Acta Scientific Clinical Case Reports

Volume 3 Issue 2 February 2022

Case Report

# Beyond GIST: A Rare Case of Small Bowel Fibromatosis (Desmoid Tumor)

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**Received:** November 11, 2021

**Published:** January 06, 2022



## Abstract

**Background:** Desmoid tumors are included in a group of disorders called fibromatosis, are benign tumors which are composed of fibrous elements. Desmoid tumors have an estimated incidence of 3.7 new cases per million people per year [1]. Desmoids may occur in the mesentery, the abdominal wall, or the retroperitoneum. Even after complete surgical resection, recurrence of desmoid tumors is high. The high recurrence rate favors the use of non-surgical therapy, such as nonsteroidal anti-inflammatory drugs and antiestrogens. Distant metastases have not been reported.

**Case Description:** A 49-year-old female came with complaints of palpable abdominal mass and abdominal pain to surgery OPD. On examination there was a large abdominal swelling on left side and CT Abdomen was recommended. CECT Abdomen and MRI Abdomen were taken and initially it was diagnosed as small bowel GIST. Laparotomy and excision of the mass was done under general anesthesia. Post-operative specimen was sent for histopathological examination. Biopsy report came out as Jejunal Gastrointestinal stromal tumor (spindle cell type) and immunohistochemistry (IHC) was recommended to rule out lymphoma. IHC report turned out to be Fibromatosis (Desmoid tumour). Patient also presented with huge right broad ligament fibroid for which patient underwent hysterectomy with bilateral salphingo-oopherectomy.

**Conclusion:** GIST and Fibromatosis (Desmoid) are indistinguishable on imaging. Both show same radiological and clinical features.

**Keywords:** GIST; Fibromatosis; Immunohistochemistry (IHC); Desmoid Tumor

## Introduction

Mesenteric fibromatosis [Desmoid] comprises of benign fibrous tissue and constitutes tumorlike lesions of the mesentery. Histologically these lesions show presence of fibrosis. They are distinctly different in pathogenesis and biologic behavior, and these pathologic and radiologic features can overlap with one another and with more common nonneoplastic and neoplastic lesions of the mesentery. Mesenteric fibromatosis is locally aggressive benign proliferative disease that may occur in association with familial adenomatous polyposis or sporadically.

The small bowel mesentery is the most common site of origin of intraabdominal fibromatosis. However other structures in mesentery such as the ileocolic mesentery, omentum, transverse or sigmoid mesocolon may be the site of origin for intraabdominal fibromatosis.

The objective of our study is to show that GIST and desmoid have the same radiological features and hence desmoid (fibromatosis) should always be considered as a close imaging differential diagnosis.

## Case Report

- A 49-year old female came with complaints of palpable abdominal mass and abdominal pain to surgery OPD. On clinical examination, there was a large abdominal swelling on left side and CT Abdomen was advised.

- Initially plain CT Abdomen was taken which showed a large lobulated mass lesion seen predominantly occupying the left abdomen with mild right paramedian extension without any calcification.
- Then contrast was injected which showed huge heterogeneously enhancing lobulated mass lesion of size≈ 21 x 12.8 x 17.5 cm seen predominantly occupying the left abdomen. There was significant mass-effect with displacement of adjacent bowel loops and mesenteric vessels; the mass lesion extended cranially up to the epigastric region and caudally up to pelvis and there was significant intralesional vasculature.
- MRI whole abdomen was also done which showed evidence of large abdominal pelvic mass on the left side with internal signal voids and multiple vascular feeders arising from branches of splenic artery and jejunal branches of SMA. On post contrast the lesion exhibited intravenous contrast uptake with few central patchy non-enhancing areas within. The lesion was seen just cranial to the upper margin of uterus with no obvious evidence of any extension into adjacent ovaries and uterine fundus. The lesion was seen to cause mild indentation over adjacent vascular structures and displacement of the rest of the bowel loops.
- With this constellation of findings, a diagnosis of small bowel GIST was made and histopathological examination (HPE) was recommended.
- Patient was operated under general anesthesia. Midline laparotomy incision from xiphisternum to pubic symphysis was done. Tumor along with 25 cm of jejunum was excised in toto and specimen was sent for HPE. Side to side jejuno-jejunal anastomosis was done.
- Biopsy report came out as Jejunal Gastrointestinal stromal tumour (spindle cell type). IHC was recommended to rule out lymphoma.

IHC report turned out to be Fibromatosis (Desmoid Tumour)

- And also, this patient had undergone hysterectomy with bilateral salphingo oopherectomy for huge right broad ligament fibroid. Hysterectomy was done dividing bilateral broad ligaments and bilateral parametrium. Vault was closed and complete hemostasis obtained.
- Post operatively patient was monitored; there were no post-operative complications and vitals were normal. Patient was discharged on 5$^{th}$ post-operative day.

