

*Case Report*

Inflammatory Malignant Fibrous Histiocytoma: A Case Report

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ABSTRACT

The rarest subtype of malignant fibrous histiocytoma (MFH) is the inflammatory type, which has peculiar clinical presentation. Its retroperitoneal position may simulate other retroperitoneal masses like xanthogranulomatous lesion, lymphoma or dedifferentiated liposarcoma. So immunohistochemical study is advised. The unique features of its presentation, multiple local recurrences and metastasis compel surgical wide excision. In this study, a case report of a 60-year-old male patient with a soft-tissue inflammatory MFH presenting as a retroperitoneal mass and a review of the current literature are presented.

Key words: Malignant fibrous histiocytoma, inflammatory type; Soft-tissue sarcoma.

INTRODUCTION

Ozello et al. first described malignant fibrous histiocytoma (MFH) in 1963. [1] A MFH is a soft-tissue sarcoma of undifferentiated mesenchymal cell origin. It is the second most common soft-tissue sarcoma in adults. It commonly occurs in the 5th and 6th decades, with a 2:1 male-to-female predominance involving most commonly extremities and retroperitoneum. [2] Malignant fibrous histiocytomas (MFHs) was newly named as 'undifferentiated pleomorphic sarcomas' in 2002 by the World Health Organization. [3] Inflammatory malignant fibrous histiocytoma (IMFH), also known as undifferentiated pleomorphic sarcoma (UPS) inflammatory variant, was initially identified as a separate category by Kyriakos and Kempson in 1976. [4] The

inflammatory type of MFH is the rarest of all and is characterized by a predominance of xanthoma cells with an intense neutrophilic infiltration. [5] Its name is derived from its distinct histological pattern. These tumors are usually bulky, characterized by multiple local recurrences and metastasis and are most commonly encountered in the retroperitoneal cavity. [4]

We present a patient who was admitted to our hospital with a retroperitoneal mass and was diagnosed as MFH of the inflammatory type- a rarest variant.

CASE PRESENTATION

A 65 yrs old male admitted to our surgical department, presented with a lump in the lower abdomen and low grade fever

since 4 months. The lump was large, firm and well defined in the lower hypogastric region.

USG: showed e/o hypoechoic well encapsulated solid lesion measuring 8.2x10x8.3cm in retroperitoneum. **USG GUIDED FNAC:** showed loosely cohesive clusters of cells with hyperchromatic and pleomorphic nuclei - s/o sarcoma. **CT** showed a large well defined right isodense soft tissue retroperitoneal mass lesion displacing right kidney measuring 13x9.5cm. No e/o any calcification or necrosis noted. On post contrast study e/o heterogenous enhancement was noted. Fat planes with surrounding organs were well maintained

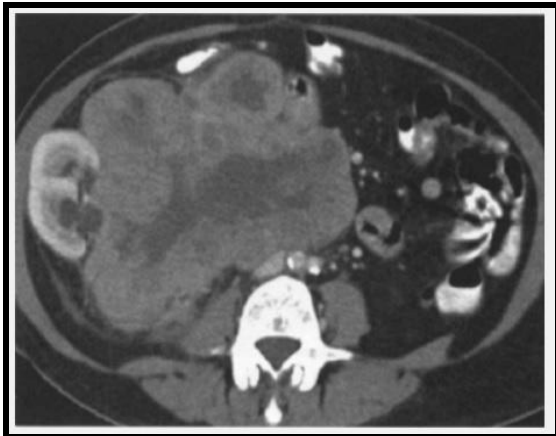


Figure 1: CT scan showing retroperitoneal mass with displacement of right kidney.

Systemic examination revealed no significant findings. Routine investigations revealed anemia, leukocytosis with neutrophilia. Clinical and radiological investigations revealed no evidence of metastasis. No significant contributory family or past history was noted. There was no history of irradiation for the prior lesion. Tumor was surgically excised and sent for histopathological examination.

