

## Primary malignant fibrous histiocytoma of the mesocolon transversum

G. COCCHIARA, M. ROMANO, G. BUSCEMI, A.I. LO MONTE, A.M. FLORENA,  
T.V. BARTOLOTTA<sup>2</sup>, A. TAIBBI<sup>2</sup>, A. DI BERNARDO<sup>1</sup>, P. DI PAOLA,  
E. MARRAZZO, A. AGRUSA, G. ROMANO

**SUMMARY:** Primary malignant fibrous histiocytoma of the mesocolon transversum.

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*Primary malignant fibrous histiocytoma (MFH) is a rare and biologically aggressive tumor. Differential diagnosis must include cystic neoplasms of the pancreas (mucinous and serous cystoadenoma or cystocarcinoma), cystic lymphangioma or mesothelioma and retroperitoneal haematoma.*

*Making the right diagnosis preoperatively may be often difficult, but MFH may be suspected when a huge retroperitoneal mass is found at imaging in elderly patients.*

*We report a case of a primary abdominal MFH originating from the mesocolon transversum.*

**RIASSUNTO:** Fibroistiocitoma maligno primitivo del mesocolon trasverso.

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*Il fibroistiocitoma primitivo maligno (FIM) è una rara neoplasia. La diagnosi differenziale include neoplasie cistiche del pancreas (cistoadenoma sieroso o mucinoso, cistoadenocarcinoma), linfangioma cistico, mesotelioma ed ematoma retroperitoneale.*

*Una corretta analisi pre-operatoria è spesso difficile ma il FIM può essere sospettato quando una voluminosa massa retroperitoneale è riscontrata in pazienti di età avanzata tramite diagnostica per immagini.*

*Viene qui descritto un caso di FIM ad origine dal mesocolon trasverso.*

**KEY WORDS:** Malignant fibrous histiocytoma - Peritoneum - Mesocolon.  
Fibroistiocitoma maligno - Peritoneo - Mesocolon.

### Introduction

O'Brien and Stout firstly described Malignant fibrous histiocytoma (MFH) in 1964 (1). MFH usually arises from soft tissue of the extremities (more frequently in the lower limb), the trunk and retroperitoneum (2). Rarely, MFH may arise from abdominal or intestinal structures, with about 40 cases described in literature (3-7).

We report a case of a primary abdominal malignant fibrous histiocytoma originating from the mesocolon transversum.

### Case report

A 79-year-old female patient was referred to the Department of General Surgery of Uremic and Organ Transplantation with a non tender, rubbery, palpable mass in the epigastrium and right hypochondrium and normal skin, dyspepsia for one month. Laboratory tests result normal except for haemoglobin (8.2 g/dL), red blood cells (3.06x10<sup>9</sup>/L), sideraemia (22 µg/mL) and serum ferritin (579 ng/mL).

Ultrasonography showed, 1 cm under skin plane, a huge mass at least 12 cm x 7 cm sized with mixed echotexture and poorly defined borders, strictly close to the left hepatic lobe (Fig. 1). Endoscopy revealed compression of the posterior wall of the gastric antrum. A Computed Tomography (CT) scan of the abdomen showed a huge and unomogeneously hypodense mass (15 cm) without calcification. The mass showed unomogeneous contrast-enhancement, after the injection of iodinated contrast agent, and was suspected originating from the pancreas and infiltrating the left lobe of the liver. Magnetic Resonance Imaging (MRI) confirmed the presence of a 15x11x6.7 cm retroperitoneal mass compressing and dislocating the posterior gastric wall and depicted the presence of septa within the

University of Palermo, Italy  
Department of General Surgery of Uremic and Organ Transplantation  
<sup>1</sup> Department of Human Pathology  
<sup>2</sup> Department of Radiology