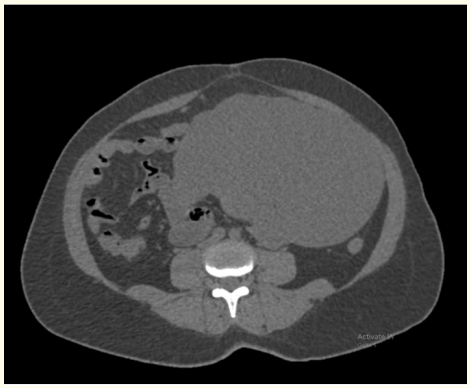


**Figure 1**

**Figure 2**

NCCT Abdomen shows large lobulated mass lesion seen predominantly occupying the left abdomen with mild right paramedian extension without any calcifications.

CECT Abdomen shows huge heterogeneously enhancing lobulated mass lesion of size≈ 21 x 12.8 x 17.5 cm seen predominantly occupying the left abdomen. There is significant mass-effect with displacement of adjacent bowel loops and mesenteric vessels.

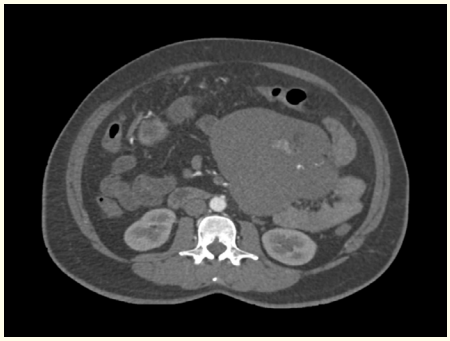


**Figure 3**

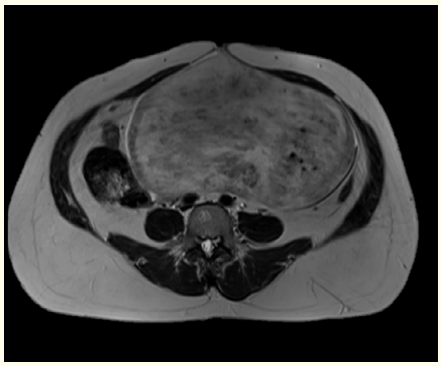


**Figure 6**

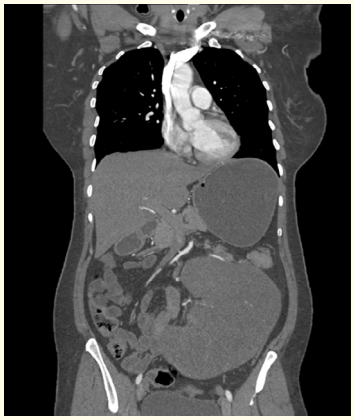


**Figure 4**

**Figure 7**

**Figure 5**

**Figure 8**

**Figure 9**

MRI Contrast whole abdomen shows evidence of large abdominal pelvic mass on the left side with internal signal voids and multiple vascular feeders arising from branches of splenic artery and jejunal branches of SMA. On post contrast sequences, the lesion exhibits intravenous contrast uptake with few central patchy non-enhancing areas within.

**Figure 10**

**Figure 11**

**Figure 12**

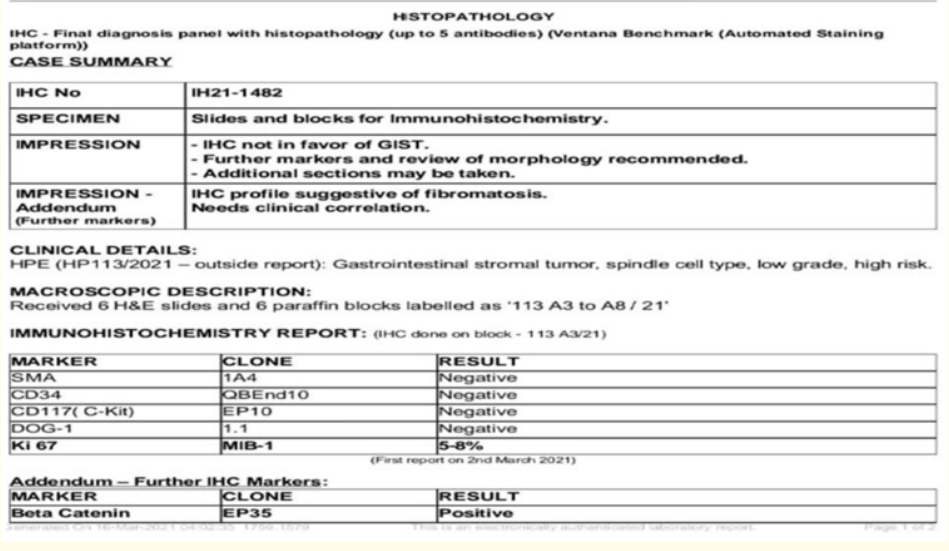
Post operative images

## Discussion

Mesenteric fibromatosis, also called intraabdominal fibromatosis or abdominal desmoid, is part of the clinical-pathologic spectrum of deep fibromatoses. The deep fibromatoses encompass a group of benign fibroproliferative processes that are locally aggressive and have the capacity to infiltrate or recur, but not metastasize. The deep fibromatoses are classified by anatomic location because they may arise from intraabdominal sites (mesenteric, pelvic, and retroperitoneal fibromatosis), the deep soft tissues of the abdominal wall (abdominal fibromatosis) and deep within extra abdominal soft tissues (extra abdominal fibromatosis).

