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Is Retroperitoneal Malignant Fibrous Histiocytoma In Female Is Easy To Ruled Out?.

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ABSTRACT

Malignant fibrous histiocytoma (MFH) is the most common soft-tissue sarcoma of late adult life occurring predominantly in the extremities and the retro peritoneum. It is much less common in the female retro peritoneum, leading to diagnostic errors. The clinical, radiographic and CT signs are non-specific. A 55-year-old woman with lower abdominal pain was clinically diagnosed as pelvic mass of unknown origin .MRI shown a retroperitoneal mass. This tumour can only be diagnosed by histology. A storiform-pleomorphic type of MHF was diagnosed from histopathology. Chemotherapy was proposed as the postoperative treatment and the patient was now on her follow up. These lesions are relatively rare and consequently difficult to study. The authors report this rare case in south India.

Keywords: Pain, mass, malignant fibrous histiocytoma, poor prognosis.

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INTRODUCTION

The first described malignant fibrous histiocytoma (MFH) case in 1963[1]. It is a soft-tissue sarcoma of undifferentiated mesenchymal cell origin. It commonly occurs in the 5th and 6th decades, with a 2: 1 male-to-female predominance. After the extremities, the retroperitoneal space is the second most common site of this tumor[2, 3]. Because the radiological findings of MFH are nonspecific, an accurate diagnosis of retroperitoneal MFH is difficult. We report a rare case of retroperitoneal MFH that was found in south India

CASE REPORT

A 55 years old Nulliparous woman presented to gyneac outpatient department with complaints of lower abdominal pain for 3 months. Patient was on antidiabetic drugs, antihypertensive and lipids lowering drugs. Patient attained her menopause 5 years before. Her per abdomen examination revealed irregularly enlarged mass arising from pelvis of about 24 weeks size, mixed in consistency, mobile but lower edge of the mass could not be made out. Bimanual pelvic examination revealed a mass arising from pelvis of approximately 24 weeks and anterior to it is a cystic, bilobed mass of variable consistency, with mobility but uterus could not felt separately .In per rectal examination a mass was felt with free mucosa. The investigations were done and given in the tabulation (table 1). The patient undergone exploratory laparotomy. The intra operative findings shown (figure 2) a large multilobulated lesion of 15 x 15cms, partly solid and partly cystic with multiple septations with smooth outer surface found behind the posterior wall of the urinary bladder and in front of the uterus arising from the retro peritoneum. The mass was highly vascularized with infiltration into the posterior wall of the bladder which bleeds on touch. Capsule was intact all over the mass except anteriorly were it was adherent (? infiltrated) to bladder. Ureters were well identified and were away from the mass.

Table 1: Investigations and Report

Parameters	Values/remark
Blood	
Group	O positive
Haemoglobin (Hb) gm/dl	10.8
Renal function test	WNL
Lipid profile	WNL
Electrolytes	WNL
Thyroid function test	WNL
Random blood Glucose(RBS) in mg	102
Glycated haemoglobin(HbA1c)in %	5.48
Viral markers(hepatitis/HIV)	NR
Ca125 marker	20.83u/ml
Urine	
Routine	WNL
USG ABDOMEN	NORMAL STUDY
USG PELVIS	B/L ovarian mass, with left larger than right Large soft tissue mass lesion or Leiomyoma with cystic degeneration arising from elsewhere
MRI ABDOMEN (figure 2)	Large heterogeneous lobulated mass lesion with solid and cystic component in the pelvis compressing and displacing the uterus posteriorly and to the left side. The lesion is also seen abutting the left ovary. Right ovary not seen separately- ovarian mass to be considered. Fibroid with calcification arising from left lateral wall of superior aspect of uterus. Uterus approximately 10x 1.2cm; endometrial thickness – 2.8 mm. Cervix and vagina - normal visualized bony structures - normal Ischiorectal fossa regions and underlying soft tissues – normal. Both ureters seen coursing posterior to the mass lesion No significant Para aortic adenopathy
PAP SMEAR	Could not be done as the cervix is not seen.

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The mass excised (debulking) with part of the capsule which got infiltrated. From the cystic areas fluid was collected and sent for analysis. Smear shows scattered spindle shaped cells with pleomorphic nuclei, multinucleated giant cells, tumor giant cells and also scattered oval to polygonal cells with hyper chromatic nuclei admixed with inflammatory cells in a hemorrhagic background with positive for malignancy. The histopathology of the section from tumor mass shows a cellular spindle cell neoplasm composed of pleomorphic spindle cells with hyper chromatic nuclei arranged in interlacing bundles, fascicles and focal storiform pattern. Extensive areas of necrosis, hyalinization, myxoid degeneration, cartilaginous metaplasia and hemorrhage are seen. Pleomorphic tumor giant cells, multinucleated cells, bizarre giant nuclei and mitotic figures are seen giving an impression of malignant fibrous histiocytoma of storiform pleomorphic type. Patient was then put on 1 course of chemotherapy with Adriamycin and cyclophosphamide given on 28 September with next cycle planned on 18 October 2014.

