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Unusual Cause of Abdominal Lump

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Clinical Combined Round

Surgery Unit III

- Clinical case discussion.
- Radiological findings.
- Operative summary.
- Histopathological findings.
- Review of Literature.

History

- 25/F .
- Abdominal distension x11 months.
- No associated pain.
- No h/o fever.
- No jaundice/haematemesis/malena.
- No loss of appetite/no loss of weight.
- Normal bowel and bladder habits.
- No previous surgeries.

General Examination

- Wt- 52 kg BMI-22.6kg/m^2
- Afebrile, normotensive.
- No pallor, jaundice, significant lymphadenopathy.

Investigations

- Routine blood investigations:WNL.
- Radiological investigations.

Surgery

- CTVS and Urology kept on stand-by.
- Procedure: Laprotomy + excision of mass along with resection of involved part ileum and ascending colon with ileocolic anastomosis.

Post operative period

- Post operative course was uneventful.
- Orally allowed day 5.
- Discharged on day 7 on normal diet.

Final Diagnosis

- Mesenteric fibromatosis with infiltration into surrounding bowel and vascular invasion.

Discussion

- Mesenteric fibromatosis-subtype of Desmoid tumors.
- Desmoid tumors are cytologically bland fibrous neoplasms originating from the musculoaponeurotic structures throughout the body.
- The myofibroblast is the cell considered to be responsible for the development of desmoid tumors.

- Overall they account for 0.03% of all neoplasms[1].

Mesentric fibromatosis

- Proliferating fibrous tissue in the bowel mesentery
- Invades the bowel or adjacent tissues with aggressive myofibroblastic proliferation
- Lacks the capacity of malignant tumorigenesis do not metastasise.

Incidence

- 8% of all desmoid tumors
- Most common primary tumor involving the mesentery. (V. Gómez Cabeza de Vaca et al "Mesenteric fibromatosis: a rare entity" Rev Esp Enferm Dig (Madrid) Vol. 101, N.° 7, pp. 506-519, 2009)
- Sex ratio W:M - 2:1
- Mean age – 35 [Figure: 1-9].

