

CASE REPORT

SOLITARY NEUROFIBROMA OF THE MESENTERY PRESENTING AS ACUTE INTESTINAL OBSTRUCTION

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ABSTRACT: INTRODUCTION: The isolated presence of neurofibromatous lesions in the gastrointestinal tract, with no associated systemic syndromes, is a rarely reported clinical entity.

PRESENTATION OF CASE: A 25-year-old lady, with no history of neurofibromatosis or other systemic disease, presented with small bowel obstruction secondary to band formation induced by an isolated mesenteric neurofibromatous mass. The patient underwent exploratory laparotomy with release of the band and removal of the mass and after a smooth recovery; she was put on a long-term follow-up schedule.

DISCUSSION: This article presents a review of the literature of this area clinical entity. Very few reports of gastrointestinal isolated neurofibromas could be found. Similarly, extra-digestive isolated lesions have been rarely reported. **CONCLUSION:** Isolated mesenteric neurofibroma is a rare pathological entity. The clinical significance of such a diagnosis lies mainly in the need of further follow up of these patients as the bowel involvement could be the first manifestation of neurofibromatosis type 1 or multiple endocrine neoplasia type 2b.

KEYWORDS: Neurofibroma, Small intestinal tumour, Intestinal obstruction, mesenteric neurofibroma.

INTRODUCTION: Neurofibromatous proliferations in the gastrointestinal tract, in general, and the small intestines, in particular, have well been described in neurofibromatosis type 1. However, the isolated presence of such lesions in the intestines with no evidence of systemic disease is a rarely reported clinical entity. Here in, we report the case of an isolated mesenteric neurofibroma in a young lady, presenting as an acute intestinal obstruction. A review of the medical literature on this rare entity follows.

CASE REPORT: This is a case report of a 25yrs female housewife presented with Chief complaints of pain abdomen and distension since 4 day. Pain abdomen was sudden in onset, gradually increasing, dull aching intermittent type of pain, no alleviating or relieving factors. h/o distension since 4 days, gradually increasing. No relief with vomiting. H/o vomiting – 2 episodes, bilious. H/o constipation for 3 days. No H/o bladder disturbances, per rectal bleeding. Patient is a mother of two children the youngest being one year. Patient was toxic, with tachycardia and hypotension. Per abdominal findings showed gross distension with guarding and rigidity, with absent bowel sounds. Per rectal examination was normal. CVS & RS showed no abnormality. Blood investigations were normal except for widal positive. X-ray erect abdomen showed small bowel obstruction, no gas under diaphragm. Patient was posted for exploratory laparotomy. Intraoperative findings showed distended bowel loops of small intestine. Band at terminal ileum

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about 20cm proximal to ileocaecal junction. Band explored to its origin and attachment. A mass was found to be at mesenteric end of the bowel which formed an adhesion to the other part of bowel thus occluding it. The mass was explored, was extraluminal, a whitish ovoid mass with smooth contours, firm in consistency, non-pulsatile, no septations, mass was excised out and rest of the adhesions released. No perforation present. Gastroduodenal and ileocaecal junctions were normal. No similar or any other masses were found in the rest of the bowel. Postoperatively the recovery of the patient was uneventful.

Pathology Findings: Section studied from mass in right iliac fossa reveals wavy nerve fibres and fibrocollagenous tissue suggestive of neurofibroma.

Outcome: Following the pathological diagnosis of an isolated small bowel neurofibroma, a thorough physical examination of the patient failed to reveal any evidence of neurofibromatosis, multiple endocrine neoplasia or other syndromes. She had a smooth post-operative course. Food intake was initiated on the fourth post-operative day and the patient left the hospital on the seventh post-operative day. After discussing the condition with the patient, a follow up schedule, consisting of 6-monthly visits, was planned with her to diagnose any emerging signs of neurofibromatosis or multiple endocrine neoplasia in the future. Three weeks postoperatively, a colonoscopy was performed that showed to be normal, ruling out any colonic polyps.

