CASE REPORTS

EXTRASKELETAL OSTEOSARCOMA OF ANTERIOR ABDOMINAL WALL: A CASE REPORT AND REVIEW OF LITERATURE

Syed Salman Ali, Muhammad Zeeshan, Iqbal Muhammad, Saeed Afzal, Shoaib Naiyar Hashmi,

Syed Naeem Raza Hamdani

Armed Forces Institute of Pathology Rawalpindi, Pakistan

ABSTRACT

We report a case of an extremely rare soft tissue tumour, extraskeletal osteosarcoma in a 62 year old Pakistani male, who presented with a slowly growing painless mass of anterior abdominal wall and died within one year of diagnosis. The clinical, radiological and pathological features of this neoplasm will be discussed, along with a review of the literature.

Keywords: Extraskeletal osteosarcoma.

INTRODUCTION

Extraskeletal osteosarcoma (ESOS) is a rare malignant mesenchymal tumour which does not involve the skeletal system directly[1]. It accounts for 1-2% of all soft tissue sarcomas and 4-5% of all osteosarcomas[2]. To date, fewer than 300 cases of ESOS have been reported[3].

CASE REPORT

A 62 year old male, known case of hypertension and a chain smoker for the last 20 years, presented to our institute in April 2015 with complaints of gradually increasing painless swelling in the left lower abdominal wall for the last 5 years and mild oozing of blood from the swelling for the last 4 months.

History goes back to 5 years, when the swelling appeared as a mild painless nodule in the left iliac fossa, which over the 5 years gradually increased in size and became a small cauliflower like growth, but the patient still ignored it, as it was not associated with any symptoms. He became worried, when blood started to ooze from the swelling. Then he reported to one of the local hospitals, where his biopsy was done and sent to our institute for review and application of immunohistochemistry (IHC). Biopsy review at our institute, reported the lesion as high grade pleomorphic sarcoma more in favour of extra skeletal osteosarcoma. Local examination of the anterior abdominal wall revealed, an irregular swelling in the left iliac fossa, measuring 10 x 6 cm, firm in consistency, attached to the overlying skin but not to the underlying structures with mild oozing of blood.

Figure-1: Gross and cut section morphology of the specimen.

Contrast enhanced CT scan of abdomen was done, which reported the lesion as either sarcoma or desmoid, along with the advice for biopsy. As bone scan already ruled out the possibility of any primary skeletal involvement, excisional biopsy was done in May 2015 and resected specimen was sent to our institute for histopathology. The cut surface of the specimen, showed a solid grey brown tumour with areas of haemorrhage and necrosis (fig-1).

The microscopy revealed multiple foci of neoplastic osteoid surrounded by atypical spindle cells having hyperchromatic nuclei with high NC ratio and a mitotic rate of about 7/10 HPF. A total of 60% of the tumor showed necrosis (fig-2). An extended panel of IHC was applied which showed positivity for osteonectin, vimentin and a Ki 67 index of about 40-

Correspondence: Dr Syed Salman Ali, Histopathology AFIP Rawalpindi Pakistan (*Email:syedsalmanali85@gmail.com)*

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50% (fig-3) and negativity for CD 99, SMA, Desmin, ER, PR and Congo red, thus giving a definitive diagnosis of ESOS. So, the patient was advised follow up chemotherapy with cisplatin, doxorubicin and methotrexate regimen, which he tolerated well initially but unfortunately he died after 2nd cycle of chemotherapy in October 2015.

DISCUSSION

ESOS was first described by Dr. Harwell Wilson in 1941[1]. It is a rare malignant mesenchymal tumor, which occurs outside the bone tissue and is formed of neoplastic cells that produce osteoid and/or cartilage[4]. It is more prevalent in males in the 6th decade of life and accounts for 1-2% of all soft tissue sarcomas and 4-5% of all osteosarcomas[2].

Most common primary site of involvement of ESOS is lower extremity (48%) followed by upper extremity (23%), retroperitoneum (17%) and trunk (11%) whereas the most common metastatic site is lungs (80%) followed by bone (8%), liver (8%), peritoneum and adrenals (<5%)[5].

There are two theories reported with regard to the mechanism behind evolution of ESOS. The tissue residue theory suggests that the mesoblastic component forms during embryonic development and then the formation of bone and osteosarcoma occurs. The metaplasia theory suggests that muscle interstitial fibroblasts are subjected to external or internal stimulation, including trauma, inflammation and metaplasia of the osteoblasts or chondrocytes, which evolves into osteosarcoma[6].