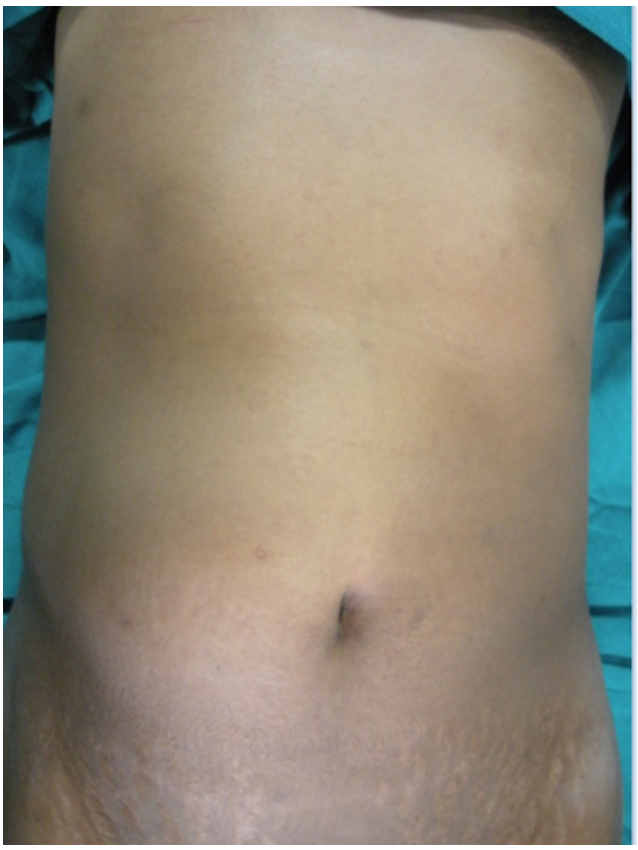

Figure 1: Abdominal examination

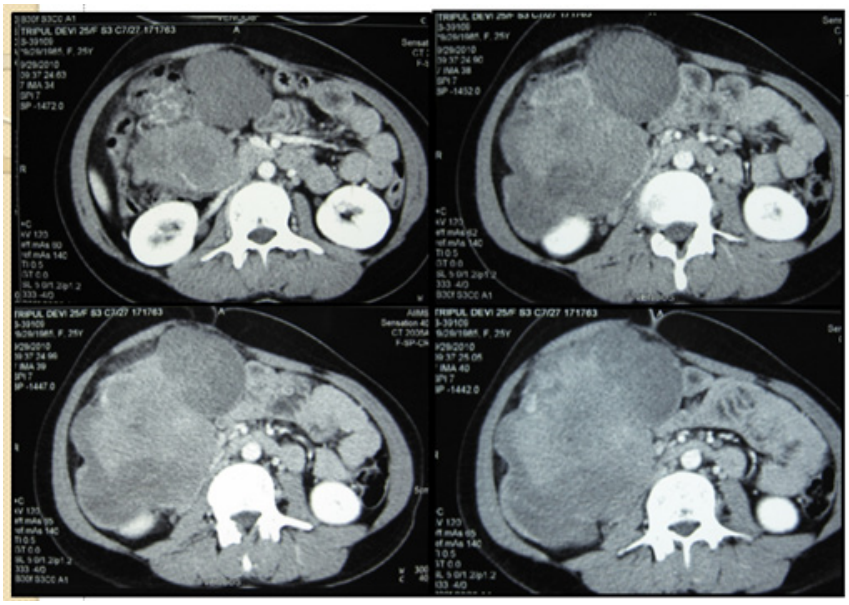

Figure 2: Investigations

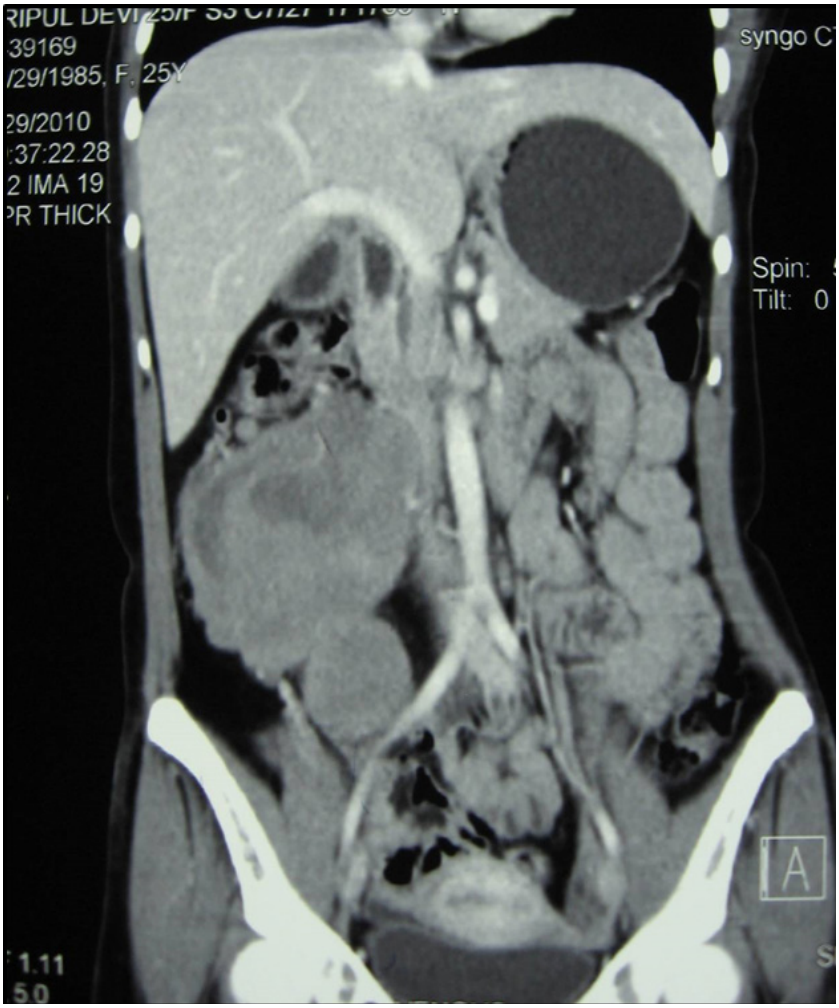

Figure 3: Investigation Surgery