DISCUSSION: Neurofibromas are benign neoplasms consisting of proliferations of all the elements in the peripheral nerves, including neuritis and fibroblasts, and the predominance of elongated, serpentine Schwann cells, with their slender, spindle-shaped nuclei. Typically, these components are dispersed in a disorderly pattern, often in a loose, myxoid stroma.^[1,2]

Neurofibromas are usually multiple upon presentation and are usually part of two autosomal dominant disorders with variable penetrance: neurofibromatosis type 1 (NF1, von Recklinghausen's disease) and neurofibromatosis type 2 (NF2, central or bilateral acoustic neurofibromatosis).^[3] However, approximately half the cases of NF1 and NF2 arise from new mutations.^[4] These disease entities have variable clinical expressions with manifestations involving the skin, nervous system, eyes, bones, gastrointestinal tract (GIT), vascular system and other body parts.^[3] While NF1 is more common than NF2, NF1 is characterized by cutaneous manifestations as café-au-lait spots and axillary freckling along a large number of nervous system tumours. On the other hand, the hallmark of NF2, and as its name suggests, is bilateral vestibular schwannomas in over 90% of patients, in to other nervous system tumours. Gastrointestinal involvement in neurofibromatosis is an uncommon entity.^[5] While the neurofibromas do not typically affect the tract in NF2, these lesions are the most common abdominal neoplasms

Encountered in NF1, affecting the GIT in 10–25% of patients.^[6] Ganglioneuromatosis and neurofibromatosis are the pathologic forms of gastrointestinal involvement.^[7]

Neurofibromas of the GIT are usually originating from either the plexus of Meissner in the submucosa or the plexus of Auerbach in the muscularis propria or even from the serosa.^[8,9] These lesions are often sessile and wide-based but also pedunculated polyps have been observed.^[8] Ganglioneuromatosis, on the other hand, refers to extended hyperplasia and

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hypertrophy of the nerve plexuses and ganglion cells in the mucosa or throughout the bowel wall,^[3] this may lead to mural thickening and eventually stricture formation.^[6] Similar lesions could be found within the bowel mesentery, these lesions may be grossly mistaken with lymph nodes.^[6] Characteristic neurofibromas have been found in the GIT in 11% of patients with NF1,^[10] according to some reports. Multiple neurofibromas are more often discovered in the jejunum, stomach, ileum, duodenum and colon according to the frequency of their appearance.^[3,10] To note, further, that NF1 is also associated with the occurrence of other neoplasms that involve the GIT, including carcinoids, somatostatinomas, leiomyomas, sarcomas and pancreatic adenocarcinomas.^[7,10]

Moreover, neurofibromas and ganglioneuromas of the GIT have also been reported in multiple endocrine neoplasia type 2b (MEN 2b), juvenile and adenomatous colonic polyposis.^[11] Consequently, the presence of gastrointestinal neurofibromatosis in association with NF1, and probably other syndromes, is not a rare clinical entity. However, it is rarely encountered as a separate pathologic entity and reports of isolated findings of neurofibromatous proliferations in patients with no additional clinical evidence of, intestinal polyposis or multiple endocrine neoplasia syndromes have been rarely documented.^[12] In these settings, isolated intestinal neurofibromatous proliferations may be the initial manifestation of NF1 or MEN 2b.^[13] The clinical presentation of isolated neurofibromatous lesions of the intestines are myriad and are dependent upon the focal or diffuse nature of the lesions, their location, their effect on gastrointestinal motility and their possible impingement on adjacent structures, resulting in abdominal pain, palpable masses, bleeding due to necrosis or ulceration of the mucosa, obstruction due to intussusception or extra-luminal pressure, diarrhoea, perforation, obstructive jaundice and obstruction of the pancreatic duct, 3 among others.^[14,15] A demonstrable aetiology is found in nearly 90% of cases in the adult population^[16] of which could be isolated intestinal neurofibromas.

