Case Report

Undifferentiated pleomorphic sarcoma: Diagnosis of exclusion

ABSTRACT

Malignant soft-tissue tumors which were designated as malignant fibrous histiocytoma are regrouped by the WHO (in 2002) under the new entity termed as "undifferentiated pleomorphic sarcoma."[1] It accounts for less than 5% of all adult soft-tissue sarcomas. Here, we report the lesion in a 70-year-old man who presented with high-grade undifferentiated pleomorphic sarcoma in the lower extremity.

Keywords: Adult soft-tissue sarcomas, malignant fibrous histiocytoma, soft-tissue sarcoma of lower extremity, undifferentiated pleomorphic sarcoma

INTRODUCTION

Undifferentiated pleomorphic sarcomas are aggressive tumors, commonly seen in adults. However histopathological pattern is very much variable in these soft tissue malignant neoplasms. We detected this case where proper clinico-histomorphological analysis coupled with immunohistochemistry (IHC) helped us to arrive at a diagnosis.

CASE REPORT

A 70-year-old male presented with swelling over the posterior aspect of the left thigh. Swelling was gradually increasing in size.

Magnetic resonance imaging findings revealed a well-defined altered signal intensity lesion with central area of necrosis in muscular plane of posterior aspect of left mid aspect of thigh involving lateral fibers of biceps femoris.

The patient was subjected to fine needle aspiration cytology [FNAC], the findings of which were suggestive of a soft tissue sarcoma.

Excision of the mass was done and was submitted to us for histopathological examination.

On gross inspection, soft-tissue mass measured 12 cm × 5 cm × 4 cm, with tumor mass of about 3 cm × 2 cm × 1 cm dimensions.

Cut section was gray brown with areas of necrosis in it [Figure 1].

Histopathological examination revealed a malignant tumor with pleomorphic bizarre cells. At places, spindled and tadpole like contour cells were seen. Many histiocytic giant cells were also noted in the sections [Figure 2, 3]. The surgical margins were free of the tumor.

Based on these microscopic findings and the site involved, differentials of pleomorphic rhabdomyosarcoma and an undifferentiated pleomorphic sarcoma were kept. Considering variable histomorphological features, immunohistochemistry (IHC) was suggested.

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Access this article online	
Website: www.asjo.in	Quick Response Code
DOI: 10.4103/ASJO.ASJO_34_17	

How to cite this article: Upasham PS, Dighavkar SP, Roplekar PM. Undifferentiated pleomorphic sarcoma: Diagnosis of exclusion. Asian J Oncol 2017;3:133-5.

Immunohistochemistry

IHC showed CD68 and S-110 protein expression in the histiocytes and histiocytic giant cells.

Tumor cells were immunonegative for smooth muscle actin (SMA), desmin S-100 protein, CD34, CK, epithelial membrane antigen, CD68, and glial fibrillary acidic protein (GFAP).

However, no photographs of IHC panel could be obtained as the patient got it done from different institutes.

Comparing all these findings, a concluding diagnosis of high-grade undifferentiated pleomorphic sarcoma was made. Moreover, the patient was referred for oncotherapy line of management.

However, nearly 2½ months later, the patient reported back with a recurrent painful lesion which was excised again.

Pathological findings showed similar gross and microscopic features as the initial lesion [Figures 4 and 5].