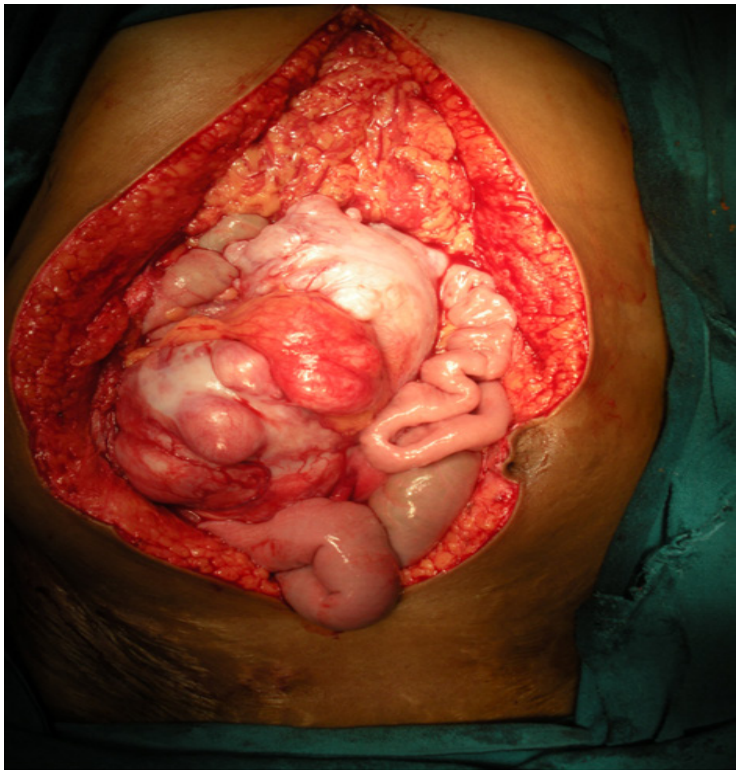

Figure 4: 16 x 15cm mass arising from mesentery of ileum. Tumour infiltrating ileocaecal junction, ileum, proximal part of ascending colon, mesentery of ileum

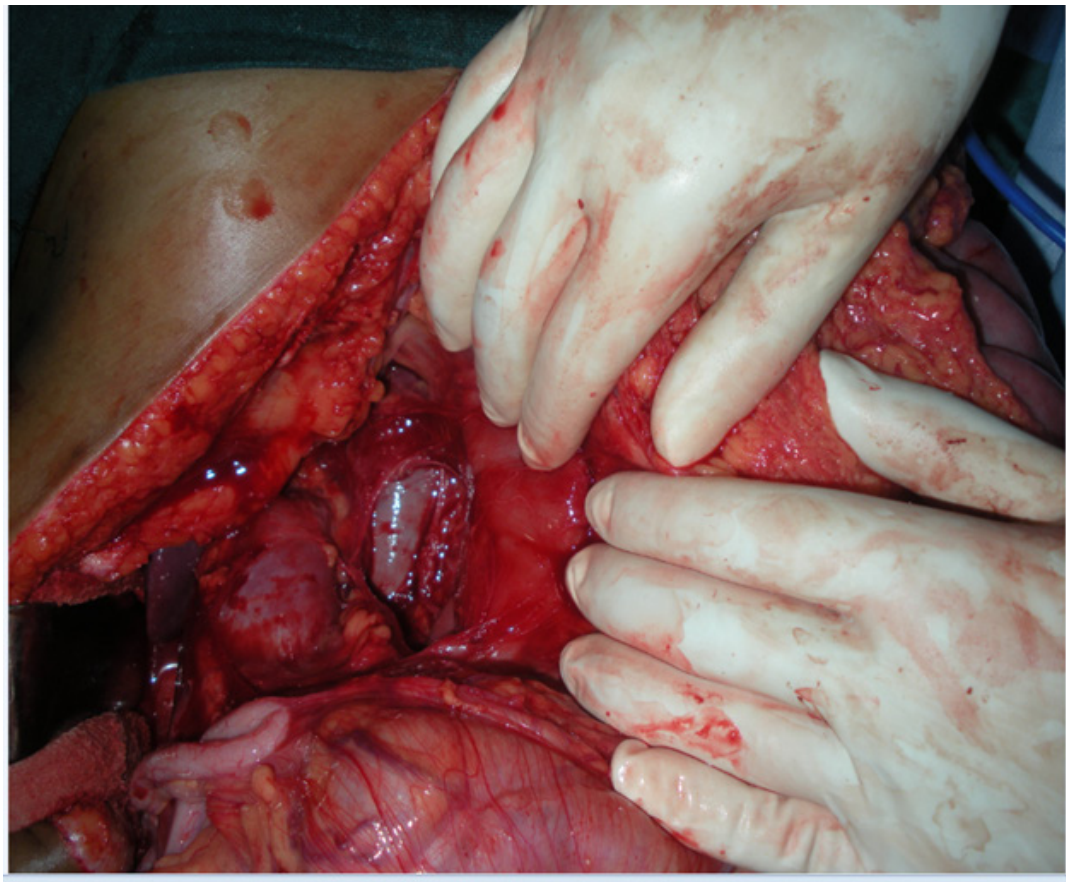

Figure 5: Tumor could be easily separated from the IVC

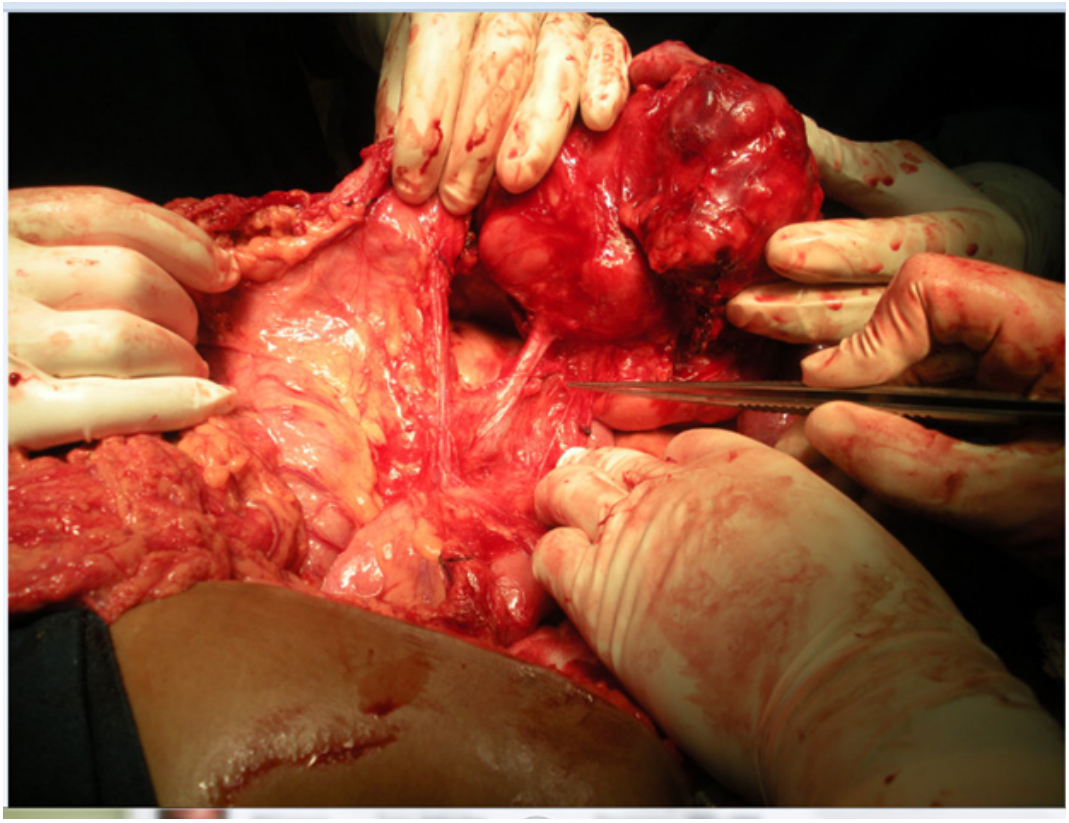

Figure 6: SMA infiltrated distal to jejunal branches ileo colic and right colic branches.