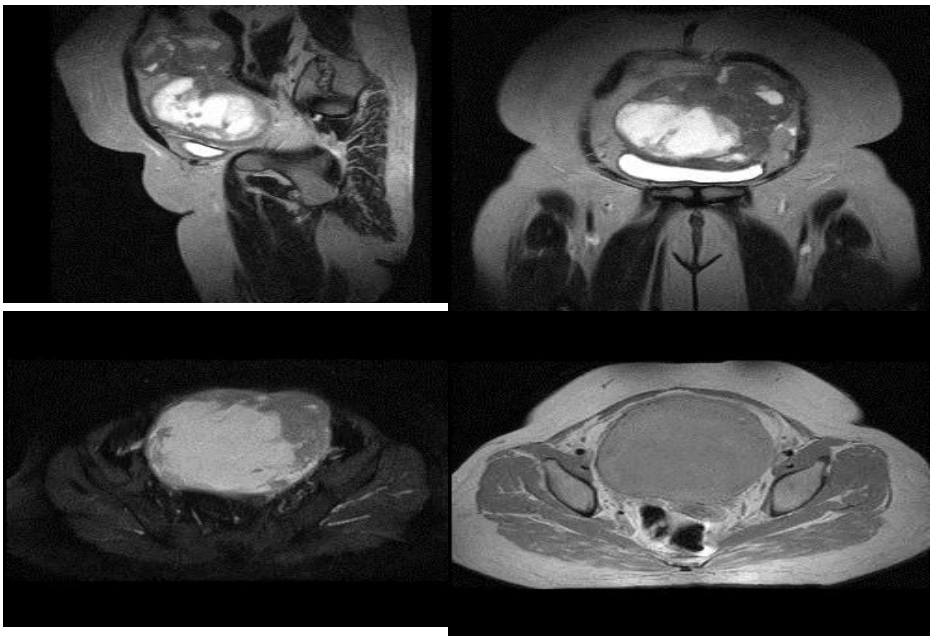

Figure 1: Magnetic Resonance Imaging of Abdomen

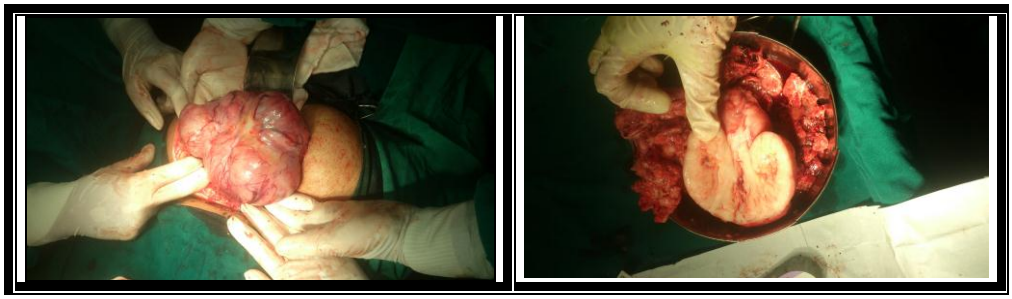

Figure 2: Intra-operative image of the tumor.

DISCUSSION

Malignant fibrous histiocytoma usually presents as a painless mass with increased intra-abdominal pressure. MFH originates from undifferentiated mesenchymal cells[2] and can be a complication of radiation exposure, trauma, various surgical incisions or varying degree burn scars, and malignancies like Hodgkin's lymphoma, multiple melanoma, and malignant histiocytosis[2].Malignant fibrous histiocytoma are tumours with a mixed structure, containing fibroblasts and histiocyte-like cells. The tumor tissue may also comprise tumour giant cells, inflammatory cells, and lipid laden xanthoma cells. The tumor may be also in many occasions interspersed with myxoid substance and elastic fibers[4]. Around five histological subtypes of MFH have been described and as follows: pleomorphic storiform (65%), myxoid (15%), giant cell (10%), inflammatory (8%), and angiomatoid (2%). MFH is most frequent in those older age group and it rarely occurs under the age of 40[4].The

prognosis of MFH is related to the histological malignity grade, the tumor size, depth, location and extension [2]. Retroperitoneal MFHs are very rare tumours that are difficult to diagnose preoperatively. The radiological findings are usually not diagnostic. Although magnetic resonance imaging (MRI) can demonstrate important characteristics of these tumours, the diagnosis is often challenging. Whenever there is no definite sign that suggests the organ of origin, the diagnosis of primary retroperitoneal tumor is likely [5]. Here MRI shows MFH as a large, lobulated, soft-tissue mass, with attenuation similar to muscles. Frequently, areas of decreased attenuation are apparent more centrally within the mass, corresponding to myxoid regions, haemorrhage, or necrosis. Haemorrhagic components are common in soft tissue MFH. Fluid-fluid levels may be seen on MRI after haemorrhage [3]. Ultra Sonography reveals a hypo echoic solid mass. This radiological finding is certainly nonspecific. In our case, MRI and USG showed a large, well-defined, soft-tissue mass with areas of decreased attenuation and focal and diffuse calcification. It is only the histology which confirms the tumour and the patient was referred to higher center for management. Outside it was diagnosed as recurrent mass lesion and the patient was given 1 course of chemotherapy with Adriamycin and cyclophosphamide given on 28 September. Next cycle is planned on 18th October.In conclusion, retroperitoneal MFHs are rare tumors that are difficult to diagnose preoperatively. The radiologic findings are usually nonspecific and are not diagnostic. The treatment of choice is complete radical surgical excision, followed by radiotherapy or hypostatic chemotherapy.

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