

A Rare Cause of Splenomegaly: Malignant Fibrous Histiocytoma

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Abstract: Here we report a 38 year old male who was evaluated for splenomegaly and diagnosed to have malignant fibrous histiocytoma of spleen, which was confirmed histopathologically. Very few cases of MFH have been reported.

Keywords: Malignant fibrous histiocytoma, spleen.

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is a soft tissue sarcoma mainly occurring in the soft tissues, especially in the extremities and trunk, and other less common sites include the retroperitoneum, and the head and neck. It is an aggressive malignancy with high potential of local recurrence and distant metastases, and surgery with radical resection of the primary tumor with negative histological margins is the treatment of choice even with recurrence or metastasis.

CASE REPORT

38 year old male presented with history of left sided abdominal pain, early satiety, tiredness for 1 month duration. He had no co morbid illness or addictions. On examination he had no pallor, icterus, clubbing, lymphadenopathy or edema. Moderate splenomegaly was present. Other systems were within normal limits.

INVESTIGATIONS



Fig-1: CHEST XRAY- Elevated left hemidiaphragm due to splenomegaly

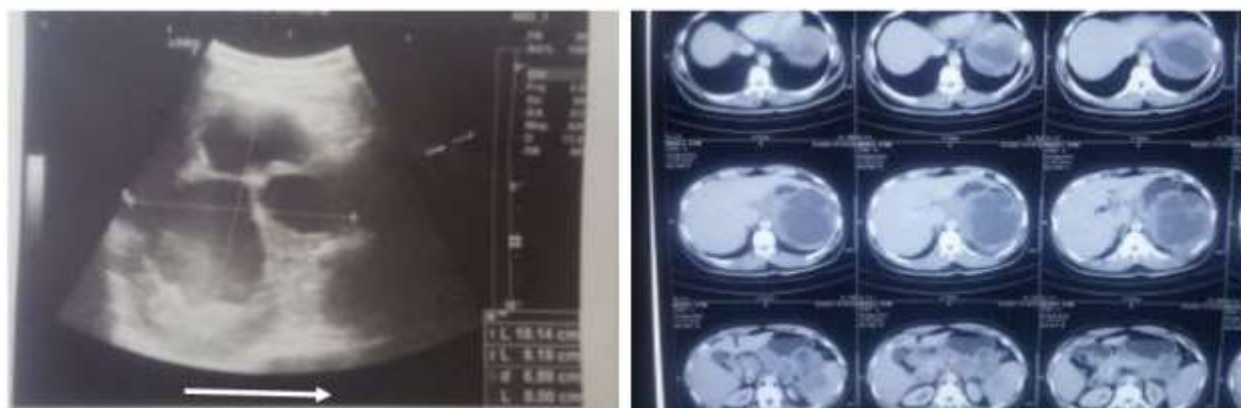


Fig-2: Usg Abdomen & Cect Thorax- Large multicystic lesion in spleen infiltrating postr wall of stomach

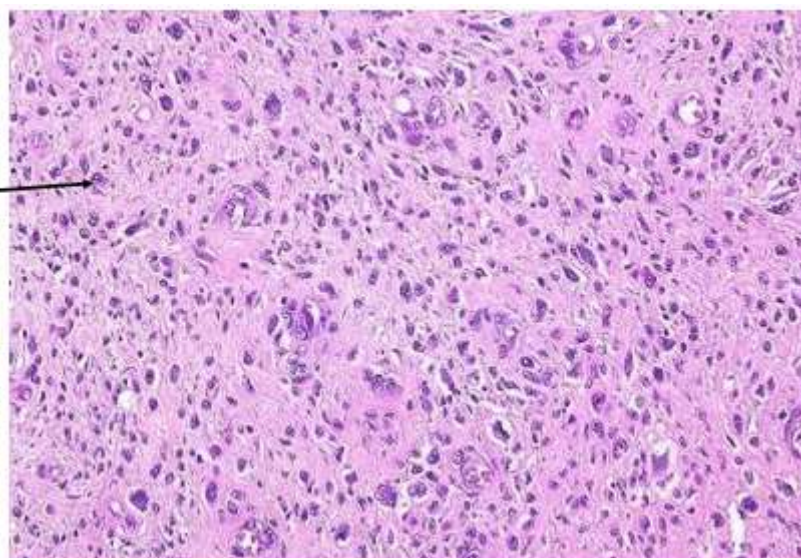


Fig-3: Histopathology of the lesion-showing pleomorphic spindle shaped cells in a storiform pattern

Table 1: Hematological Report

Hemoglobin	12 g/dl
Total count	11000/cmm
Platelet count	3.2 L/cmm
MCV	79 Fl
ESR	60 mm in 1 st hr
Blood urea/ serum creatinine	17 mg/dl/0.4mg/dl
Total protein/albumin	7 g/dl/4.2g/dl
SGPT/ALP	46/51 IU/L
Chest xray	Elevated left hemidiaphragm

USG abdomen revealed a large cystic lesion measuring 11x 10x 8 cm in the splenic parenchyma. CECT abdomen confirmed that the lesion is arising from spleen and is extending to posterior wall of stomach with abdominal lymphadenopathy without much contrast enhancement. Fnac from the lesion showed spindle shaped cells with plump vesicular nucleus and cytoplasm-possibility of malignant fibrous histiocytoma. Patient underwent splenectomy with gasrotojejunostomy. Histopathological examination revealed an infiltrating highly cellular lesion with pleomorphic spindle cells arranged in intersecting

fascicles and storiform pattern at places. Frequent mitoses and extensive areas of necrosis along with tumor giant cells and histiocyte like cells were also identified. Final diagnosis of storiform variant of malignant fibrous histiocytoma arising from spleen was made.

DISCUSSION

Primary MFH of the spleen is especially rare. The incidence of splenic MFH in all primary spleen neoplasms is difficult to evaluate because the cases are distinctly rare. According to the cases reported, splenic

MFH mainly occurs in the individuals aged from 40 to 60 years old, and there seems to be a slight male predominance [1]. There is no typical clinical symptom, but abdominal pain, splenomegaly, weight loss and fever were the prominent features in about 70% of the patients. Ultrasonography (USG) and computed tomography (CT) are the two most valuable imaging techniques. However, neither USG nor CT is specific for MFH. There are no suggestive imaging patterns that can be used for the diagnosis prior to surgery. These tumor cells consist of histiocyte-like and fibroblast-like cells with multinucleated giant cells [2]. There are positive stainings for lysozyme, vimentin, CD68, and alfa-1 antitrypsin.

MFH has been categorized into five types, based on the histopathologic subtype, including storiform-pleomorphic, myxoid, inflammatory, giant cell and angiomatoid variants [3]. However, only the subtypes of storiform, pleomorphic and inflammatory variants have been reported in the MFH of the spleen in the previously reported literature. Multinucleated giant cells, foam cells and inflammatory cells are also usually observed [4]. The accurate diagnosis of MFH depends on an accurate differential diagnosis from other sarcomas, observing karyomorphism and differential figures, and immunohistological staining.

In our case, we could not get a definite preoperative diagnosis of the splenic tumor according to the physical examination and CT. The final diagnosis was confirmed by the results of the histopathology and immunohistochemistry. Splenectomy is the treatment of choice, and the role of radiotherapy and chemotherapy as adjuvant therapeutic modality is not yet clear in retroperitoneal and visceral sarcomas [5].

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