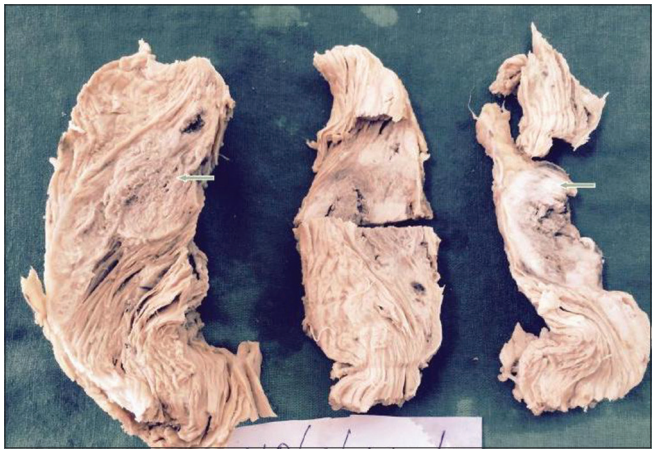

Figure 1: Cut section of the tumor mass

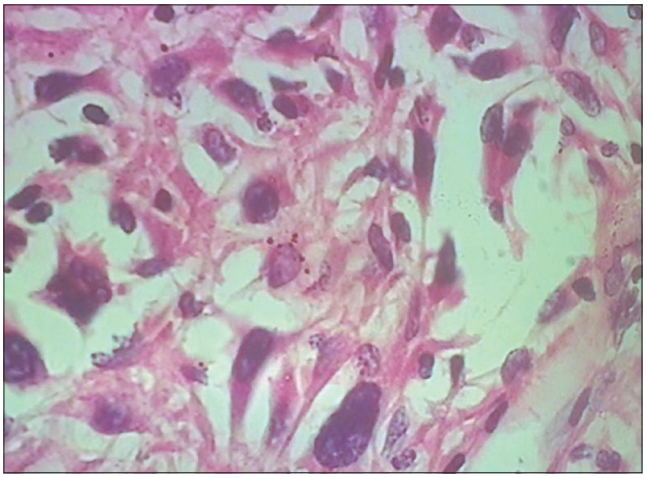

Figure 3: Undifferentiated pleomorphic sarcoma: showing anaplastic and variable cellularity

On attempts to know the follow-up, we got the information that the patient had succumbed to the disease, and cause of death informed to us was multiorgan failure.

DISCUSSION

Undifferentiated pleomorphic sarcomas have high incidence of local recurrence and metastasis.[1]

Accounting for <5% of adult soft-tissue sarcomas,[2] these neoplasms have also been reported in children.[3]

Individuals in the age group of 50–70 years are mostly affected with common site being the lower extremities.[4] However, there have been reports of other rare sites involved by these neoplasms such as retroperitoneum[4] male breast,[5] mediastinum,[1] cutaneous,[6] and pelvic region.[7]

CONCLUSION

These undifferentiated sarcomas are tumors which do not show distinct line of differentiation and obvious morphological features on histopathology. complete

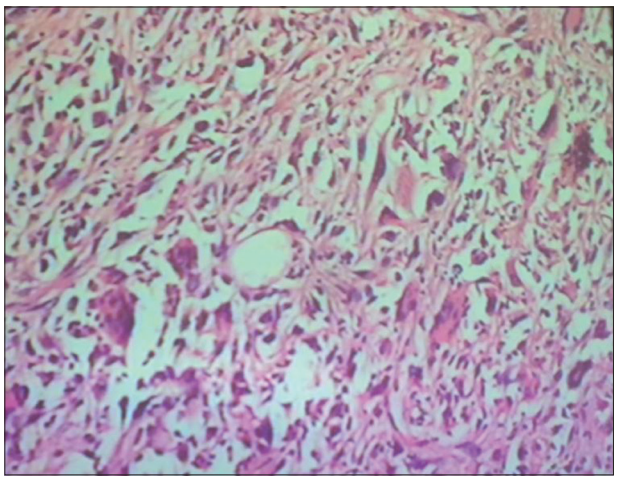

Figure 2: H- and E-stained section from the tumor showing histiocytic cells

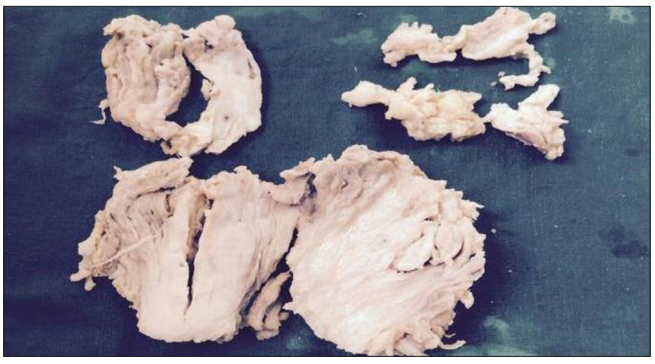

Figure 4: Gross morphology of the recurrent lesion

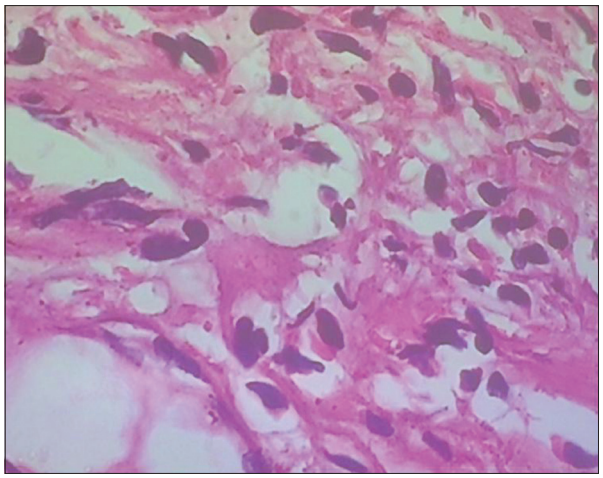

Figure 5: Microscopic features in a recurrent lesion (H and E)

workup is recommended to exclude other soft –tissue sarcomas and classify the lesion into this renamed high grade entity.[8]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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