Mucosal Benign Epithelioid Tumor Of Jejunum: A Case Report
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ABSTRACT: Mucosal benign nerve sheath tumor is derived from nerve sheath which is like “Schwann cell hamartoma” but cannot be classified as schwannoma or neurofibroma. These are rare group of gastrointestinal tumors mainly reported to occur in large intestine and bladder. We report a extremely rare case of mucosal benign epithelioid tumor of jejunum in a 20 yrs old female presenting as acute abdomen.

KEYWORDS: Mucosal tumor, Gastrointestinal stromal tumor, Biopsy

Introduction: Primary neurogenic neoplasms rarely involve the small intestine, considering the 35 different varieties of small bowel tumors that have been compiled from literature[1]. Mucosal benign epithelioid nerve sheath tumors have not been well described in the gastrointestinal tract and other mucosal sites. Most of these reported tumors can be seen in colonic and bladder mucosa. These tumors have a mean of presentation at the age of58.6years with slight female predominance.[2] Many a times these benign tumors appear as small polyp on colonoscopy. We present a benign mucosal epithelioid tumor arising from jejunum with clinical presentation of acute abdomen.

Case Report: A 20 years old female presented with acute abdomen of 1 day history with symptoms of pain around umbilicus and 2 episodes of vomiting. On examination she had tachycardia and tenderness over umbilical and hypogastrum regions with bp-100/60mm of Hg. USG shows a oval well defined hypoechoic solid lesion seen in lower abdomen infraumbical region to the left of midline. CECT shows a isodense inhomogeneously enhancing soft tissue exophytic tumor of size 5.8*4.2*4.3 arising from small bowel likely to represent gastrointestinal stromal tumor. There is no lymphadenopathy with moderate amount of free fluid in the pelvis. Initially we proceed with laparoscopic exploration. But due to dense adhesions we converted it into formal laparotomy with lower midline incisions. After entering into abdominal cavity a tumor...
arising from jejunum approximately 7 feet distal to D-J junction with dense adhesions with surrounding bowels was seen. [Figure-1]

After clearing all adhesions carefully segmental resection of bowel having tumor carried out with end to end anastomosis. [Figure-2] The postoperative period was uneventful and orally allowed on 4th postoperative day. Biopsy sent to 3 different labs. Histopathology from all the 3 labs reported uniform epithelioid morphology with spindle component lacking ganglion cells and axons. These cells are very strongly positive for S100 and negative for C-kit, CD34, SMA, Calponin, HMB45, Melan A. There are no mitoses or necrosis. The patient is followed for 8months without any complication.

Mucosal benign epithelioid nerve sheath tumor is defined by strong intramucosal infiltrate of S100 positive epithelioid cells lacking other neural elements[3]. These tumors usually occur sporadically with no relationship with familial syndromes like Von Recklinghausen’s syndrome[4]. Most of the reported cases are incidental finding. When found they should be resected to prevent complication like infection and obstruction.

Surgical resection is curative

References:


Discussion: