CASE REPORT

# Pleomorphic Undifferentiated Sarcoma of The Mesentery Presenting as Intestinal Obstruction: A Case Report

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## ABSTRACT

**Introduction:** The affection of abdomen by Pleomorphic Undifferentiated Sarcoma (PUS) , previously known as Malignant Fibrous Histiocytoma (MFH) is very rare. It is most commonly seen in extremeties and retropeitoneum with only a few cases of mesenteric MFH reported till date.

**Case Report:** An elderly female presented as sub-acute intestinal obstruction in the emergency department. Abdominal examination revealed slightly distended abdomen with raised bowel sounds. Contrast enhanced CT scan revealed malignant thickening of the ileal loops. Laparotomy was done and an exophytic growth was present in the terminal ileum adhered to the urinary bladder and omentum. Resection of ileal segment was carried out and subsequent histopathology suggested mesenteric MFH.

**Conclusion:** PUS of mesentery is a rare and highly malignant tumour with an unclear pathogenesis. No standard treatment protocols are available for management. Despite of its rarity, cases are reported and so there is a need to consider mesenteric MFH as a rare but a definite cause of intestinal obstruction in elderly patients.

**Key words**: Pleomorphic Undifferentiated Sarcoma, mesenteric, intestinal obstruction

**How to cite this article:** Agarwal A, Garg C, Agarwal M. Pleomorphic Undifferentiated Sarcoma Of The Mesentery Presenting As Intestinal Obstruction: A Case Report. Int J Cont Med Res. 2014;1(2):79-82

**Source of Support: Nil**

**Conflict of Interest: None**

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## INTRODUCTION

Malignant fibrous histiocytoma (MFH), now better known as pleomorphic undifferentiated sarcoma (PUS) was first described in 1963 by Ozello et al as malignant histiocytoma and fibrous xanthoma.[1] Later, in 1964 the origin of the tumor was ascertained to be fibroblastic as well as histiocytic.[2] Since, the origin of MFH has been a topic of debate, recently. It is now regarded as an pleomorphic undifferentiated sarcoma (PUS) and essentially represents a diagnosis of exclusion.[3]

Five histological subtypes of MFH have been described. These are pleomorphic storiform, myxoid, giant cell, inflammatory, and angiomatoid. The most common are pleomorphic storiform and myxoid types. The first two subtypes tend to be high-grade neoplasms, while the others are usually low-grade sarcomas. The karyotypic abnormalities in MFH are usually complex, with multiple numerical and structural rearrangements. The pathogenesis of MFH has not been clarified to date. However, it has been recognized as a complication of radiation, resulting from chronic postoperative repair, trauma, surgical incisions or burn scars.[4]

MFH occurs late in adult life, mostly around 6$^{th}$ to 7$^{th}$ decades. Males are more frequently affected than females. Lower extremities (49%) followed by upper extremities (19%), retroperitoneum (16%) and peritoneal cavity (5% -10%) are the frequent sites involved.[5] Visceral organs and the gastrointestinal tract are very rarely involved.

There are only meagre case reports describing MFH in these locations.[5]

MFH of mesentery presenting as intestinal obstruction is not reported till date.[6] Here we report a case of Mesenteric PUS presenting as subacute intestinal obstruction highlighting the need to consider this albeit rare diagnosis in the mind of surgeons while operating for intestinal obstruction in patients in their late stages of life.

## CASE PRESENTATION

A 50 year elderly female presented with complaints of on and off constipation for 2 months and pain abdomen with non passage of stools for 2 days. Patient gave history of frequent non bilious vomiting during this period. There was no history of weight loss, evening rise of temperature or passage of blood in stools. General physical examination revealed moderate pallor. Abdominal examination revealed slightly distended abdomen which was non tender but with raised bowel sounds. Hence, a clinical diagnosis of subacute intestinal obstruction was suggested.

Routine hematological investigations were within normal limits except for haemoglobin level which was 7.1 gm%. Ultrasound Abdomen showed circumferential short segment thickening of bowel loops (3.4 cm X 3.9 cm) in lower and central abdomen with loss of mural stratification and a large nodal mass adjacent to it with a cystic lesion in the region of posterior wall of urinary bladder. An impression of malignant bowel thickening was made.

Contrast enhanced CT scan abdomen showed short segmental necrotic thickening of ileal loops with necrotic exophytic growth of 6.4 cm X 4 cm and mild luminal compromise associated with dilated bowel loops, which were in close relation to anterior abdominal wall and urinary bladder along with invasion of fat planes. No evidence of peritoneal or retroperitoneal lymphadenopathy was seen. A final Impression of malignant thickening of ileal loops was thus, ascertained.

