Case Report

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# Giant Omental Fibromatosis Presenting as Pelvic Mass

Oncology Section

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## ABSTRACT

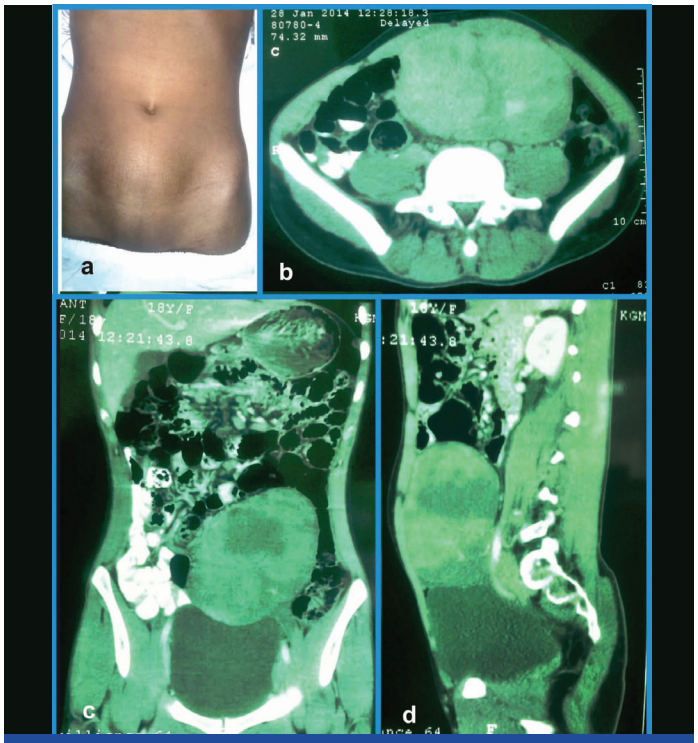
Omental fibromatosis (abdominal desmoids) is a rare benign but locally aggressive neoplasm characterized by mass like or infiltrative growth of fibrous tissue. It usually arises from the abdominal wall or the extremities, however rarely it may also arise in the omentum, ileocolic mesentery, transverse or sigmoid mesocolon and ligamentum teres. Here, we present an 18-year-old male, who presented with lower abdominal pain and palpable lump in hypogastric region. Computed tomography of the abdomen showed large heterogeneous mass in lower abdomen, possibly arising from mesentery with regional adenopathy. Patient underwent exploratory laparotomy with a preoperative diagnosis of mesenteric tumour possibly gastrointestinal stromal tumour (GIST). Histopathological examination revealed the lesion as omental fibromatosis. To the best of our knowledge, very few cases of omental fibromatosis are noted in literature. Here, we describe a rare case of giant omental fibromatosis which resembled mesenteric GIST clinically but finally diagnosed as fibromatosis by histomorphology and immunohistochemistry (IHC). The present article describes fibromatosis of greater omentum and the difficulty in preoperative diagnosis, as it is frequently misdiagnosed as GIST.

**Keywords:** Desmoids, Fibromatosis, Mesentery, Omentum

## CASE REPORT

An 18-year-old male patient was admitted with pain in lower abdomen and lump since one year. No other significant history noted. Abdominal examination revealed large mobile palpable lump in hypogastrium approximately 12 x 8cm in dimensions, with well defined borders [Table/Fig-1a]. Laboratory investigations and the tumour markers (AFP, LDH and Beta HCG) were within the normal range.

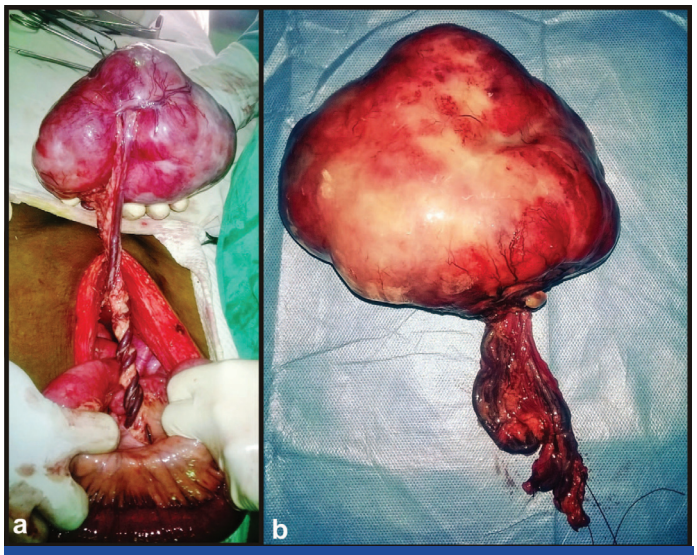
Ultrasonography (USG) and computed tomography (CT scan) showed well-defined lobulated mass in lower abdomen measuring 14 x 10cm, displacing adjacent structures and heterogeneous enhancement in solid part with non enhancing cystic areas on contrast study [Table/Fig-1b-d]. Fine needle aspiration of the lump revealed round, oval to spindle shaped cells with atypia suggestive of mesenchymal neoplasm. Patient underwent exploratory laparotomy with a preoperative diagnosis of mesenchymal tumour. Laparotomy revealed large encapsulated mass approximately 18 x 10 cm arising from greater omentum [Table/Fig-2a]. En bloc excision of mass with omentum was done. Gross examination showed globular soft tissue mass measuring 17 x 15 x 13 cm [Table/Fig-2b]. Cut surface of the specimen was fasiculated and pinkish gray in color [Table/Fig-3]. Microscopic examination revealed elongated and spindle shaped cells with minimal nuclear pleomorphism; collagenisation and perivascular inflammation [Table/Fig-4a,b]. Immunohistochemistry study showed tumour cells negative for CD 117 [Table/Fig-5a], DOG-1[Table/Fig-5b], smooth muscle actin (SMA) [Table/Fig-5c], S-100 and CD-34 [Table/Fig-5d]. Final diagnosis of fibromatosis was done. Postoperative course was uneventful and patient is disease free at 18 month of followup.