Histopathological Examination:

Gross: Received specimen of large, encapsulated, glistening greyish white mass, of size 12x10x9 cm. One similar lobulated mass was attached to it of size 4x3 cm. cut

section showed fleshy yellowish white surface with focal areas of haemorrhage and necrosis [Fig. 2].

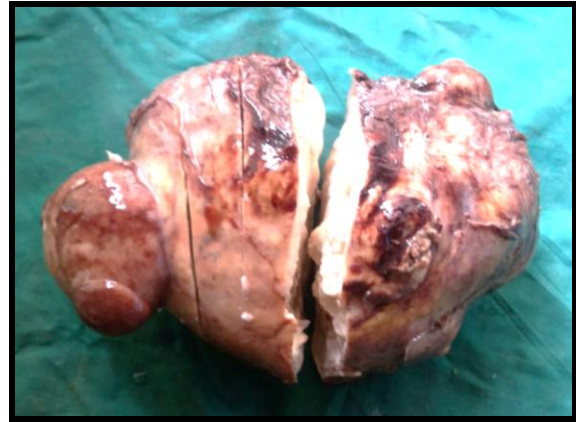


Figure 2: A solid encapsulated mass of size 12 × 10 × 9 cm, C/S: yellowish white.

Microscopy: The tumor was composed of spindle cells having large pleomorphic hyperchromatic spindle shaped nuclei. These spindle cells were arranged in fascicular and storiform pattern. There were large groups of xanthoma cells and dense infiltration of neutrophils, eosinophils, plasma cells masking the tumour cells. Stroma was scanty, myxoid with areas of haemorrhage and necrosis [Fig.3].

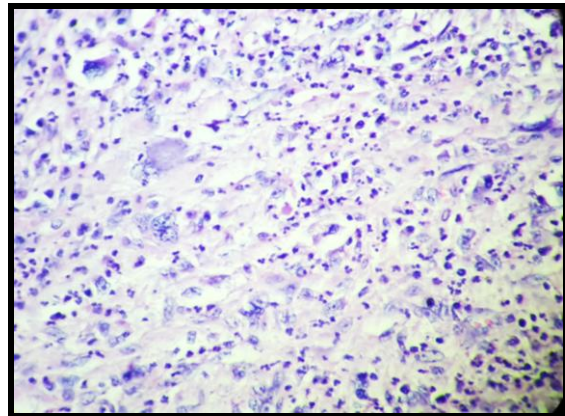


Figure 3: Microphotograph showing tumor composed of spindle cells, xanthoma cells and dense infiltration of neutrophils, plasma cells masking the tumour cells.

On histopathology, diagnosis was given as Inflammatory malignant fibrous histiocytoma or Undifferentiated Pleomorphic Sarcoma, Grade 3/3 (coindre grading system^[6]), Inflammatory type.

The immunohistochemical study was advised. Immunostain showed strong, diffuse vimentin positivity. Tumor cells were immunonegative for LCA, CD30, CD68, MPO, smooth muscle actin, desmin, and MDM2 [Fig. 4 & 5].

Based on this, final histopathological diagnosis was given as Retroperitoneal Undifferentiated Pleomorphic Sarcoma-Inflammatory type, Grade 3/3 (Coindre grading system).

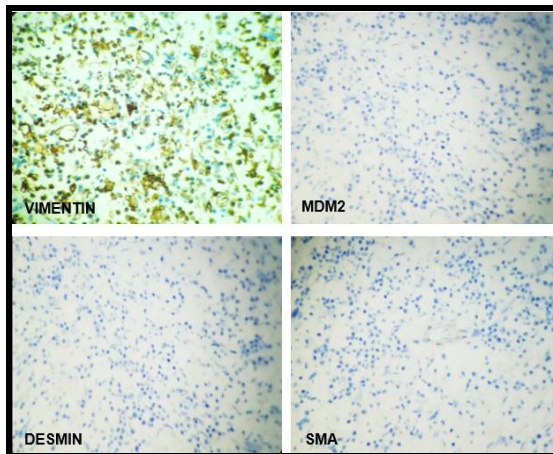


Figure 4: IHC showing positive reactivity to vimentin & negative reactivity to MDM2, DESMIN, & SMA.

