fig.2).

The diagnosis of GCT was made and the patient underwent a surgery in January 2014. The tumor was excised in whole with two ribs and the chest wall defect was repaired by simple approximation of ribs.

Gross examination revealed a nodular tumor measuring 7.8 x 6 cm, bearing two ribs.

This tumor destroyed one of the ribs and infiltrated the soft tissue in the anterior side. The cut surface of the tumor was half-solid, half-cystic with blood-filled cavities (Fig.3). Microscopic examination revealed a heterogeneous tumor (Fig.4) with areas that simulated GCT and others that had the same appearance as ABC.

The presence of atypical round-to-spindle shaped cells proliferation with the osteoid matrix allowed us to confirm the diagnosis of TOS.

The histological examination confirmed the infiltration of soft tissue that was seen macroscopically (Fig.5). After surgery, the patient made an uneventful recovery. Then, she received 6 courses of adjuvant chemotherapy (Doxorubicin-Cisplatin-Ifosfamide alternating with Doxorubicin-Ifosfamide) with good tolerance.

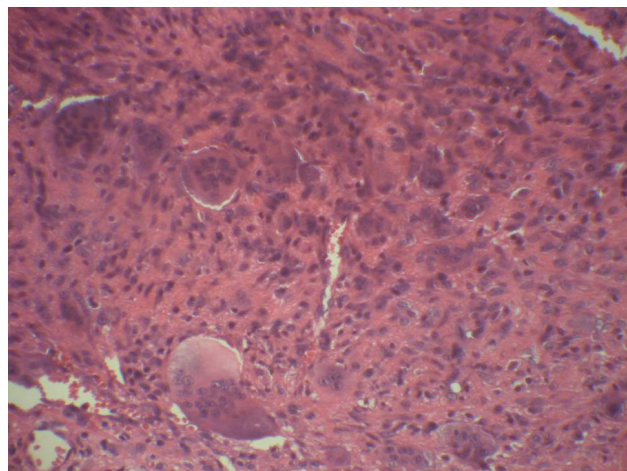


Fig.2: Biopsy of the tumor showing many osteoclastic giant cells scattered on a background of mononuclear cells with round regular nuclei (HE x 400)

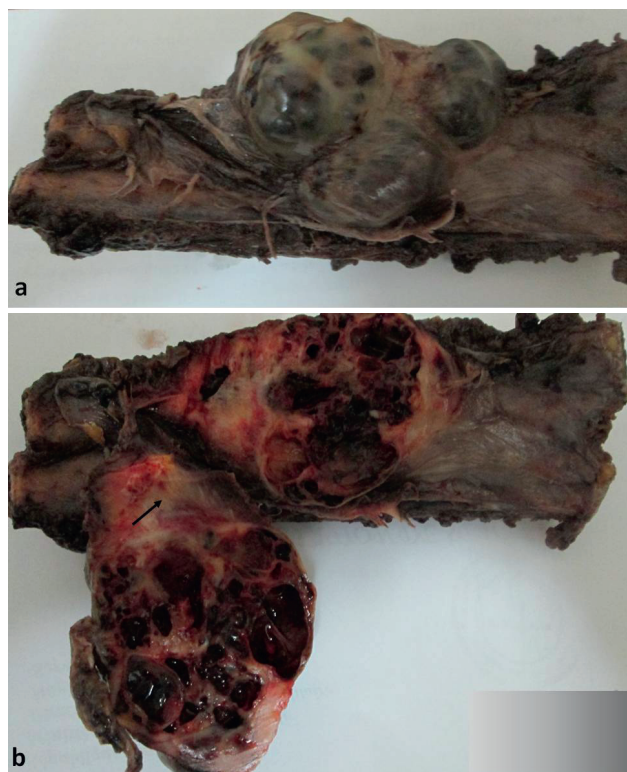


Fig.3: (a): Gross appearance of the tumor with 2 ribs attached. (b): The cut surface of the tumor showing a solid portion (→) with blood-filled cavities separated by fibrous septa.