The diagnosis of ESOS must be made using a combination of the clinical manifestations, radiological and pathological findings. Clinically, it usually presents as a slowly growing painless mass. Radiology will reveal a soft tissue mass with variable amounts of mineralization and lack of skeletal involvement. Histopathologically, it shows reverse zonal pattern in which malignant spindle cells with marked nuclear atypia surrounds varying amounts of neoplastic osteoid and/or cartilage[2].

Immunohistochemically, the expression of antigens in ESOS varies in the reported cases.

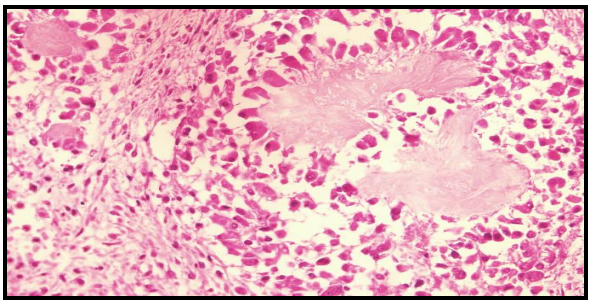

Figure-2: Photomicrograph of ESOS (Haematoxylin-Eosin original magnification 40x).

However, it shows positivity for osteonectin and vimentin whereas negativity for epithelial markers[3]. Today, molecular analysis may resolve the diagnostic dilemma in ambiguous cases. Fluorescence in situ hybridization (FISH) analysis has revealed the amplification and/or overexpression of 2 oncogenes namely MDM2 and CDK4 in ESOS[7].

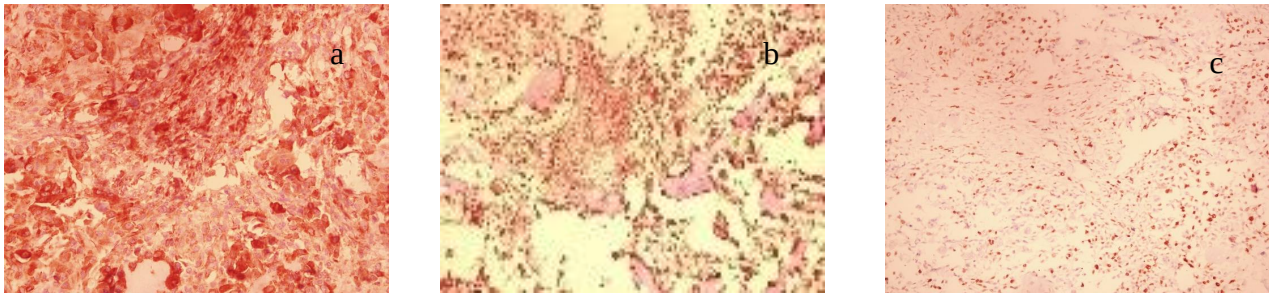

Figure-3: Photomicrograph of ESOS. (a) Osteonectin original magnification 40x. (b) Vimentin original magnification 20x. (c) Ki 67 original magnification 20x.

Histologically, ESOS is divided into 6 subtypes depending upon the predominance of the type of matrix as osteoblastic, chondroblastic, fibroblastic, malignant fibrous histiocytoma-like, talengiectatic and well differentiated[6]. Differential diagnosis to be considered includes myositis ossificans, parosteal osteosarcoma, ossifying fibromyxoid tumour, synovial sarcoma and malignant melanoma[7].

Wide resection is the treatment of choice for extraskeletal osteosarcoma. Adjuvant chemotherapy

and/or preoperative radiation therapy may be useful, although extraskeletal osteosarcoma seems relatively chemoresistant compared to osseous osteosarcomas[8].

Prognosis of ESOS is usually poor. The 5-year survival rate is 37% or less. Although partial spontaneous regression of extraskeletal osteosarcoma has been reported in a few cases. Approximately 50% of the tumors recur locally and lung metastases develop within 3 years after diagnosis. Tumor size is an important prognostic factor. Patients with tumors >5 cm usually have an unfavorable clinical course. The histological subtypes of ESOS have also been related to prognosis. The fibroblastic and chondroblastic subtypes may have a slightly better prognosis compared to the other subtypes[9].

CONCLUSION

The diagnosis of ESOS must be made using a combination of the clinical manifestations, radiological and pathological findings. Clinically indolent lesions sometimes turn out to be malignant on histopathology, which is a gold standard for diagnosis. Role of immunohistochemistry has become vital and reviews in difficult cases are not uncommon, which are meant for quality assurance and as a learning tool.

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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