DOI: 10.7860/JCDR/2025/79577.21393

Case Report

Radiology Section

Unravelling the Mystery of Mesenteric Fibromatosis: A Rare Case Report

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ABSTRACT

Mesenteric Fibromatosis (MF) is a proliferative fibroblastic lesion of the small intestinal mesentery. It constitutes 8% of all desmoid tumours, which represent 0.03% of all neoplasms. Although histologically benign, MF can invade locally and recur after excision. It occurs sporadically or in association with Familial Adenomatous Polyposis (FAP) mutation as a component of Gardner's syndrome. The presenting features of MF include an asymptomatic abdominal mass, abdominal discomfort or pain, bowel or ureteral obstruction, intestinal perforation, fistula, or functional impairment of ileoanal anastomosis following colectomy in FAP cases. We report the case of a 29-year-old male who presented with a swelling on the right side of the umbilicus for six months, associated with dull aching pain for two months. Fine Needle Aspiration Cytology (FNAC), Ultrasonography (USG), and Contrast Enhanced Computed Tomography (CECT) findings were inconclusive. Exploratory laparotomy revealed a mass measuring approximately 6×5×4 cm in the ileal mesentery, which was excised along with 20 cm of ileum. Histopathology confirmed the diagnosis of MF. Considering the rarity of this tumour and the diagnostic and therapeutic challenges associated with it, we present this case to add to the existing literature.

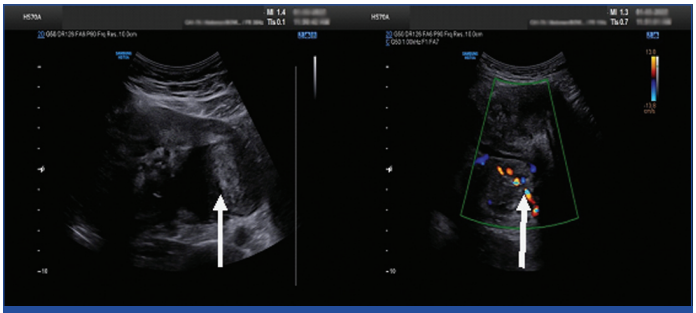
Keywords: Desmoid, Familial adenomatous polyposis, Neoplasms

CASE REPORT

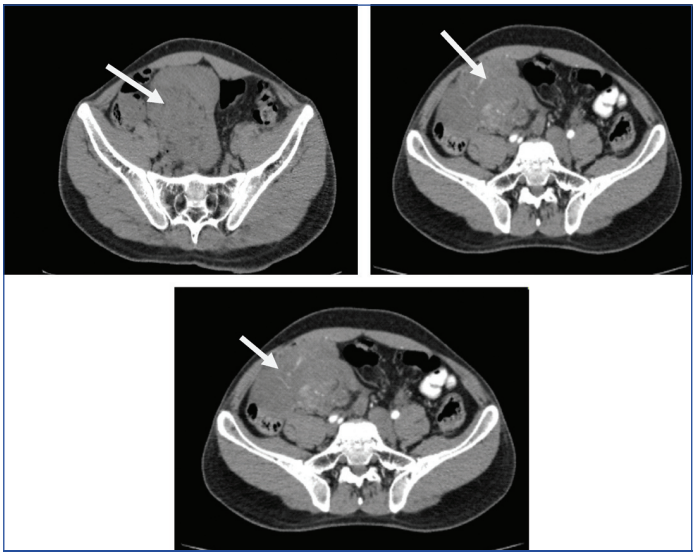
A 29-year-old man complained of a dull soreness that had been present for two months, along with swelling on the right-side of the umbilicus for six months. There were no additional symptoms, and the swelling was progressively getting larger. There was no concomitant fever or other abdominal or extra-abdominal symptoms. There was no personal history of abdominal trauma, Familial Adenomatous Polyposis (FAP), or colon cancer in the family. Examination of the umbilical region revealed a spherical intraperitoneal lump measuring approximately 5×4 cm. It had a smooth surface with distinct borders, and was firm, mobile, slightly tender, yet it lacked fixity on respiratory movement. Routine haematological and biochemical investigations were normal.

An intra-abdominal abscess or cyst was suggested by the abdominal ultrasonography [Table/Fig-1]. In a background of blood, Fine Needle Aspiration Cytology (FNAC) showed a few monolayered clusters of muscle fibres, mucin strings, and colonic mucosal cells; no atypical or malignant cells were observed. FNAC may not be able to distinguish spindle cell lesions such as Gastrointestinal Stromal Tumours (GISTs), MF, or leiomyomas. In some cases, FNAC findings may be inconclusive, necessitating further diagnostic procedures. Given these limitations, a Contrast Enhanced Computed Tomography (CECT) scan was performed to assess the lesion's size, location, and involvement of adjacent structures. An oval hypodense lesion (+25 HU), which was modestly enhancing (+46 HU), was visible in the intraperitoneal cavity with retroperitoneal extension on CECT, suggesting a GIST or leiomyoma [Table/Fig-2]. Upper gastrointestinal endoscopy and colonoscopy revealed no polyps.

