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Solitary intestinal neurofibroma with no associated systemic syndromes causing intussusception: Case report and literature review

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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 48-year-old lady, with no history of neurofibromatosis or other systemic disease, presented with small bowel obstruction secondary to an ileo-ileal intussusception induced by an isolated ileal neurofibromatous mass. The patient underwent a segmental enterectomy 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 ileal 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.

1. 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. Herein, we report the case of an isolated ileal neurofibroma in a middle-aged lady, presenting as an ileo-ileal intussusception. A review of the medical literature on this rare entity follows.

2. Report of a case

2.1. History

A 48-year-old lady presented to our hospital with a 10-day history of intermittent, colicky, epigastric abdominal pain radiating occasionally to the whole abdomen, exacerbated by food intake. She is known to suffer from peptic ulcer disease (PUD) with a small, sliding-type, hiatal hernia. She has undergone laparoscopic cholecystectomy and appendicectomy several years before and, upon presentation, was on no medications. After normal chest X-ray, abdominal computed tomography (CT) scan and laboratory studies, an upper gastrointestinal endoscopic exploration showed severe, diffuse gastritis with antral and prepyloric erosions. A gastric biopsy showed non-atrophic glands and moderate chronic inflammation with few crypts showing *Helicobacter pylori* (*H. pylori*) organisms, suggesting type B gastritis.

So, the patient was diagnosed as suffering from severe gastritis and was started on *H. pylori* eradication therapy; however, she showed no symptomatic improvement. Three days later, while still at the hospital, the patient's abdominal pain worsened and she started to develop global abdominal distension with clinical deterioration suggesting a distal gastrointestinal obstruction. An erect abdominal X-ray showed multiple air/fluid levels. Feaculent material drained upon inserting a naso-gastric tube. A repeat, urgent abdominal CT scan (Fig. 1) showed severe distension of the stomach, duodenum, jejunum and ileum with fluid stasis due to intestinal intussusception at the level of the terminal ileum caused by an ileal tumour resulting in this obstructive syndrome.

Thence, the surgery team was consulted and surgical exploration was decided in view of the patient's presentation and worsening clinical condition. During the laparotomy, around 500 cc of clear ascitic fluid was drained and largely distended jejunal and ileal loops were encountered. Upon running the small bowels, an ileo-ileal intussusception (Fig. 2) at some 60 cm from the ileocecal valve was identified and reduced and an intra-luminal mass could easily be palpated, the mesentery looked normal with no evidence of vascular compromise. No lymph nodes or other palpable intra-luminal masses could be discovered. A segmental entrectomy, including about 10 cm of the ileum from either side of the mass along with the associated mesentery was performed. Continuity of

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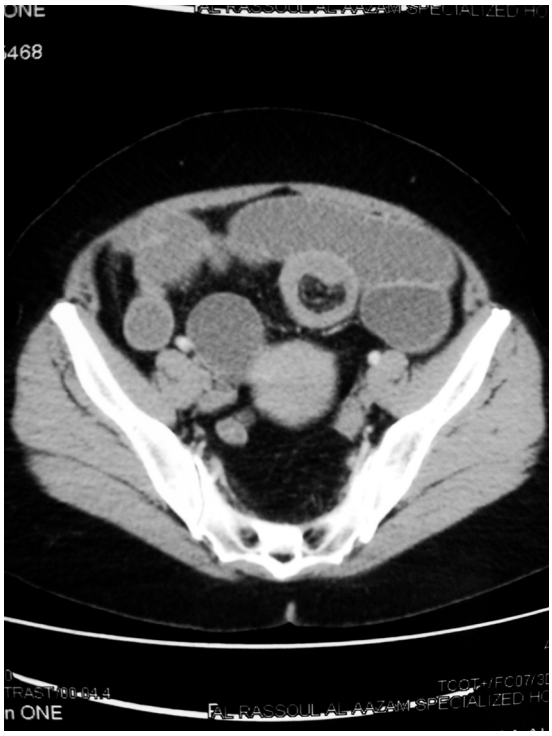

Fig. 1. Computed tomographic scan image. Computed tomographic scan image showing the site of ileal intussusception (target sign) and grossly dilated small bowel loops.

the bowels was re-established by a hand-sewn, side-to-side anastomosis.