Most intraabdominal desmoids arise in the mesentery, usually in the mesentery of the small bowel. Desmoids are the most com-

**Figure 13**



HISTOPATHOLOGY

IHC - Final diagnosis panel with histopathology (up to 5 antibodies) (Ventana Benchmark (Automated Staining platform))

CASE SUMMARY

| | |
|---|---|
| IHC No | IH21-1482 |
| SPECIMEN | Slides and blocks for Immunohistochemistry. |
| IMPRESSION | - IHC not in favor of GIST.<br>- Further markers and review of morphology recommended.<br>- Additional sections may be taken. |
| IMPRESSION - Addendum (Further markers) | IHC profile suggestive of fibromatosis.<br>Needs clinical correlation. |

CLINICAL DETAILS:
HPE (HP113/2021 – outside report): Gastrointestinal stromal tumor, spindle cell type, low grade, high risk.

MACROSCOPIC DESCRIPTION:
Received 6 H&E slides and 6 paraffin blocks labelled as '113 A3 to A8 / 21'

IMMUNOHISTOCHEMISTRY REPORT: (IHC done on block - 113 A3/21)

| MARKER | CLONE | RESULT |
|---|---|---|
| SMA | 1A4 | Negative |
| CD34 | QBEnd10 | Negative |
| CD117( C-Kit) | EP10 | Negative |
| DOG-1 | 1.1 | Negative |
| Ki 67 | MIB-1 | 5-8% |

(First report on 2nd March 2021)

Addendum – Further IHC Markers:

| MARKER | CLONE | RESULT |
|---|---|---|
| Beta Catenin | EP35 | Positive |

**Figure 14**

mon primary tumor of the mesentery [2]. Most of these tumors occur sporadically. The incidence of abdominal wall and mesenteric desmoids in patients with Gardner's syndrome ranges between 4% and 29% and the tumors typically occur after abdominal surgery [1]. Desmoid tumors may be also associated with trauma and estrogen therapy [2].

Desmoids may occur in all age groups but they are typically seen in the third and fourth decades of life [1,3]. Women are affected more commonly than men [1,3]. However, in the largest study in the literature, with 130 cases, 55% of the cases occurred in men [2].

Most patients with desmoid tumors are clinically asymptomatic, and the tumors cause little or no focal symptoms until late in their course. Patients may present to a physician because of a palpable mass, abdominal pain or gastrointestinal bleeding [2]. Desmoids are usually locally aggressive lesions. Some complications that have been reported include small-bowel obstruction and hydronephrosis [4].

On gross histopathologic examination, desmoid tumors are usually circumscribed lesions, but they may have irregular or infiltrating borders. On the cut surface, they are white and coarsely trabeculated, resembling scar tissue. Desmoids are usually larger than 5 cm when they are discovered, and they may be larger than 15 cm. In 10-15% of cases, desmoids are multiple [5]. Histologically, desmoid tumors are composed of stellate fibroblastic cells or bland spindled embedded in a collagenous stroma without evidence of neural or muscular differentiation. This tumor may infiltrate adjacent viscera or tissues in the periphery [2].

On ultrasound, desmoid tumors have variable echogenicity, appearing as masses of low, medium or high echogenicity with smooth and well-defined margins [3].

In CT most desmoid tumors appear as well-circumscribed homogeneous masses that may be isodense or hyperdense relative to muscle. Some cases of heterogeneous masses with infiltrative outer margins are seen. Desmoid tumors may enhance after injection of intravenous contrast [1]. On MRI, desmoid tumors appear as masses of low signal intensity relative to muscle on T1-weighted images. On T2-weighted images, desmoid tumors show variable signal intensity relative to muscle [3].

Even after complete surgical resection these desmoid tumors have a tendency to recur. The high recurrence rate favors the use of non-surgical therapy, such as nonsteroidal anti-inflammatory drugs and antiestrogens. Distant metastases have not been reported.

## Conclusion

GIST and Fibromatosis (Desmoid) are indistinguishable on imaging. The objective of our study is to show that GIST and desmoid have the same radiological and clinical features and hence desmoid (fibromatosis) should always be considered as a close imaging differential diagnosis.

## Bibliography

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