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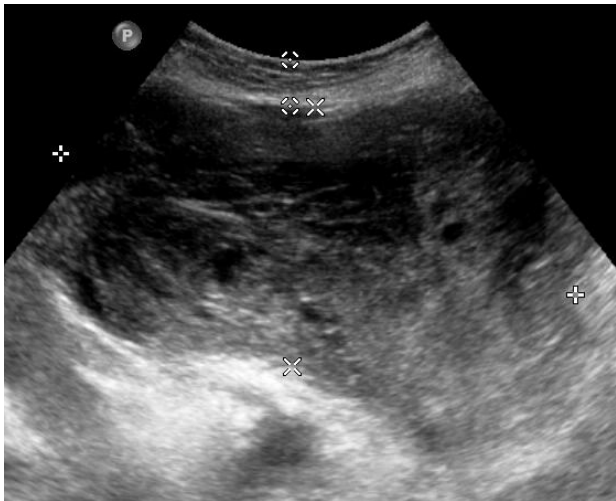


Fig. 1 - Ultrasound examination shows a large mass with mixed echotexture and poorly defined borders.

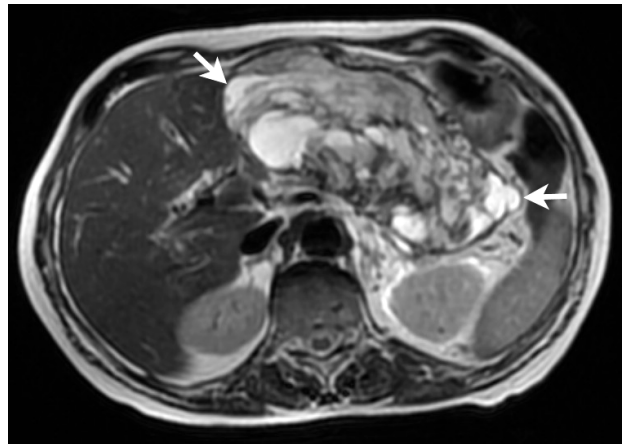


Fig. 2 - T2-weighted FSE MR image depicts a huge multiloculated abdominal tumour with unomogeneous and high signal intensity, bounded by thin hypointense capsule (arrows).

lesion, which also showed a thin but incomplete wall (Fig. 2). The mass was markedly unomogeneous either before and after the injection of a Gadolinium-based contrast agent, with faint peripheral contrast-enhancement. Septa also showed contrast-enhancement, but the lesion was substantially hypovascular (Fig. 3). Superior mesenteric artery was patent but dislocated laterally. No dilatation of intrahepatic and extrahepatic bile ducts or Wirsung was appreciable. The mass did not show clear cleavage planes from the pancreas and a mucin producing pancreatic tumor was first supposed.

At surgery, a 15 cm sized mass, strictly adherent to the posterior wall of gastric antrum, body and tail of the pancreas and mesocolon transversum, was detached from the stomach and a left pancreatectomy with sparing of the spleen was performed.

Macroscopic evaluation reported a 14x14x6 cm tawny yellow crumbly mass with a smooth capsule; at cut section a dense tissue with large haemorrhagic areas, often cystic, was observed. Microscopic evaluation showed a mixed cellular pattern, consisting of pleomorphic cell elements presenting high grade nuclear atypia, large and pleomorphic nuclei, mitotic index 8/10 High Power Field (HPF), multinuclear giant cells (osteoclastic-like cells) and spindle-shaped cells.

The immunochemistry profile of tumor cells showed positivity for vimentin and CD68 (PGM1), and negativity for S100 protein, cytokeratin, desmin and actin (Fig. 4a). These findings were consistent with a pathological diagnosis of malignant fibrous histiocytoma (giant cell type) (Fig. 4b). Chronic pancreatitis was also pathologically diagnosed.

Patient underwent a six month chemotherapy with gemcitabine. Positron Emission Tomography (PET) examination (88 MBq of <sup>18</sup>F-FDG), performed four months after surgery, revealed accumulation of the radio-tracer in VII segment of the liver and in the epigastrium consistent with still viable tumoral tissue as already showed by CT follow-up. Twenty-one months after surgery the patient is under clinical follow-up.

## Discussion

MFH usually occurs in the seventh decade of life presenting as painless mass originating from fibroblasts of soft tissues (5, 6). Histologically, five subtypes of MHH

are described: pleomorphic, storiform, myxoid, giant cell, inflammatory and angiomatoid. The pleomorphic and myxoid subtypes are more frequent and show a higher grade of malignancy when compared with the other subtypes.

