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Fibromatosis of The Appendix Presenting With Right Iliac Fossa Mass:- A Rare Case Report

AUTHOR

Abstract
Intra-abdominal fibromatosis are uncommon benign neoplasms which originate from musculoaponeurotic structures of the body and primarily affects the mesentery or retroperitoneum. However, rarely, they can arise from the intestinal wall and mimic gastrointestinal malignant neoplasm. A rare case of proliferative fibroblastic lesion involving the appendix and meso appendix presenteting as RIF mass with its origin ,diagnosis and differential diagnosis are discussed here .

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I. Case Report

A 33 year old female was presented in our OPD with complain of swelling in right lower abdomen since 8 months. She noticed this swelling 8 months back which was insidious in onset and gradually progressive in nature .There was no history of abdominal pain, fever ,vomiting ,bleeding per rectum, and bleeing per vagina .There was no history of anorexia and significant weight loss .There was no changes in her bowel and bladder habits .There was no history of previous abdominal surgery or trauma .

She had 3 children with normal vaginal delivery with history of multiple Miscarriages. She has normal menstrual cycle with normal flow .

On examination :-

She was conscious ,oriented to time person and place at time pr presentation .

she was thin built with BMI 19.5

Her systematic examination was normal .

Local examination :-

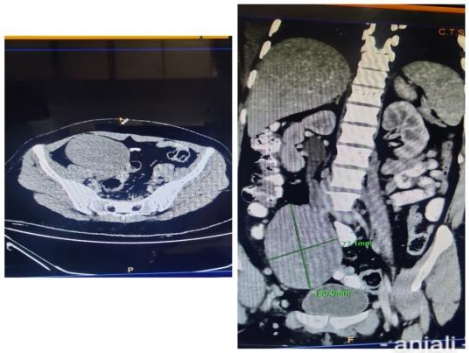
There was right illac fossa mass approximately size of 10 x 10 cm palpated. Mass was non tender , mobile ,firm in consistency with ill defined margin . Patient was thoroughly investigated .

All routine baseline investigations were found to be normal.

USG whole abdomen revealed ;-A well defined predominately hyperechoic mass of size 10.2 X 5.8 cm noted in right illac fossa .On color Doppler it shows arterial flows .An eccentric component is also noted in mass.- suggestive of necrosis .

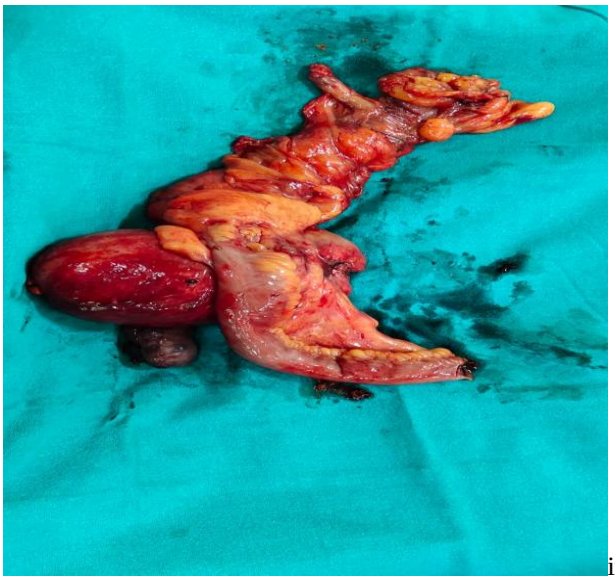
There was modereate right hydronephrosis with dilated right ureter (6.4 mm) and is traced upto the inferior aspects of RIF mass .

Contrast enhanced computerized tomography showed -well defined low-attenuating lesion with areas of soft tissue density within showing progressive enhancement of the soft tissue component in the right iliac region abutting the right adnexa, however appearing separate from the right ovary-may represent a mesenchymal lesion arising either from the broad ligament or from the retroperitoneum in the right iliac region.