2.2. Pathology

Macroscopically, the ileal segment measured 24 cm × 4 cm × 5 cm. When the bowel loop was incised, a well-delineated, intra-luminal mass arising from the bowel wall was identified measuring about 3.5 cm × 3 cm × 3 cm, this mass was spherical and pedunculated, having a smooth surface with a hard consistency; grossly, no hard or infiltrative areas related to the mass could be identified (Fig. 3). The pathology report revealed a normal intestinal wall except for a central, well-delineated, fibrous, spindle-cell neoplasm in different planes consistent with a neurofibroma.

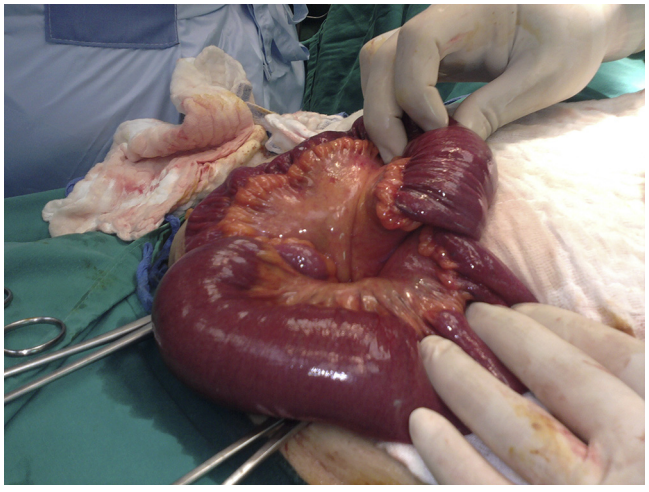

Fig. 2. Intra operative picture. The site of the ileo-ileal intussusception at some 60 cm proximal to the ileo-cecal valve.

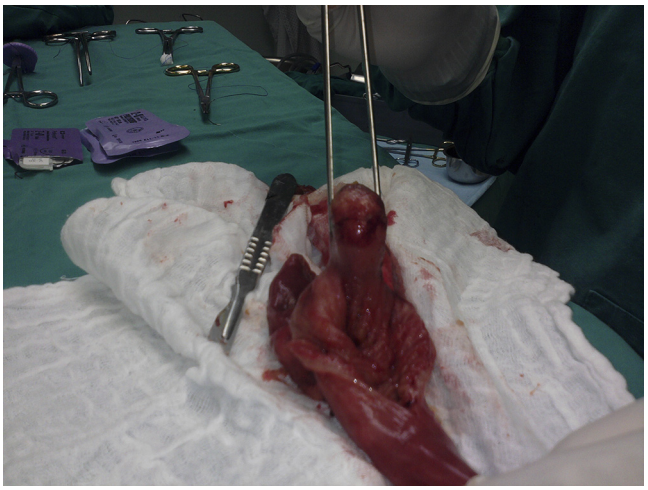

Fig. 3. Macroscopic view. A macroscopic view of the ileal neurofibroma.

2.3. 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 post-operatively, a colonoscopy was performed that showed to be normal, ruling out any colonic polyps.

3. Discussion

Neurofibromas are benign neoplasms consisting of proliferations of all the elements in the peripheral nerves, including neurites 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 characterised by cutaneous manifestations as café-au-lait spots and axillary freckling along with 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 addition to other nervous system tumours.[4]

Gastrointestinal involvement in neurofibromatosis is an uncommon entity.[5] While the neurofibromas do not typically affect the gastrointestinal tract in NF2,[5] 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