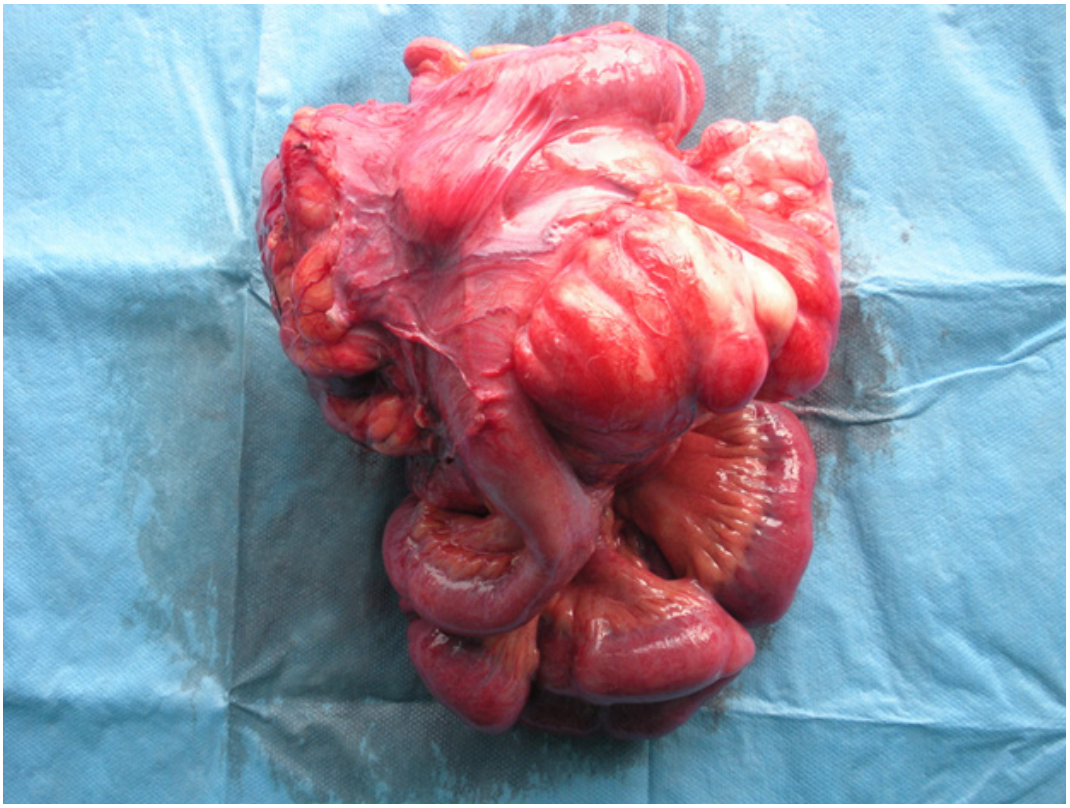

Figure 7: Mass excised enbloc with wide proximal and distal margins along with involved segment of ileum, ileocaecal junction, and ascending colon. Stapled ileo colic anastomosis done.