During elective laparotomy, a 6×5×4 cm mass in the ileal mesentery was discovered. The mass and 20 cm of ileum were removed to achieve R0 margins, and two layers of an end-to-end anastomosis were performed. Histopathological analysis revealed uniform, slender spindle-shaped cells resembling myofibroblasts. These cells were arranged in long, sweeping fascicles within an abundant collagenous stroma. The nuclei were elongated, pale-stained, and vesicular with finely dispersed chromatin. There was no evidence of atypical mitotic figures. The stroma was rich in collagen, and the tumours had ectatic blood vessels with thin walls. Immunohistochemistry revealed diffuse positivity for smooth muscle actin and beta-catenin (both nuclear and paranuclear staining). The neoplastic cells tested negative for H-Caldesmon, CD117, CD34, S100 protein, and desmin. On the basis of morphological and immunohistochemical findings, the probable diagnosis was Deep Fibromatosis (DF) or Desmoid Type Fibromatosis (DTF) of conventional pattern.



[Table/Fig-1]: A fairly well-defined heterogenously hypointense mass lesion showing minimal internal vascularity on colour Doppler.



[Table/Fig-2]: A Contrast-Enhanced Computerised Tomography (CECT) showed an oval hypodense (+25 HU) and mildly enhancing (+46 HU) focal lesion in the intraperitoneal cavity with retroperitoneal extension.

After an uncomplicated surgical phase, the patient was discharged on the 10th day. Following surgery, adjuvant radiation therapy (50 Gy) was administered.

DISCUSSION

Less than 1% of retroperitoneal masses and less than 3% of all soft tissue tumours are desmoid tumours, sometimes referred to as DT or DTF. They are a rare subset of deep fibromatoses, making up fewer than 3% of all soft tissue tumours, 0.03% of all neoplasms, and less than 1% of retroperitoneal masses. Although they are locally aggressive, they do not have the ability to spread. A few cases of spontaneous cystic degeneration in DF have been reported, but they remain uncommon. Desmoid tumours in the abdominal region can develop intra-abdominally, with the small bowel mesentery being the most frequent location, or from the soft tissue of the deep abdominal wall [1-5]. Although they can occur at any age, they are most common in the third and fourth decades of life. Abdominal desmoids are more prevalent in females, especially those of reproductive age; however, they can affect both sexes [6].

The clinical indicators of desmoid tumours vary depending on the tumour's location and can include a wide spectrum of symptoms. They can range from minor, incidental lesions to aggressive, fast-growing abdominal masses that may prove fatal within a few years or months. The majority of patients present with an asymptomatic mass in the abdomen; however, some may also experience paresthesia, intestinal, vascular, ureteric, or neurological obstructive symptoms, or abdominal discomfort. The size of the tumour does not necessarily correlate with the level of pain. Acute abdominal discomfort, bleeding, or peritonitis due to bowel perforation can all result from mesenteric desmoid tumours [1].

FAP may be linked to desmoid tumours. The prevalence of desmoids in FAP is between 10% and 20%, which is almost 850 times higher than in the general population. Intra-abdominal desmoid tumours are one of the leading causes of death for individuals with FAP and a significant cause of morbidity. Factors such as trauma, particularly abdominal surgery like preventive colectomy, a family history of desmoids, hormonal variables, and the location of the Adenomatous Polyposis Coli (APC) germline mutation have all been associated with FAP-related desmoids. Desmoid tumours in FAP patients may be identified prior to, concurrently with, or subsequent to the FAP diagnosis [7].

For aggressive fibromatosis, Radiotherapy (RT) can be a useful treatment option. It can be employed as the sole treatment for unresectable tumours as well as for resected tumours that have local recurrences. In the past, radiotherapy was only applied to patients who refused surgery or in cases of unresectable fibromatosis. In more recent times, RT has been used as the primary therapeutic option for individuals encountering complications following surgery. Good long-term local control rates of 70-93% with RT were observed in a number of studies. In the meantime, there is not enough data to support adjuvant RT [8].

Because MF has a significant chance of local recurrence, even after total surgical resection, regular follow-up is essential to its management. Studies indicate that recurrences can happen months to years after therapy; in some cases, they have been reported as late as 117 months following surgery. Therefore, a systematic follow-up protocol is necessary to identify recurrences early and treat them promptly. According to guidelines, clinical evaluations and imaging tests, such as Magnetic Resonance Imaging (MRI) or Computed Tomography (CT) scans, should be performed every three to six months during the first two to three years after treatment, followed by annual assessments [9].

Bahlouli N et al., reported a similar case of a 38-year-old male who presented with an abdominal mass [1]. Imaging revealed two distinct tumours: a solid mass near the left lateral iliac pedicle and a large cystic mass within the abdominal cavity. An elective exploratory laparotomy was conducted to excise both masses, and histopathological analysis confirmed that both lesions were desmoid tumours. The patient exhibited an exceptionally rare cystic form of mesenteric desmoid tumour, accompanied by a solid inguinal mass. Imaging revealed a large intra-abdominal mass with both cystic and solid components, extending to the mesenteric root and displacing adjacent structures. Conversely, in our case, the patient presented with a firm, mobile mass near the umbilicus, initially suspected to be an intra-abdominal abscess or cyst. Imaging and FNAC were inconclusive, leading to an exploratory laparotomy that revealed a 6×5×4 cm mass in the ileal mesentery.

CONCLUSION(S)

Desmoid tumours are uncommon; however, they should be taken into account when making a differential diagnosis for intra-abdominal masses in middle-aged patients. It is crucial to adopt a multidisciplinary approach and conduct long-term follow-up to monitor recurrence. In cases where complete surgical excision is not feasible, radiotherapy should be considered as a treatment alternative.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Mar 31, 2025
- Manual Googling: May 19, 2025
- iThenticate Software: May 27, 2025 (19%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Mar 26, 2025**
Date of Peer Review: **Apr 23, 2025**
Date of Acceptance: **May 29, 2025**
Date of Publishing: **Aug 01, 2025**