Case Report

Magic box – not only flesh but also with a fatal bone: A rare case of extraskeletal intra-abdominal osteogenic sarcoma

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ABSTRACT

Extraskeletal osteogenic sarcoma is a rare tumor with a poor prognosis. It is histologically characterized by the formation of malignant osteoid, however, there is no association with the axial or appendicular skeleton. It comprises less than 2% of the soft-tissue sarcomas. Postulated risk factors for the development of extraskeletal osteogenic sarcoma are middle and elderly aged patients, history of radiation in the past, and trauma. Here, we report a rare case of extraskeletal intra-abdominal osteogenic sarcoma in a 56-year-old female who presented with a complaint of on and off abdominal pain for the past month. The patient presented with a large retroperitoneal soft-tissue mass with areas of calcifications on CT. Positron emission tomography-CT revealed a large mass with diffuse fluorodeoxyglucose uptake with no other lesion elsewhere in the body. A CT-guided tru-cut biopsy was taken and immunohistochemistry was done on the same. This was proven as osteogenic sarcoma on immunohistochemistry. The patient was started on palliative chemotherapy as the mass was not resectable.

Key words: Conventional osteogenic sarcoma, Extraskeletal osteogenic sarcoma, Retroperitoneal sarcoma

Extraskeletal intra-abdominal osteogenic sarcoma is a rare malignant mesenchymal tumor not in continuation with the appendicular or axial skeleton. To date, less than 300 cases have been reported [1]. Extraskeletal osteogenic sarcomas are rare tumors with a prevalence of 4% of osteogenic sarcoma and 1% of all soft-tissue sarcomas [2]. Here, we present a rare case of intra-abdominal osteogenic sarcoma in a 56-year-old female who presented at our institute. In view of this rarity and difficult location, this case needs a mention in the literature.

CASE REPORT

A 56-year-old female presented at our hospital complaining of on and off abdominal pain for the past 1 month. She also had associated nausea, generalized weakness, loss of appetite, history of weight loss, and decreased frequency of micturition for the past 1–2 months. She was a known diabetic and hypertensive for the past 10 years and was on regular medication for that.

On examination, the patient was European Cooperative Oncology Group status 1. Her vitals were stable. She had a significant history of weight loss of approximately 20 kg from the past 1–2 months.

Her routine blood investigations revealed increased serum creatinine and serum urea levels (serum creatinine 52 mg/dl and serum urea 2.30 mg/dl). Serum sodium was low (129 mmol/L). The other blood investigations were within normal limits.

Fluorodeoxyglucose-positron emission tomography computed tomography (PET/CT) showed a hypermetabolic irregular mass lesion with calcification in the retroperitoneum predominantly in the left para-aortic region encasing the left mid ureter with the left hydronephrosis and with indistinct fat planes with the aorta, forth part of the duodenum, and proximal jejunum. Hypermetabolic mesenteric and omental nodularities with surrounding fat stranding were noted with minimal ascites with low-grade metabolic activity (Fig. 1). For a definite diagnosis, the patient was planned for a CT-guided biopsy.

A CT-guided biopsy was performed from the retroperitoneal mass. Two-three tiny linear cores were obtained. Histopathology showed two-three linear cores infiltrated by poorly differentiated cells invading the adjacent fibrocollagenous tissue. Areas of necrosis were noted. No areas containing osteoid were seen. It was reported as poorly differentiated malignancy (Fig. 2).

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Immunohistochemistry was advised for definitive subtyping and diagnosis.

Immunohistochemistry showed neoplastic cells showing diffuse and strong immunoreactivity for Vimentin and moderate and diffuse immunoreactivity for SATB-2 (Fig. 3a) while they were immunonegative for MDM-2, SMA, and desmin (Fig. 3b-d). Thus, a diagnosis of extraskeletal osteogenic sarcoma was made. In view of her extensive disease and comorbidities, the patient was advised palliative chemotherapy. The patient is on palliative chemotherapy with stable disease.

DISCUSSION

The overall incidence of retroperitoneal sarcoma is less. The most common retroperitoneal sarcomas are the liposarcomas followed by leiomyosarcomas. Extraskeletal intra-abdominal osteogenic sarcoma is an extremely rare entity comprising only 4% of osteogenic sarcomas and 1.2% of all soft-tissue sarcomas [2-4]. Extraskeletal osteogenic sarcoma most commonly occurs in the thigh followed by the upper limb. Retroperitoneum is a very uncommon site as reviewed in the literature [5,6]. Other than the above-mentioned sites, the incidence of extraskeletal osteogenic sarcoma has been reported in the larynx, kidney, esophagus, small intestine, liver, heart, urinary bladder, parotid gland, and breast [3]. Until now, only two cases of intra-abdominal osteogenic sarcoma have been reported [1,7]. The incidence of conventional osteogenic sarcoma has been mostly related to the bones of the appendicular skeleton and axial skeleton occurring in the first two decades of life. The patients with extraskeletal osteogenic sarcoma usually belong to the fourth and fifth decade [3-5]. Even our patient was 56 years old.

