Neuromuscular and vascular hamartoma as an unusual cause of small bowel obstruction

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A B S T R A C T

INTRODUCTION: Neuromuscular and vascular hamartoma (NMVH) is a rare, controversial lesion of the intestine, with only 23 cases reported in the English literature since its initial description in 1982.

PRESENTATION OF CASE: A 59 year old female suffering from longstanding Crohn’s disease with chronic stricture presented with symptoms of small bowel obstruction. Contrast studies demonstrated massive dilatation of the proximal small bowel. Laparotomy identified a 5 cm long stenotic segment of ileum, with grossly distended jejunum and ileum proximally. Pathology determined the stricture’s aetiology as a neuromuscular and vascular hamartoma of the small intestine.

DISCUSSION: NMVH is a benign lesion of hamartomatous origin. Its very existence is questionable due to histological similarities with several reactive pathologies, such as Crohn’s and diaphragm diseases.

CONCLUSION: NMVH could be confused with a spectrum of chronic inflammatory bowel conditions, but this report establishes it as a distinct cause of chronic bowel obstruction.

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1. Introduction

Neuromuscular and vascular hamartoma (NMVH) is an uncommon lesion of the small intestine, often presenting clinically as non-specific abdominal pain, recurrent obstructive symptoms, or occult gastrointestinal bleeding.

First described by Fernando and McGovern in 1982, this benign lesion consists of an aberrant proliferation of muscular, neural and vascular elements indigenous to the small intestine [1]. Since its initial description, there have been only 23 cases reported in the English literature [1–6]. NMVH can occur as single or multiple strictures, or single or multiple polypoid-like mass lesions [2,3], affecting a variable length of bowel. Generally arising from jejunum or ileum, one reported NMVH originated from the caecum [4]. Radiologically, the involved bowel may demonstrate findings consistent with a stricture, intussusception, or a polypoid mass. No age or gender predilection appears to exist, with one case of NMVH having been reported in a 12-year-old child [5]. Surgical resection is curative [5], and there have been no documented recurrences of NMVH.

Similar histological features have been seen in association with a range of chronic reactive conditions, and it has been suggested that the NMVH is not a distinct entity, but is in fact a ‘burnt out’ form of Crohn’s disease or another chronic inflammatory process [2,3,6]. NMVH-like proliferations have been described in relation to diaphragm disease, radiation enteritis, ischaemic enteritis, and Crohn’s disease.

2. Case presentation

A 59 year old woman presented for specialist outpatient review with chronic small bowel obstruction. She described a ten year history of increasing abdominal pain, distension and persistent vomiting after oral intake. The patient had a past medical history significant for Crohn’s disease diagnosed 16 years earlier. She had undergone two previous laparotomies, with an ileocolic resection and subsequent ileal resection for Crohn’s fistula and perforation, respectively. The patient had also developed Crohn’s related cirrhosis with a variable coagulopathy and bouts of hepatic encephalopathy. A pre-operative abdominal ultrasound revealed ascites, portal hypertension and splenomegaly. There was no history of non-steroidal anti-inflammatory drug (NSAID) use or radiation exposure.

A pre-operative small bowel series demonstrated massive chronic dilatation of jejunum and ileum, with extremely slow transit of contrast medium and collapsed bowel distally, indicating almost complete small bowel obstruction (Fig. 1). Small bowel
obstruction due to either adhesions or Crohn’s stricture was suspected.

The patient was admitted to hospital 2 months pre-operatively with acute decompensation of chronic liver disease and metabolic encephalopathy. Surgery was scheduled once the encephalopathy had resolved with lactulose, rifamixin, and protein load reduction, ascites was minimized with diuretics, and coagulation studies had normalized. At operation, a midline laparotomy was performed. Extensive serosal adhesions were divided and a transition point identified in the ileum. Small bowel proximal to this point was grossly distended to a diameter of 12 cm. The transition point was a 5 cm long stenotic segment of ileum, not typical of Crohn’s disease. This segment was resected, along with a 15 cm segment of the pre-stenotic ileum. A stapled anastomosis was performed using a side-to-side, functional end-to-end technique. The patient’s post-operative course was uneventful. The abdominal distension resolved slowly and the obstructive symptoms settled.