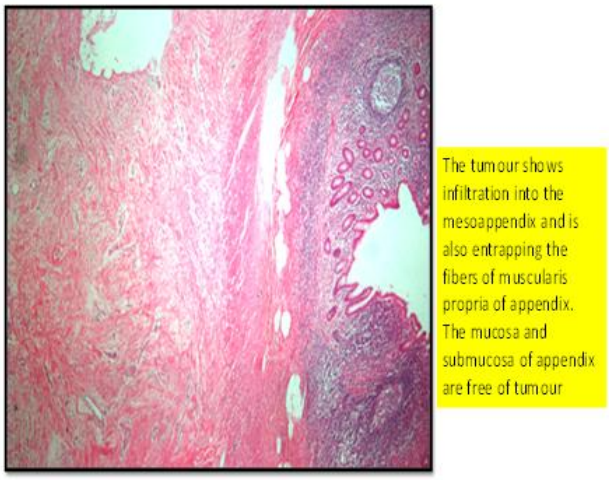

CECT whole abdomen

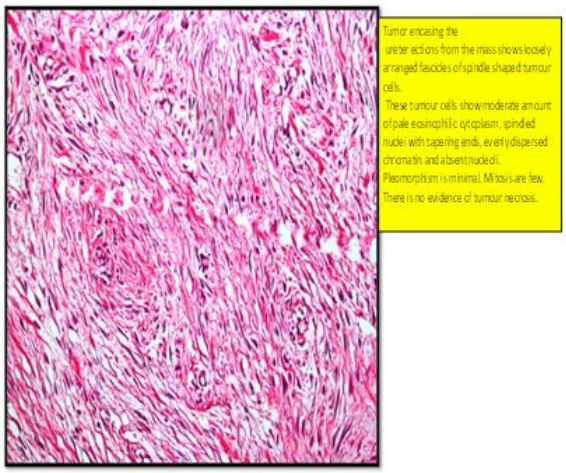
Patient was placed for explorary Laparatomy which revealed a mass size of 10X10 cm arising from wall of appendix and shows tip of appendix at periphery .Mass was adherent to ileo- cecal junction ,ileum ,right ovary and 1/3 rd part of right ureter. There was right hydronephrosis . complete excision of mass with right hemicolectomy with ileo-transverse anatomoses was done .right ureter reconstruction was done using boari's flap method. There was no free fluid in peritoneum .Histopathologic examination confirmed the diagnosis of intrabdominal fibromatosis arising from appendix . Ki67-< 1%. Desmin, CD34, CD117, STAT 6 and ALK-1 are negative

Post operative period was uneventful and patient recovered well .we followed up patient upto 6 months .

 intraoperative (specimen)

HISTOPATHOLOGY :-





II. Discussion:-

Mass in the right iliac fossa has innumerable differential diagnosis ranging from mass arising from abdominal wall to peritoneal cavity to retroperitoneum. These include common disease processes like appendicular mass or abscess, ileocaecal tuberculosis, carcinoma caecum, intussusception, and lymphoma to some rare diagnosis like retroperitonealsarcoma, iliac artery aneurysm, or chondrosarcoma of iliac crest. In females right iliac fossa mass may be due to ovarian cyst or tuboovarian mass. The present case discusses the rare possibility of appendicular fibromatosis presenting as a rightiliac fossa mass because it has never been mentioned in this differential diagnosis.

Fibromatosis also known as desmoids tumors ,are local aggressive benign neoplasms which originate from musculoaponeurotic structures of the body .They may have a peripheral or intra –abdominal localization.

While intra- abdominal fibromatosis genereally originates from mesentery of small intestine but they can invole retroperitoneum ,transverse mesocolon and omentum . Rarely ,it can also arise from intestinal wall (submucosa) .They can mimic as gastrointestinal stromal tumor (GIST).Absence of reaction with CD117 and S-100 is key point of differential diagnosis between fibromatosis and GIST as in the present case .1

Intra –abdominal fibromatosis are local aggressive neoplasm , which do not show metastatic tendency .While most cases have an asymptomatic course ,urgent surgical interventios have been reported due to reasons such as intestinal obstruction ,perforation and abscess formation.2

As there is no classical Symptoms related to appendicular fibromatosis, the diagnosis is confirmed only after the histological analysis of the tumor. Imaging remains the mainstay of preoperative investigations to establish a working diagnosis of fibromatosis . CT scan is considered the first line imaging modality for identifying, characterizing, and staging fibromatosis. On CT, these tumors appear as a soft tissue mass displacing/involving surrounding viscera, usually appearing as encasement of bowel loops . Although the mass may appear well circumscribed, it often has irregular margins reflecting its infiltrative nature.3

Microscopically, appendicular fibromatosis is characterized by loosely arranged fascicles of spindle shaped tumor cells. Thes e tumor cells show moderate amount of pale eosinophilic cytoplasm ,spindled nuclei with tapering ends, evenly dispered chromatin and absent nucleoli. Pleomorphism is minimal . The mitotic count is relatively low with no evidence of necrosis and nuclear dedifferentiation . Wide field surgical excision is the first-line treatment for most appendicular fibromatosis 4. As noted in our case, the majority of these lesions require resection of the adherent surrounding structure. surgical excision is the gold standard primary treatment for appendicular fibromatosis .To conclude, appendicular fibromatoses may present with bizarre clinical features and demonstrate a wide spectrum of imaging and histological spectra. while managing a patient with an abdominal mass and consider this entity in the differential diagnosis of a right iliac fossa mass as well.

To our Knowledge ,this is the first documented case of appendiuclar fibromatosis ,adherent to wall of cecum and right ureter .

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