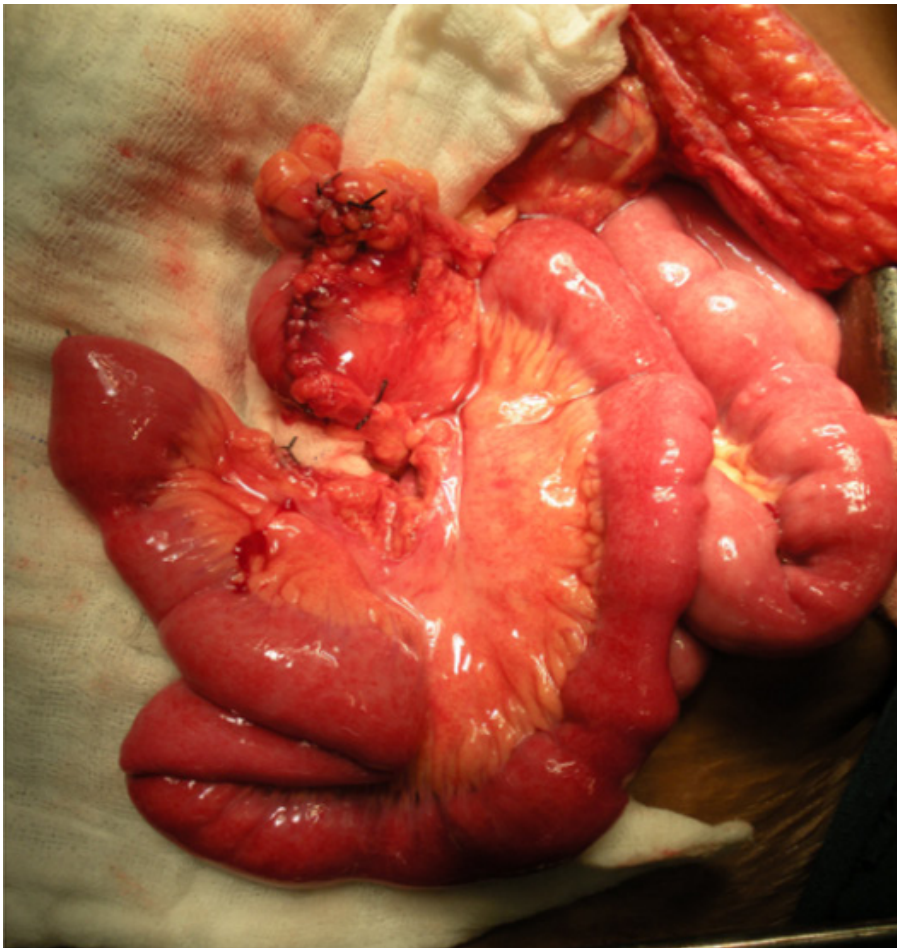

Figure 8: Resected ends of ileum and transverse colon

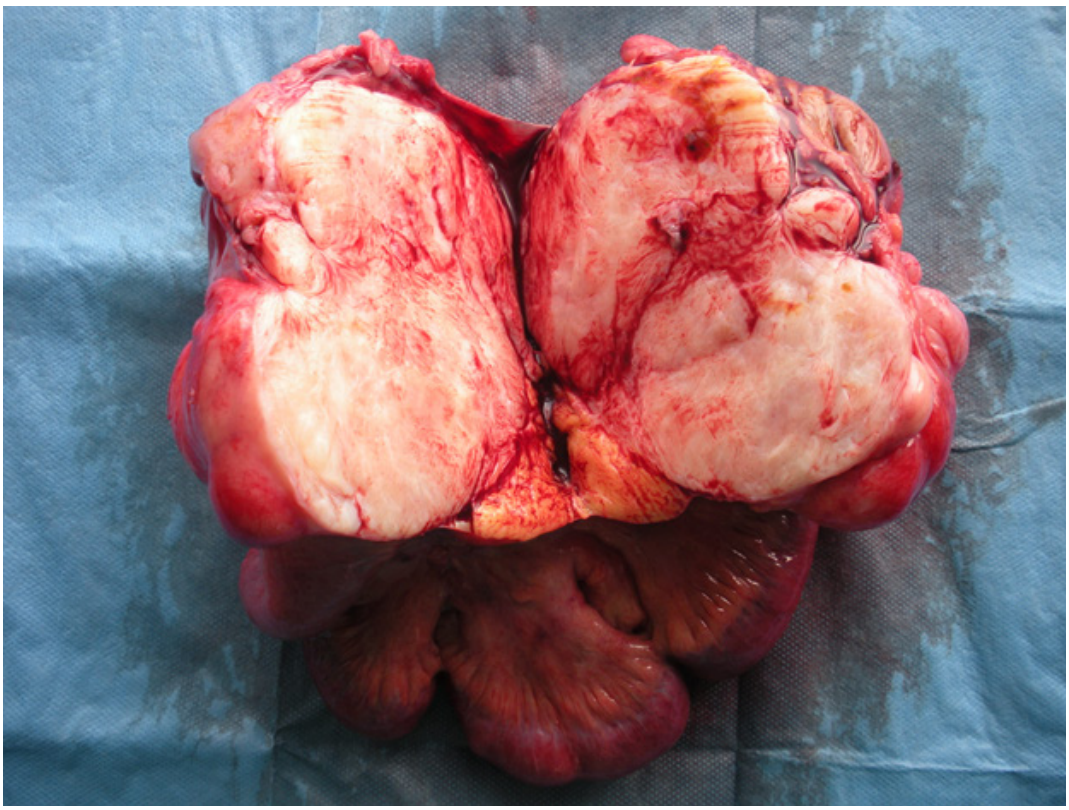

Figure 9: Cut section of tumour

Aetiology

- Unknown
- Associations
    - o prior trauma or abdominal surgery (10-12%)
    - o with FAP (Gardner's syndrome) (25%)
- Sporadic cases seen
    - o Probable endocrine etiology:
        - the relative increased prevalence in perinatal women;
        - tumour regression after menopause;
        - regression with tamoxifen therapy [2].

Location

- Mesentery of the small bowel is frequently involved;
- Can also originate from
    - o ileocolic mesentery,
    - o gastrocolic ligament,
    - o Greater omentum
    - o Retroperitoneum [3].

Clinical presentation

- Mass per abdomen
- Acute pain abdomen
- Ureteric /bowel obstruction [4].

Investigations

- CT
- MRI
- Histopathology
- Intra-abdominal lesions with spindle-cell morphology -rare, the similarity of their histological appearance to that of other lesions frequently leads to misdiagnosis
- Mesenteric fibromatosis -CD117 ,typically expressed more weakly than in GIST, actin , and desmin in keeping with myofibroblastic differentiation but lacked CD34, S-100, and keratin.
- "Beta-catenin immunohistochemistry separates mesenteric fibromatosis from gastrointestinal stromal tumor and sclerosing mesenteritis."[5].

Treatment

- Curative.
- Palliative.
- Curative:
    - o Surgical resection – complete resection with wide margin-BEST RESULTS

Palliative:

- Chemotherapy.
- Radiotherapy [6].

Radiotherapy

- Adjuvant modality.
- Post-operative radiation therapy is recommended for
    - o positive margins identified histologically.

- recurrences.

- Along with systemic chemotherapy [7].

Chemotherapy –Cytotoxic

- Combination regime.
- Doxorubicin - based chemotherapy (with dacarbazine or cyclophosphamide
- and vincristine) .
- Actinomycin-D-based chemotherapy.
- Combination of methotrexate with a vinca alkaloid [8].

Chemotherapy-Non Cytotoxic

- Tamoxifen –no correlation between response to tamoxifen and ER status.
- NSAIDs
  - first-line treatment in patients with a high risk general condition.
  - FAP-associated desmoids Interferons..
- Anika Hansmann Cancer 2004;100:612–20.
- High-Dose Tamoxifen and Sulindac as First-LineTreatment for Desmoid Tumors.
  - 120 mg tamoxifen & 300 mg sulindac.
  - 3/3 sporadic tumours showed cessation of growth .
  - 10/13 cases associated with FAP showed partial or complete remission [9].

Summary

- Mesenteric fibromatosis is a rare cause of abdominal symptoms.
- May be associated FAP or Gardener's syndrome.
  - Surgical excision with a negative free margin is the treatment of choice.
- Palliative:Radiotherapy/chemotherapy/hormonal therapy.
- Regular followup may be considered in view of locally recurrent nature of disease [10,11,12].

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