Exploratory laparotomy was done and per-operatively greater omentum was found adhered in pelvis to ileal loops. An exophytic growth was present in the terminal ileum 20 X 12 X 5 cm in size that was adhered to the urinary bladder with omentum adhered over it (Figure 1). A metastatic focus 1 X 1 cm in size was present 6 feet proximal to ileo-caecal junction (Figure 2).

Wide excision of the ileal growth with bladder repair with omentectomy with lymphadenecto-my with end ileostomy with mucous fistula was done and sample was sent for histopathology.

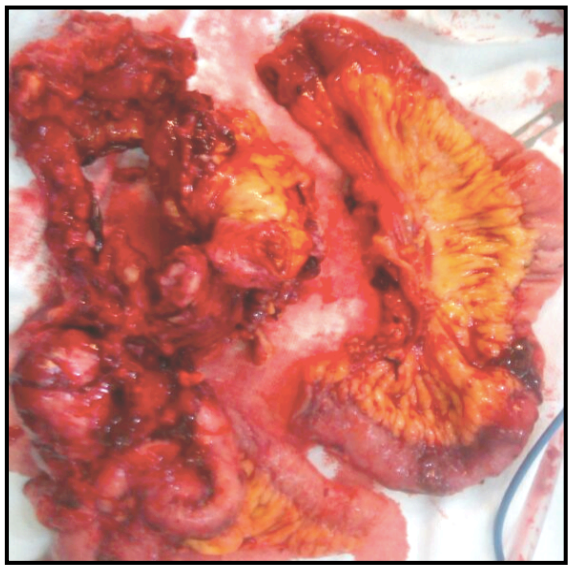

**Figure 1:** Shows resected specimen with exophytic growth in the mesentery of ileum.

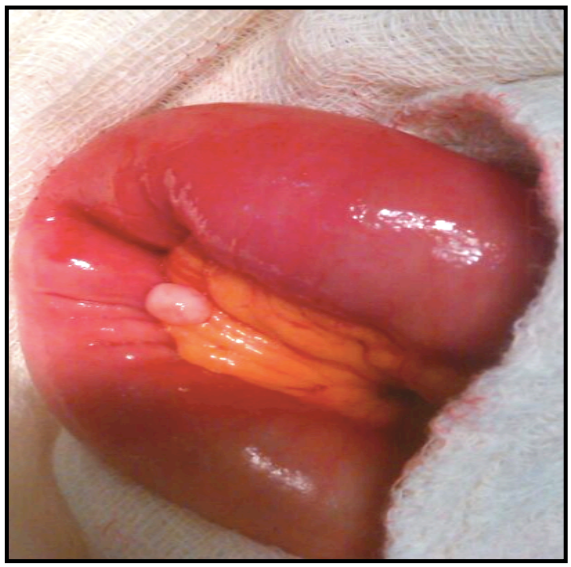

**Figure 2:** Shows a 1X1 cm creamish white metastatic focus which was 6 feet from the ileocaecal junction.

Grossly, on histopathological examination the specimen showed part of ileum with a

circumferential tumour attached to its serosal surface with omentum adhered to it. Microscopically sections under hematoxylin and eosin staining showed pleomorphic malignant cells arranged in a storiform pattern with giant cells with several histiocytes. Omentum showed metastasis with infiltration by tumour cells. A confirmatory diagnosis of mesenteric malignant fibrous histiocytoma (Pleomorphic storiform type) was thus, made (Figure 3). Postoperative period remained uneventful and patient is kept on follow-up without evidence of recurrence for the last 6 months.