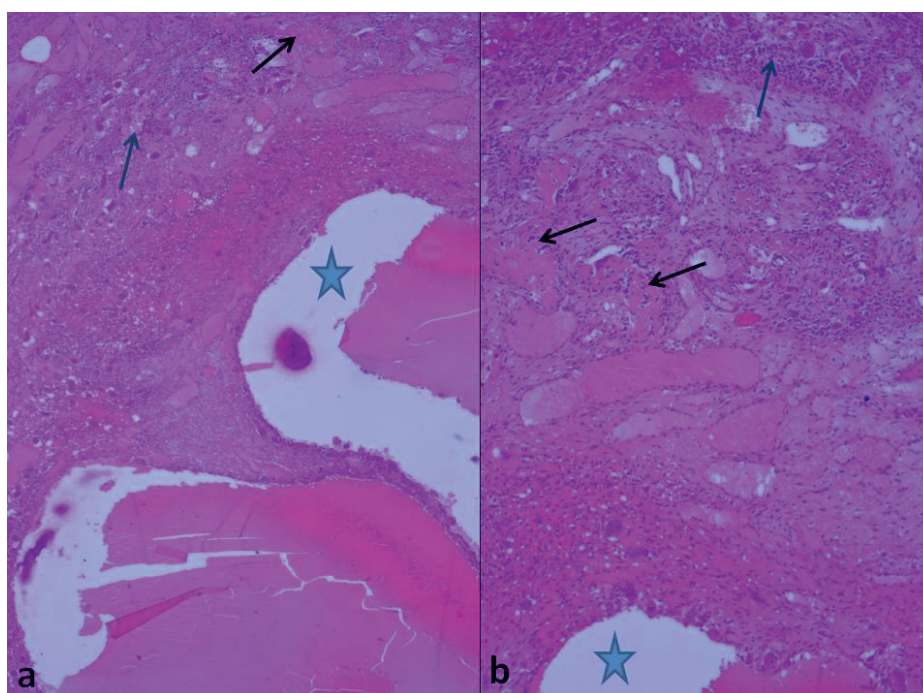


Fig.4: (a) + (b): Low appearance of the different histological features of TOS: blood-filled cystic spaces (*) surrounded by thick septa containing a giant cell proliferation (—>) with osteoid matrix (—>). (HE x 200)

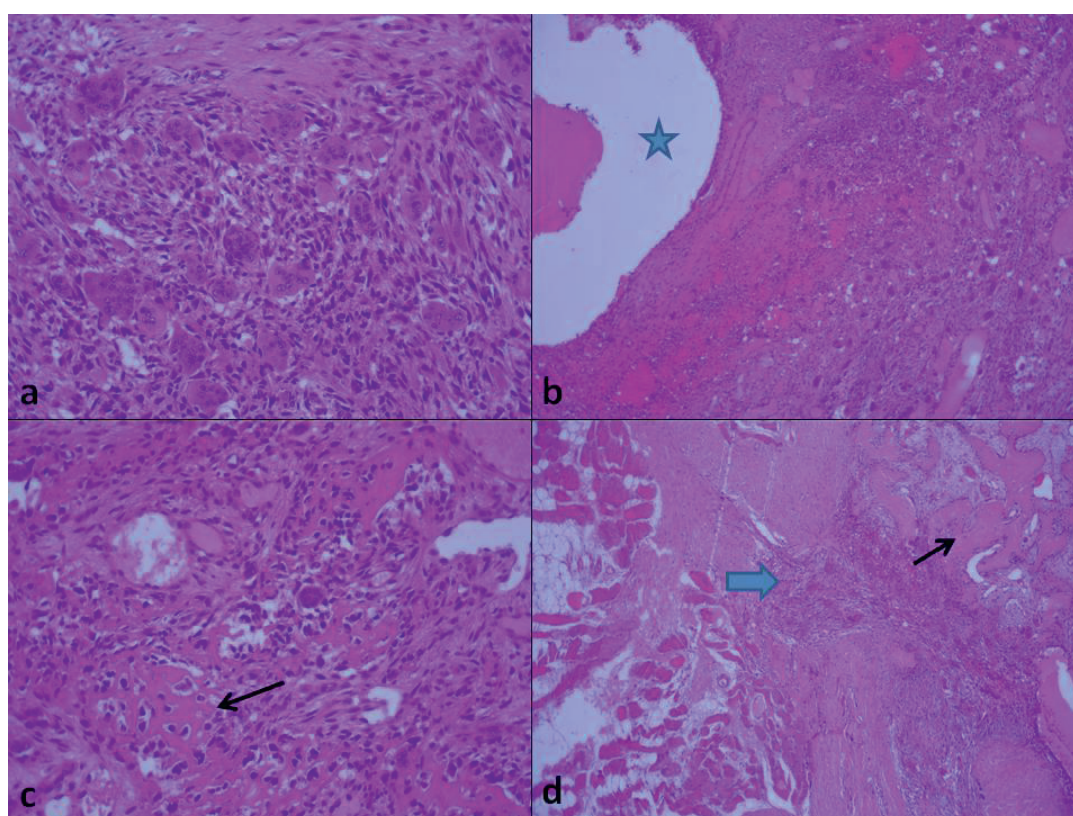


Fig.5: (a): Area resembling GCT similar to that seen on biopsy (HE x 400). (b): ABC-like area with blood-filled cavernous spaces separated by fibrous septa without any atypical stromal cells or osteoid matrix (HE x 200). (c): Note the presence of osteoid matrix (—>) (HE x 400). (d): Infiltration of the soft tissue by the tumor (—>) (HE x 200).