Primary abdominal or intestinal MHF are quite rare and clinical symptoms are usually not specific and occur late despite MHF size may range from 2 up to 20 cm (3-6). Abdominal pain, dyspepsia, fever, diarrhea, weight loss and, ultimately, a palpable abdominal mass



Fig. 3 - At contrast-enhanced GRE T1-weighted image the mass shows a moderate and heterogeneous uptake of contrast medium mainly at the periphery and within the septa (arrow) and dislocates the stomach on the left (arrowheads).

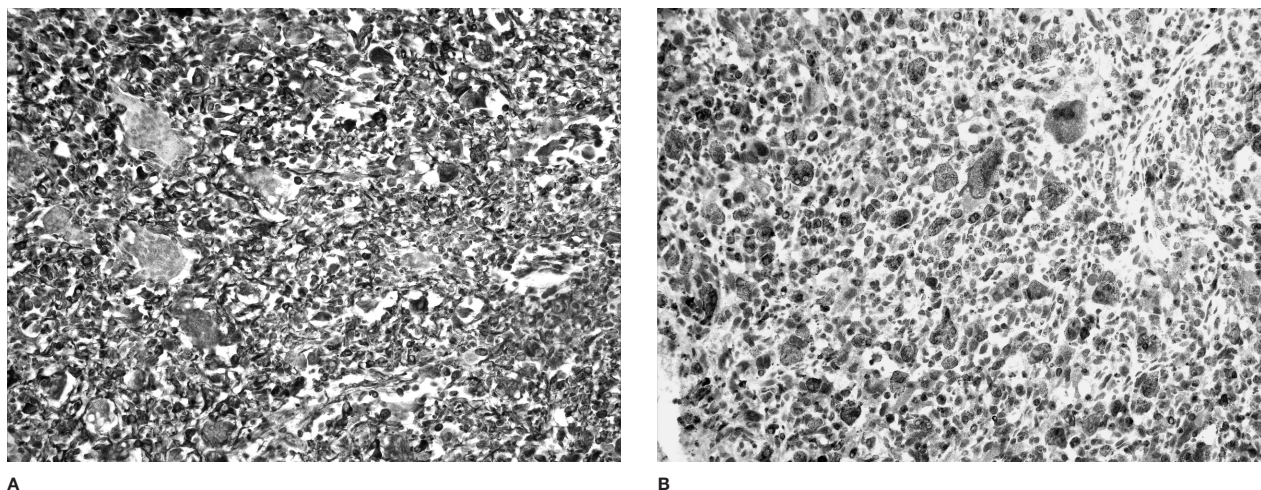


Fig. 4 - A) Vimentin immunohistochemistry staining shows a diffuse and intense positivity of the majority of the neoplastic cells, while giant multinucleated cells prove negative (magnification 200x). B) The giant multinuclear cell component of the tumor stains positive for CD68 immunohistochemistry reaction (magnification 200x).

are often described. Our patient had a huge palpable abdominal mass dislocating adjacent organs, such as stomach and liver. The patient also complained of anorexia and dyspepsia. A microcytic hypochromic anemia with increased serum level of ferritin was also present.

MHF are difficult to diagnose and do not show specific imaging findings (8). According to literature data and our experience, ultrasound examination may show a hypoechoic, mixed (because of tumoral necrosis) or anechoic pattern with internal hyperechoic septa (9, 10). CT usually depicts MFH either as well or ill defined mass, with attenuation values similar or slightly lower than normal muscle. Hypodense areas within the mass may be appreciable owing to necrosis or haemorrhagic infarction (9-12). In about 16% of abdominal MFH, especially pleomorphic, CT may demonstrate the presence of peripheral calcifications (12). MHF may invade adjacent organs, such as bowel, pancreas and kidney, making difficult to ascertain the precise site from which the mass originates (11).

In our case neither US nor contrast-enhanced CT was able to make a precise diagnosis. MRI concluded for a suspected cystic neoplasm of the pancreas. Cystic

appearance of MFH has been reported, sometimes because of bleeding within the lesion (11, 13-16). Hence, anemia may occur, as in our patient, who had microcytic hypochromic anemia with increased serum level of ferritin. All imaging modalities showed a close relationship between the mass and the left hepatic lobe, but laparotomy ruled out any invasion of the liver. Histology with immunohistochemistry demonstrated a primary abdominal malignant fibrous histiocytoma originating from the mesocolon transversum.

