Case Report

Isolated Nerve Sheath Tumor Of Jejunum Presenting As Acute Abdomen

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Abstract

Nerve sheath tumors of gastrointestinal tract are rare and occurs more frequently as systemic manifestations of generalized neurofibromatosis type-1(NF-1).In neurofibromatosis-1 lesions may be scattered or solitary tumor. Occasionally ,such tumors may be the initial presenting feature of NF-1.Rarely ,cases of isolated nerve sheath tumor of small intestine presenting with bleeding and obstruction have been documented. We report a 19years male presenting with nerve sheath tumor of jejunum in acute abdomen. His investigations show no features of NF-1.

Keywords: nerve sheath tumor, neurofibromatosis, isolated, intestinal.

Introduction

Nerve sheath tumors of small intestine may manifest in various forms and accordingly given different descriptive terms including intestinal neurofibromatosis, ganglioneuromatosis, neuronal intestinal dysplasia and diffuse plexiform neurofibromatosis. These lesions may be seen in 25% cases of neurofibromatosis-1 and multiple endocrine neoplasia type 2b.There occurrence without NF-1 and MEN-2b are very rare1. No consensus has been made yet whether these isolated lesions represent different phenotypic manifestations of the neurocutaneous or multiple endocrine neoplasia syndromes or whether they represent separate and distinct entities.

Case Report

A 19 years old male presented to us with acute abdomen of 1 day history with symptoms of pain around umbilicus and 2 episodes of vomiting. On examination he had tachycardia and tenderness over umbilical and hypogastrium regions with bp-90/60mm of Hg. He had rigidity and rebound tenderness more marked in lower abdomen. USG shows a oval well defined hypo echoic solid lesion seen in lower abdomen infraumblical 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]

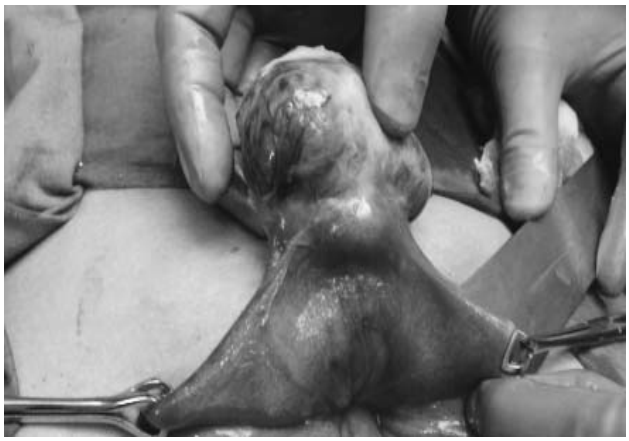

Figure I Intra operative photograph showing tumor arising from jejunum

After clearing all adhesions carefully segmental resection of bowel having tumor carried out with

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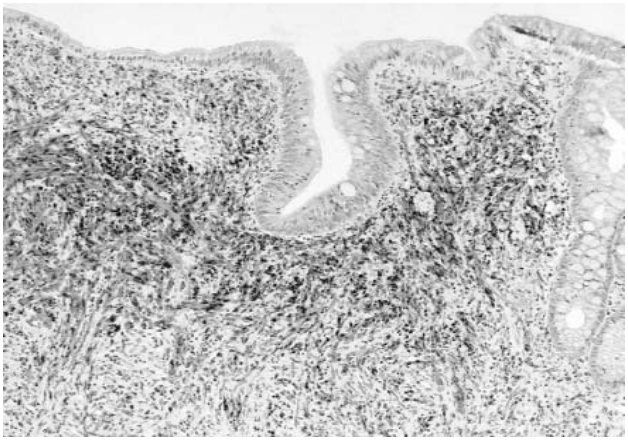

Figure II Immunohistochemistry showing strong s-100 reaction

end to end anastomosis. [Figure-II] The postoperative orally allowded on 4th postoperative day and recovery was uneventful .Biopsy sent to 3 different labs. Histopathology from all the 3 labs reported uniform epithelioid morphology with spindle component . These cells are very strongly positive for S100[Figure-3] and negative for C-kit, CD34, SMA, Calponin, HMB45, Melan A. There is no mitoses or necrosis. The patient is followed for 8months without any complication.

Discussion

Isolated nerve sheath tumors are benign neural lesions of intestinal tract which may be initial presenting feature of NF-1 and MEN tepe2b. Affected patient may present with altered bowel habit2, abdominal pain3, intestinal obstruction4 or with palpable abdominal mass. In setting of NF-1 associated clinical findings including dermal neurofibromas, café-au-lait spots and lisch nodules may be seen. In MEN type 2b thickened lips, marfanoid habitus and medullary carcinoma of thyroid may be seen. Isolated nerve sheath tumors commonly involve colon , terminal ileum , and appendix5. Surgical excision is the treatment of choice for these lesions These cases need to be followed up closely to exclude NF-1 and MEN type2b in future.

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