Radiologically, the differential diagnosis of single or multiple nodular neurofibromatous lesions is wide and includes many epithelial and stromal neoplasms as well as nodular lymphomas.^[13] Yet, diffuse intestinal ganglioneuromatosis may mimic, radiologically, Crohn's disease,^[17,18] intestinal lymphoma or carcinoid tumour with diffuse infiltration of the bowel wall.^[6] The endoscopic appearance of these lesions depends on the focal or diffuse nature of the lesions. Most lesions can be approached endoscopically, and endoscopic biopsies are a mainstay of the diagnosis. But as neurofibromas arise deep to the epithelium, the biopsies may yield only unaffected overlying bowel mucosa or minimally diagnostic superficial lesional tissue mainly when lesions are small in size.

Macroscopically, these tumours are firm and solid in consistency, and white to tan in colour. Haemorrhage and necrosis are exceptional, but superficial ulceration has been reported^[13] in both focal and diffuse entities.^[19] Endoscopic resection of colonic lipomatous polyps and laparoscopic resection of benign bowel tumours causing ileal and/or ileocolic intussusception has a role in very selected settings,^[20] care must be taken during trocars insertion and insufflation specially in cases of intestinal dilatation in order to avoid bowel injury. So, the primary therapeutic option of isolated neurofibromatous proliferations of the intestines is surgical,^[16] depending on the location and size of the lesions.

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For asymptomatic, incidental findings during endoscopy, no further treatment may be required.^[13] Otherwise, resection of the lesions is dictated by patient's symptoms and operability. In all cases, a correct diagnosis has considerable implications for further management as the bowel involvement could be the first manifestation of neurofibromatosis.^[21] A similar case of an isolated mesenteric neurofibroma has been reported by Margo et al. 123 in 1980. The patient presented, however, with an ileocolic intussusception. Moreover, single or multiple neurofibromas have rarely been reported in literature. These lesions were found in the soft palate, oesophagus, stomach, gallbladder, common bile duct, small bowel and the mesentery, colon and the anal canal with no evidence of associated systemic disease.^[22] The presence of isolated neurofibromas outside the GIT has also been documented. These rare cases involved the kidney, spermatic cord, nasal cavity, palatine tonsil, parapharyngeal space, larynx, humerus, submandibular salivary gland, conjunctiva, retroperitoneal space, cranial ventricles and chest wall.

CONCLUSION: Isolated mesenteric neurofibroma is a rare pathological entity. The clinical significance of such a diagnosis lies mainly in the need of further follow up of these patients as the bowel involvement could be the first manifestation of neurofibromatosis type 1 or multiple endocrine neoplasia type 2b.



FIG. 1

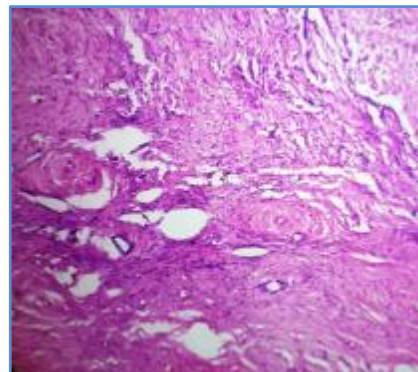


FIG. 2

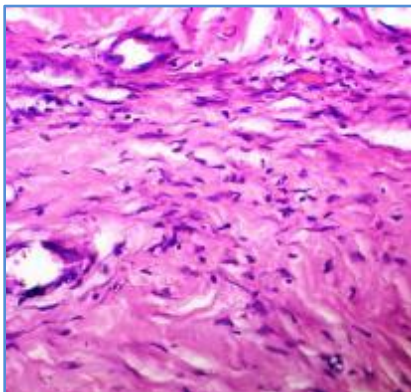


FIG. 3



FIG. 4

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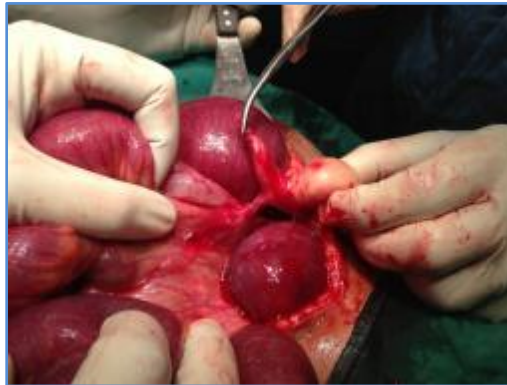


FIG. 5

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