## Conclusion

Primary abdominal MFHs are rare and biologically aggressive malignancies. Differential diagnosis must include cystic neoplasms of the pancreas (mucinous and serous cystoadenoma or cystocarcinoma), cystic lymphangioma or mesothelioma and retroperitoneal haematoma. Making the right diagnosis preoperatively may be often difficult, but MFH may be suspected when a huge retroperitoneal mass is found at imaging in elderly patients.

## References

- O'Brien JE, Stout AP: Malignant fibrous xanthomas. *Cancer* 1964, 17:1445-1455.
- Enzinger FM, Weiss SW: Malignant fibrohistiocytic tumors. In: *Soft tissue tumors*. Mosby-Year Book Inc., 351-380, St Louis, MO. 1995.
- Udaka T, Suzuki Y, Kimura H, Miyashita K, Suwaki T, Yoshino T: Primary malignant fibrous histiocytoma of the ascending colon: report of case. *Surgery Today* 1999, 29:160-164.
- Singh DR, Aryya NC, Sahi UP, Shukla VK: Malignant fibrous histiocytoma of the rectum. *Eur J Surg Oncol*, 1999, 25:447-448.
- Okubo H, Ozeki K, Tanaka T, Matsuo T, Mochinaga N: Primary malignant fibrous histiocytoma of the ascending colon: report of a case. *Surg Today* 2005, 35: 323-327.
- Hiraoka N, Mukai M, Suzuki M, Maeda K, Nakajima K, Hashimoto M et al: Malignant fibrous histiocytoma of the cecum: report of a case and review of the literature. *Pathos Int* 1997,

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- 47: 718-724.
7. Hasegawa S, Kawachi H, Kurosawa, Obi Y, Yamanaka K, Nakamura K et al: Malignant fibrous histiocytoma in the ileum associated with intussusception. *Dig Dis Sci* 2004, 49: 1156-1160.
  8. Huang CC, Ko SF, Ng SH, Lee TY, Wan YL, Lin JW et al: Cystic malignant fibrous histiocytoma of the gastrocolic ligament. *Br J Radiol* 2001, 74: 651-653.
  9. Ros PR, Viamonte M Jr, Rywlin AM: Malignant fibrous histiocytoma; mesenchymal tumor of ubiquitous origin. *AJR* 1984, 142: 753-759.
  10. Bruneton JN, Drouillard J, Rogopoulos A, Laurent F, Normand F, Balu-Maestro C et al: Extraretroperitoneal abdominal malignant fibrous histiocytoma. *Gastrointest Radiol* 1988, 13:299-305.
  11. Goldman SM, Hartman DS, Weiss SW: The varied radiographic manifestations of retroperitoneal malignant fibrous histiocytoma revealed through 27 cases. *J Urol* 1986, 135: 33-38.
  12. Ko SF, Wan YL, Lee TY, Ng SH, Lin JW, Chen WJ: CT features of calcifications in abdominal malignant fibrous histiocytoma. *Clin Imaging* 1998, 22: 408-413.
  13. Weiss SW, Enzinger FM. Malignant fibrous histiocytoma: an analysis of 200 cases. *Cancer* 1978, 41: 2250-2266.
  14. Castillo M, Davis PC, Takei YD, Schwartzberg DG, Hoffman JC Jr: Intracranial cystic malignant fibrous histiocytoma in a child: sonographic and CT findings. *Pediatr Radiol* 1990, 20: 194-195.
  15. Berry AD 3rd, Reintjes SL, Kepes JJ. Intracranial malignant fibrous histiocytoma with abscess-like tumor necrosis. *J Neurosurg* 1988, 69:780-784.
  16. Panicek DM, Casper ES, Brennan MF, Hajdu SI, Heelan RT. Hemorrhage simulating tumor growth in malignant fibrous histiocytoma at MR imaging. *Radiology* 1991, 181: 398-400.
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