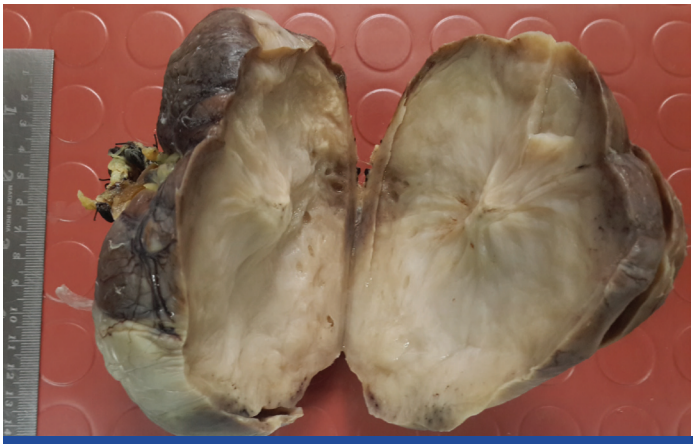

**[Table/Fig-1]:** a) Abdominal examination revealed palpable lump in pelvis. b-d) Computed tomography (CT scan) showed well-defined lobulated, heterogenous mass in lower abdomen

## DISCUSSION

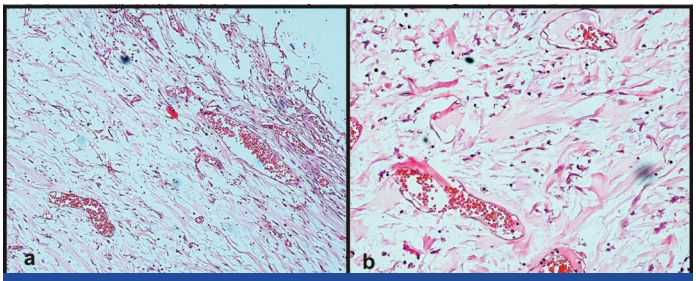
Primary peritoneal, omental and mesenteric tumours are relatively uncommon [1]. The first case of primary omental tumour was reported



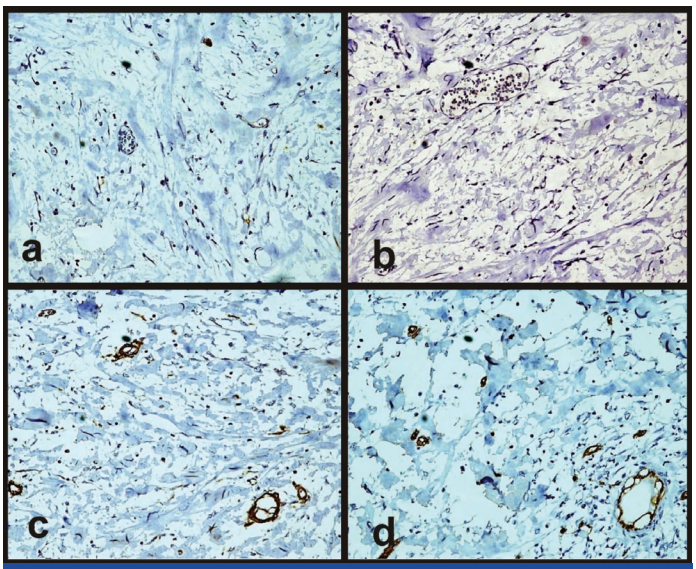

**[Table/Fig-2]:** a) Exploratory laparotomy revealed large encapsulated mass arising from greater omentum & b) Specimen revealed globular soft tissue mass arising from greater omentum



**[Table/Fig-3]:** Cut surface was fasiculated and pink gray in color




**[Table/Fig-4]:** (a) & (b) Microscopy revealed hypocellular lesion composed of spindle cells with minimal nuclear pleomorphism among bundles of dense collagen (H.E. X10 & X20 respectively)