III. DISCUSSION

TOS is a rare variant of osteosarcoma. Patient demographics, clinical symptoms and lesion locations are similar to those in conventional osteosarcoma. In a series of 40 patients with TOS, 68% were males with a mean age of 24 years^[10]. TOS most frequently involve the metaphyses of long bones, especially the femur. The involvement of the rib is so uncommon and accounts for 1.5% of all locations of TOS^[8, 11]. However, Liu et al^[7] provided an overview

of the case reports of TOS that has been published in the literature so far, and no case of TOS occurring in the rib has been presented.

The most clinical signs of TOS include pain, soft tissue mass and pathological fracture.

On a chest x-ray film, the characteristic appearances of TOS include asymmetric expansion, geographic lysis of bone, and an aggressive growth pattern with cortical destruction and minimal peripheral sclerosis. Pathologic



fracture is also frequent^[12]. However, it is sometimes difficult to differentiate whether the lesion is from the rib, lung or pleura. Therefore, CT scan is the best choice as it can reveal the origin, location, component and extend of the tumor. TOS shows as a heterogeneous mass with pure lytic large bone destruction. Extension into soft tissue is often seen. Fluid levels representing hemorrhage are seen in 49% of lesions and osteoid matrix in the intraosseous and/or soft-tissue components of the lesion is seen in 85% of cases^[10, 13]. MR imaging shows thick peripheral, septal and nodular enhancement surrounding the cystic spaces. These enhancing areas should be biopsied.

Macroscopically, TOS is mainly composed of aneurysmally dilated blood-filled cavities with a solid portion. When the blood is washed away, the cyst demonstrates many thin fibrous septa, giving it a sponge-like appearance. Most resections reveal evidence of cortical destruction and extension of the tumor into the surrounding soft tissue.^[8, 9, 11, 13] Histologically, the blood-filled cystic spaces do not demonstrate an endothelial lining. The septa separating those spaces contain highly malignant atypical mononuclear tumor cells. Those cells show nuclear hyperchromasia, pleomorphism and highly mitotic activity including atypical mitoses. The presence of benign multinucleated giant cells admixed with the atypical stromal cells is a common microscopic feature^[14]. A delicate and lacelike osteoid matrix is identified between the malignant cells in some cases; however, this feature is not essential to establish the diagnosis of TOS^[8, 12].

The main and common differential diagnoses are aneurysmal bone cyst (ABC) and giant cell tumor (GCT). ABC is a benign lesion that has radiological and macroscopical appearances similar to TOS^[15]. Only the microscopic examination can aid to have the definitive diagnostic. Histologically, ABC shows multiple blood-filled cavernous spaces with no endothelial lining separated by fibrous septa. These septa are characterized by the proliferation of bland spindle cells intermixed with occasional multinucleated giant cells and immersed in fibromyxoid matrix with inflammatory cells and hemosiderotic macrophages. The mitotic activity is easily identified in the spindle cell component but atypical mitoses should not be seen. The presence of atypical or malignant stromal cells identified in TOS is never seen in ABC and helps exclude the latter condition^[8, 16, 17]. In some cases, extensive sampling and a careful search are essential in order to demonstrate malignant cells^[8]. The other diagnostic consideration is GCT, which is located in the epiphyseal regions of long bones. Its occurrence in the rib is so uncommon. The characteristic histo-pathological appearance is of round to oval polygonal or elongated mononuclear cells evenly mixed with numerous osteoclast-like giant cells. The nuclei of the stromal cells are very similar to those of the osteoclasts. Mitotic figures are invariably present. Atypical mitoses are not however seen and their presence should point to a diagnosis of a giant-cell-rich sarcoma. In addition, it is rare to see septa within cystic giant cell tumors of bone^[8, 18, 19]. In our case, the tumor was initially misdiagnosed as a GCT, the reason for which may be that some of the characteristic histologic

findings were overlooked because of the limited sample volume obtained by biopsy.

The treatment of TOS is similar to conventional osteosarcoma. It generally consists on wide surgery excision followed by adjuvant chemotherapy that increases the chance of relapse-free survival. Surgery should include resection of the full thickness of the chest wall with wide margins that may include the adjacent ribs, intercostal muscles, pleura and vertebrae. If the preoperative definite diagnosis of TOS is established, neoadjuvant chemotherapy might become one of the alternatives for treatment. Recent studies show that employing neoadjuvant chemotherapy with surgery in TOS leads to a similar or even better prognosis than conventional OS^[7, 8, 20].

IV. CONCLUSION

TOS is a rare variant of OS. It needs to be considered in the differential diagnosis for a lytic bone tumor that appears to be malignant even in location where its occurrence is very uncommon such as the rib. The imaging appearance of prominent fluid-filled hemorrhagic spaces simulates that of aneurysmal bone cysts. That is why biopsy should be obtained from the enhancing nodular areas in order to sample the malignant stromal cells.

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