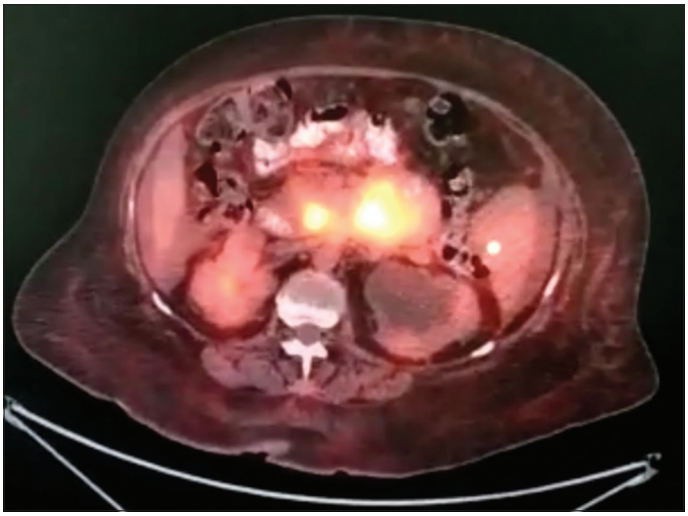

Figure 1: Positron emission tomography image shows large fluorodeoxyglucose avid retroperitoneal mass measuring approximately 10 × 8 × 6 cm

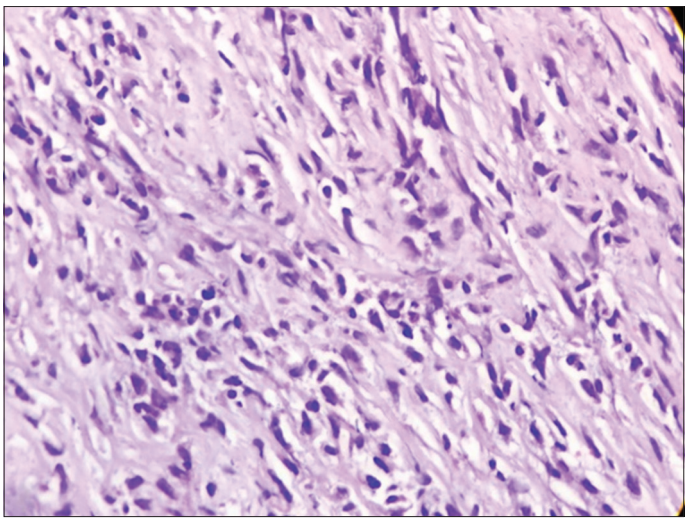

Figure 2: H & E section shows malignant mesenchymal cells, spindeloid in shape with nuclear pleomorphism, and atypia (×40)

The etiopathogenesis of extraskeletal osteogenic sarcoma remains unknown. Few have postulated the exposure to radiation and trauma as the causative factors, however generally, the cause remains unknown [8,9]. Our patient did not have any of the above histories.

The diagnosis is often challenging because a majority of the patients present with very vague symptoms or at times no symptoms at all. Typically, the patients present with advanced disease because of late presentation at the time of diagnosis. One-third of the patients present with intra-abdominal mass associated with a vague pain. Renal function tests might be obliterated, just like in this case if the mass is compromising renal functions. Usually, the rest of the blood parameters are within normal limits [10]. In this case, PET-CT showed a large intra-abdominal mass with areas of calcification along with omental and mesenteric metastasis.

Histomorphology showed sheets of malignant mesenchymal cells along with areas of necrosis. The differentials to be considered in this case are leiomyosarcoma, liposarcoma, and osteogenic sarcoma. Osteoid was not documented in the submitted biopsy; hence, the diagnosis of osteogenic sarcoma was made after immunohistochemistry.

Immunohistochemistry shows diffuse and strong immunoreactivity for Vimentin and SATB-2 while the tumor cells are immunonegative for AE1/AE3, CD-45, desmin, SMA, and MDM2 ruling out other differentials of retroperitoneal malignancies such as high-grade lymphoma, metastatic deposits of epithelial malignancy, leiomyosarcoma, and liposarcoma.

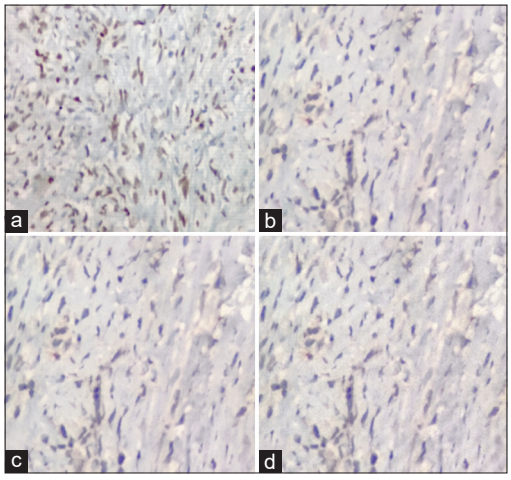

Figure 3: (a) Section shows diffuse and moderate immunoreactivity of tumor cells to SATB2. Tumor cells are immunonegative for (a) desmin, (c) MDM2, and (d) SMA

The overall survival of extraskeletal osteogenic sarcoma is poor with a cause-specific survival rate at 5 years less than 25% [11]. Resection of the tumor is best to have local disease control and to reduce the symptoms, however, it does take care of the metastasis and overall survival [12].

Systemic chemotherapy is the treatment of choice, however, its efficacy has not been proven by clinical trials due to the rarity of this entity. Radiotherapy is opted to achieve temporary palliation [12]. Goldstein-Jackson *et al*. in their article have proposed the use of multiagent aggressive chemotherapy [12]. The patient is on palliative chemotherapy with stable disease. More information needs to be obtained concerning the clinical outcome for appropriate management, planning, and prognostic estimation.

CONCLUSION

Here, we report an extremely rare case of extraskeletal intra-abdominal osteogenic sarcoma arising in the retroperitoneum with extensive omental and mesenteric metastasis. A suspicion of this entity is advised when an intra-abdominal soft-tissue tumor with abundant areas of intratumoral calcification or ossification is seen.

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