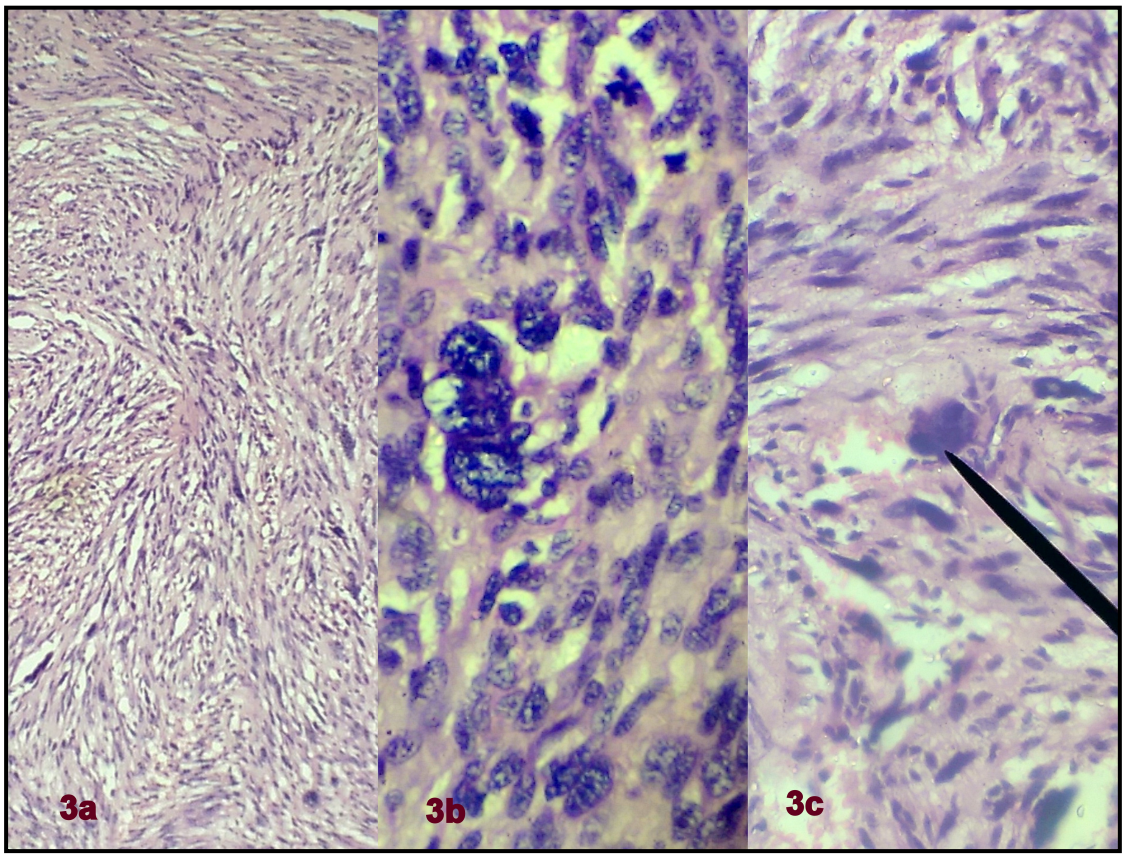


Figure 3: a) Shows storiform pattern of atypical spindle shaped cells (H&E X 40), b) Shows tumor giant cells and marked nuclear pleomorphism (H&E X 400), c) Shows bizarre hyperchromatic nucleus (H&E X 400).

## DISCUSSION

MFH accounts for 20-24% of soft-tissue sarcomas, making it the most common soft tissue sarcoma occurring in late adult life. The combination of infrequent occurrence, varied pathologic features, uncertain histogenesis, numerous subtypes and the many potential sites of presentation makes these tumors a challenge for the diagnostician, surgeon and oncologist. The origin of MFH has been a topic of debate. It is now regarded as an undifferentiated pleomorphic sarcoma and essentially represents a diagnosis of exclusion.[3]

Extremeties followed by the retroperitoneum are amongst the most frequent locations. Origin is typically from either the deep fascia or skeletal muscles of the region. Cases of MFH have been reported in the past to be located in the pulmonary region, hepatic, renal, bladder, scrotal, vas deferens, heart, aorta, GIT, orbit, CNS, facial sinuses, nasal cavity, oral cavity, nasopharynx, and soft tissues of the neck.[7]

Very few cases of abdominal MFH have been reported, from liver, omentum, ileum, psoas muscle, kidney, spleen, mesocolon, urachus and especially mesentery.[5] No more than 30 cases have been reported in literature so far.[8]

Basso and Pisanelli [9] have reported two such cases in the past. These patients presented with

abdominal pain and lump, unlike our case where there was no palpable lump abdomen. Diagnosis was confirmed after exploratory laparotomy followed by histopathological study of resected specimens. One of the patient though had a recurrent disease on a 2 year follow up, which has not been reprted with our case so far.

Nakayama et al [10] described a case of MFH of mesorectum. Patient presented with lower abdominal tumor that had been gradually increasing in size, unlike our case which presented as acute abdomen. Clinical examination revealed a firm, irregular, fixed, painless, child-head-sized tumor located in her lower abdomen. Computed tomography (CT) and magnetic resonance imaging (MRI) of the abdomen revealed a polycystic tumor at the lower abdomen which was 15 × 13 × 11 cm in diameter and encased the colorectum to the left back side. The tumor in our case was also circumferentially encasing the ileum from the serosal surface. The tumour was excised by low anterior resection and diagnosis of MFH was confirmed by histopathology.

Thus, to our knowledge MFH of mesentery presenting as intestinal obstruction has not been reported till date.[6] and we emphasise the need to consider this diagnosis for patients of intestinal obstruction especially, in their late stages of life.

## CONCLUSION

MFH in mesentery is rare, but cases are being reported in literature. No standard guidelines are available for the surgeons, radiotherapists and medical oncologists for the treatment of this disease despite the advancements in these respective fields. The current modality of choice being, surgical resection followed by radiotherapy in some cases. Studies are also needed to evaluate the pathogenesis of this disease so that in future it could be prevented or diagnosed early to reduce the mortality of this highly metastatic diseases.

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