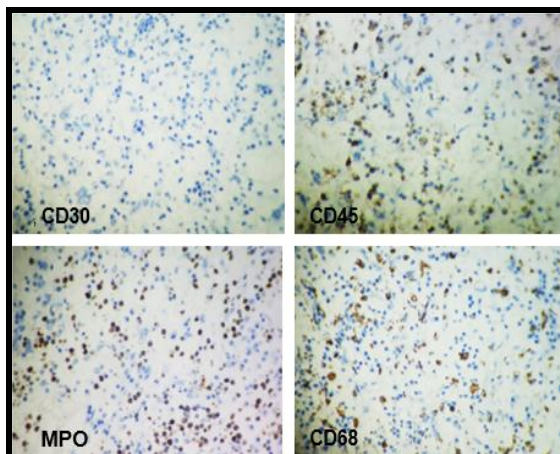


Figure 5: IHC showing negative reactivity to CD68, CD30, CD45, and MPO.

DISCUSSION

MFH is the commonest type of soft-tissue sarcomas, representing 28% of all sarcomas. It was first described in 1964 as ‘malignant fibrous xanthomas’ after an extensive analysis of 53 cases which

possessed histiocytic and fibroblastic elements with a storiform pattern, accompanied by inflammatory, xanthomatous, and pleomorphic giant cells. MFHs occur more frequently in the extremities, followed by the abdominal cavity and the retroperitoneum. These may also be found in any part of the body, including the digestive tract, spermatic cord, kidney, pancreas, spleen, lung, thymus, ovaries, and gallbladder. [5] Five histological subtypes of MFH have been described: pleomorphic storiform (65%), myxoid (15%), giant cell (10%), inflammatory (8%), and angiomatoid (2%). [2]

Inflammatory malignant fibrous histiocytoma/undifferentiated pleomorphic sarcoma inflammatory variant was first described over 30 years ago as ‘inflammatory fibrous histiocytoma’, due to the intense acute inflammatory infiltrate in the absence of known infectious agents and unassociated with tissue necrosis. It commonly affects the retroperitoneum and, less frequently, the extremities. However, due to the rarity of this entity, information about its presentation, clinical course, and outcome is very scarce. [4,8]

Patients with retroperitoneal tumors develop constitutional symptoms including anorexia, malaise, weight loss, and signs of increasing abdominal pressure. Occasionally, fever and leukocytosis with neutrophilia or eosinophilia dominate the clinical presentation of this disease. This symptom has been documented for the inflammatory type of malignant fibrous histiocytoma, although rarely it occurs with the other subtypes. [7]

The differential diagnosis consists of distinguishing this tumor from 1) non neoplastic xanthomatous /xanthogranulomatous lesions (absence of mitotic activity). 2) dedifferentiated liposarcoma (MDM2 positive) 3) lymphoma (neoplastic cells of IMFH display phagocytosis of neutrophils which is

typically not seen in lymphoma, also symptoms of fever and leukemoid reaction occasional characteristic of IMFH are rare with lymphoma). Importantly neoplastic cells in IMFH express vimentin but not various leukocyte lineage markers (CD15,CD20,CD45). To summarise, “vimentin only” immunoreactivity without any other specific or distinct expression of cell line markers can indicate a diagnosis of MFH. [7]

Imaging modalities are helpful in providing data about the size, location, vasculature, extension of the tumor, and in designing preoperatively the extent of the surgical excision. But definitive diagnosis can be made only by biopsy, which should be taken at multiple sites to avoid misdiagnosis. [5]

Surgical wide excision with tumor-free margins remains the principal modality for treatment adjuvant therapy, like Chemotherapy is not effective and radiotherapy is limited by toxicity to the adjacent organs. [4]

CONCLUSION

Inflammatory MFH represents a unique subtype of MFH, which is extremely rare. It has a poor prognosis with high recurrence and low survival rates. Inflammatory MFH should be included in the differential diagnosis in patients with a large retroperitoneal mass. The best therapeutic approach is an aggressive wide surgical resection of the tumor mass en bloc.

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