**[Table/Fig-5]:** (a) Immunohistochemical examination (20x), Tumour cells showed negativity for CD117. (b) Tumour cells stained negative for DOG-1. (c) Tumour cells negative for SMA. (d) Tumour cells stained negative for CD-34

| SL NO | Author | Year | Country | Title |
|---|---|---|---|---|
| 1 | Anthony C et al., [2] | 1993 | California | Atypical desmoids tumour of the abdomen: A case report |
| 2 | Y. Pakosy et al., [3] | 1998 | Turkey | Omental fibroma: CT AND USG finding. |
| 3 | Domenico lusco D et al., [4] | 2005 | Italy | Giant fibroma of the lesser omentum: report of a rare case. |
| 4 | Masanori Ono et al., [5] | 2009 | Japan | Fibroma of the Omentum Resembling an Ovarian Tumour in the Pelvis |
| 5 | Ahmed, T et al., [6] | 2011 | Bangladesh | A 30-year-old female with lower abdominal lump. |
| 6 | Present case | 2014 | India | Giant omental fibromatosis presenting as pelvic mass |

**[Table/Fig-6]:** Cases of omental fibromatosis reported in the literature till date

by Stout and Cassel in 1942. Generic term "Fibromatosis" was first described by Stout and Raffle in 1961 and these unusal tumours account for 0.03% of all tumours. Based on extensive literature review, the incidence of primary omental tumours/ fibromatosis is unknown. Till date, 5 cases of primary omental tumours have been reported worldwide [Table/Fig-6].

Omental fibromatosis is a rare clinical entity also termed as abdominal desmoids. It is a benign proliferative disorder and locally aggressive. Fibromatosis have distinct biological behavior, characterized by initial rapid growth, followed by stability or even regression. Majority of fibromatosis occur sporadically, or in association with Gardener syndrome, familial adenomatosis polyposis coli (FAP) and bilateral ovarian fibromatosis. Risk factors include pregnancy, previous abdominal surgery, trauma and estrogen therapy [7]. Exact etiology and natural course is not well-understood. Majority of fibromatosis occur in women of child bearing age but there is no gender or racial predominance. Based on literature review, Yannopoulos et al., [8] revealed that both males and females were affected with equal frequency and Burke et al., [9] reported that the tumour was common in male with mean age of 41 y.

Most intra-abdominal fibromatosis affect mesentery of small bowel but uncommon in mesocolon and omentum. Mesenteric fibromatosis accounts for approximately 8% of all cases, while incidence and biology of omental fibromatosis is not well-known [1]. Most patients with omental fibromatosis are asymptomatic, discovered incidentally and may present with vague abdominal pain and palpable mass. These tumours usually lie in lower abdomen and are freely mobile with well-defined borders. Ultrasonography (USG) or computed tomography (CT SCAN) may be helpful in diagnosis but ultimately, final diagnosis is made by excision and histological examination. Differential diagnosis includes cysts, sclerosing mesenteritis, mesenteric panniculitis, or tumours originating from smooth muscle, neuronal tissue, adipose and stromal tissue of gastrointestinal tract. They rarely present with complications such as small bowel obstruction, bleeding, bowel perforation or fistula formation due to its ability to infiltrate adjacent structures. Intraabdominal fibromatosis should be treated mainly with surgery and medical therapy carries minimal benefit which includes chemotherapy, targeted agents, NSAIDs and antiestrogen therapy. Surgical approach includes wide local excision of these tumours as they have tendency toward local recurrence. Wide local excision with resection of adjacent structures is recommended [10]. Patients with fibromatosis have a prolonged survival even in advanced cases and some authors recommend a trial of observation with antiestrogens, nonsteroidal anti-inflammatory drugs such as sulindac with limited role of targeted agents such as Imatinib.

Majority of patients with fibromatosis have been misdiagnosed as GIST clinically and can be differentiated on immunohistochemistry (IHC) studies. CD 117 expressed commonly in GIST's can be positive in up to 75% cases of mesenteric fibromatosis, therefore IHC for beta catenin or DOG-1 should be included in the panel besides CD117 [11].

## CONCLUSION

Primary tumours of the omentum are rare and there is little information available in the literature. Omental fibromatosis is usually benign tumour but locally aggressive. Fibromatosis should be considered in differential diagnosis of pelvic and abdominal lesions. Majority are asymptomatic, and difficult to diagnose based on clinical presentation and radiological investigation. Final diagnosis is usually made on histopathology and immunohistochemistry (IHC) studies. Surgical excision is the only curative method of treatment with ill-defined role of medical or radiation therapy. Despite curative surgery, fibromatosis are prone for local recurrence.

## ABBREVIATIONS

AFP- Alpha fetoprotein, LDH- lactate dehydrogenase,

Beta-HCG- human chorionic gonadotropin, SMA- smooth muscle antigen,

CD- cluster of differentiation, DOG- discovered on GIT.

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