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ARIPEX

ORIGINAL RESEARCH PAPER

BETA CATENIN NEGATIVE GASTRIC FIBROMATOSIS

KEY WORDS: Beta catenin,

General Surgery

Fibromatosis, Gastrointestinal stromal tumor.

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Gastric fibromatosis-Locally aggressive tumor, rarely metastasise, hereby presenting a case of 45 year old female who manifested with complaints of vomiting, clinically unremarkable with imaging showing features suggestive of gastrointestinal stromal tumor, proceeded with surgical resection of extraluminal mass, biopsy of which reported FIBROMATOSIS OF KELOID TYPE; Betacatenin negative

BACKGROUND

Gastrointestinal fibromatosis is a rare tumor that represents about 0.03% to 0.1% of all neoplasms. Fibromatosis is a histologically benign growth of fibroblastic and myofibroblastic cells, with a potential to recur and invade local organs. It can occur as a superficial or deep form. Visceral fibromatosis and superficial fibromatosis are histologically similar. They both have alterations in the WNT signalling pathway, usually 70-80% fibromatosis are betacatenin positive (4) but mutations in the APC or -catenin gene do not occur in superficial fibromatoses.

Abdominal Fibromatosis can be:(a) Abdominal wall Fibromatosis- on the walls of abdomen (b) Extra abdominal Fibromatosis- Outside the abdomen/abdominal wall (c) Intra abdominal Fibromatosis- Within the abdomen in mesentery, pelvis, retroperitoneal locations(3).

Here by presenting a case of Betacatenin negative gastric(visceral) fibromatosis.

Case Presentation And Workup

Female in her 40s presented with complaints of multiple episodes of vomiting following food intake for one week. Clinical examination is unremarkable. Routine labs were found to be normal.

Upper gastrointestinal endoscopy - showed submucosal mass (Figure 1) with Histopathology reporting Chronic Gastritis. Contrast enhanced computed tomography showed well defined homogeneous extra luminal solid mass of size 2.9x3.4x3.9 cm arising from the greater curvature of stomach (proximal body region) with loss of fat plane with greater curvature of stomach. No significant contrast enhancement noted.No evidence of fat/ calcifications / hemorrhage/ necrosis within-Features suggestive of Gastro intestinal st romal tumor (Figure 2).



Figurel 94

Treatment

Patient was taken up for surgical excision of tumor. Intraoperatively- 4x5 cm mass arising from the greater curvature of stomach adherent to surrounding omentum was found and proceeded with Surgical excision of extra luminal mass (Figure 3). Histopathology showed : FIBROMATOSIS OF KELOID PATTERN with Betacatenin - negative (Figure 4).





Follow Up

Being highly aggressive tumor, 6 month followup showed no recurrence.

DISCUSSION

Fibromatosis may be due to mutations in APC / Beta Catenin / Both. Can also be sporadic due to previous surgeries and Trauma. Aggressive Fibromatosis is a abnormal hyperplasia of fibroblasts / myofibroblasts in deep soft tissues. WHO(2013)- INTERMEDIATE LOCALLY INVASIVE TUMOR (rarely metastasise). Gastric Fibromatosis case may also present with complaints of abdominal pain , distention, vomitings, sometimes a palpable mass. If erodes mucosa leads to ulcers, Gastro intestinal bleeding causing hematemesis, also can lead to Bowel perforation, obstruction, fistula formation, ureteral obstruction, mesentric vessel compression and causing further complications.

Differential Diagnosis

One of the major differential working diagnosis will be GIST (Gastrointestinal stromal tumor) due to its similar manifestations, to be differentiated by pathological differences (1). GIST presents as soft, lobulated mass with haemorrhage, necrosis and calcification within and Beta Catenin negative where as fibromatosis is a firm ,tan ,homogenous mass with no evidence of hemorrhage, necrosis and calcification and 70-80% of fibromatosis will be Beta Catenin positive. Other differentials like Inflammatory Fibroid

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polyp, Schwannoma, Leiomyoma, Inflammatory myofibroblastic Tumor, Schirrous carcinoma stomach, Follicular dendritic cell carcinoma, Inflammatory malignant fibrous histiocytoma, Myofibroma/Myofibromatosis, Solitary fibrous tumor of stomach should be ruled out histologically.

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