Macroscopically, the resected section of ileum displayed an area of annular stenosis 45 mm in length, and a luminal diameter reduced to 9 mm (Fig. 2). The specimen displayed serosal adhesions at the level of the stenosis. Histopathological analysis of the stenosed portion of small bowel demonstrated features consistent with NMVH of the small bowel. Within the submucosa, there was focal thickening due to disorganized bundles of smooth muscle, proliferation of peripheral nerve bundles, thick-walled blood vessels, and scattered plasma cells and mast cells. Focally ectatic lymphatic vessels were present (Fig. 3). The mucosa appeared unremarkable, without fissures or cobblestone formation. No granulomas were present, and there was no evidence of malignancy. The resection margins of the specimen displayed normal small bowel mucosa.

One year following laparotomy, the patient remains well.

3. Discussion

NMVH is a benign, non-epithelial hamartoma: a submucosal proliferation of mature, although disorganized, elements normally found in the small bowel. These lesions demonstrate haphazard, aberrant fascicles of smooth muscle from the muscularis mucosa, bundles of non-mylinated nerve fibres with scattered abnormal ganglion cells, and hemangiomatous vascular channels [1,2]. Debate exists regarding the hamartomatous nature of this disorder, given that many of the features of NMVH are seen in a range of reactive conditions, including Crohn’s disease, ischaemic enteritis, radiation enteritis and NSAID-induced small bowel strictures (so-called ‘diaphragm disease’) [2,6,7].

Shepherd and Jass have questioned the hamartomatous origin of NMVH, as similar pathological features may be seen in Crohn’s disease. They described four cases, three of which involved an unequivocal diagnosis Crohn’s disease, and all of which displayed histological features similar to NMVH. They suggested that NMVH may represent “an unusual histologic consequence of inflammatory bowel disease, predominantly Crohn’s disease” [6]. However, their cases showed an absence of fissuring ulceration and transmural inflammation, both of which are typical hallmarks of active Crohn’s disease. They suggested that the particular pathological features seen in their four cases may be those of a chronic, ‘burnt out’ form of Crohn’s disease [3,6].

The pathological changes seen in Crohn’s disease can be varied and non-specific, but the distinctive features are transmural chronic inflammation, non-caseating granulomas, and lymphoid aggregates within the bowel wall. The mucosa typically demonstrates a cobblestoning appearance of fissures and oedematous mucosa. In the advanced stages of Crohn’s disease, neuromuscular hypertrophy, submucosal fibrosis and serosal fibrosis, are commonly seen [2]. Occasionally, the transmural inflammation and fissuring which are classically described with Crohn’s disease may be absent, and connective tissue changes (such as fibrosis, muscular hypertrophy, neuronal and vascular changes) represent the only remaining elements of a chronic ‘burnt out’ disease process. It is worth noting that only 5 out of the 23 documented cases of NMVH exhibited prominent fibrosis on histological examination [2,3], suggesting that the remaining 18 cases without significant fibrosis are unlikely to represent a chronic reactive inflammatory condition, or a burnt-out form of Crohn’s disease.

Prominent fibrosis is also a hallmark feature of ischaemic and radiation small bowel strictures, as well as diaphragm disease [2,6]. Fibrosis was not a feature in the case presented here.
NMVH is indeed a very rare lesion. Its very existence is controversial. The debate is ongoing whether this is truly a distinct hamartomatous lesion, or a non-specific reactive phenomenon. However, it is clear that the NMVH is an abnormal, non-neoplastic proliferation of elements normally found within the submucosa of the gastrointestinal tract, and as such, it does fit the criteria of a hamartoma. NMVH has certain features that mimic those commonly associated with inflammatory bowel disease and other reactive conditions, but not closely enough to be conclusive of those disorders. We believe that in the absence of conclusive historical features of Crohn’s disease, or clinical suspicion of diaphragm disease, radiation or ischemic enteritis, NMVH must be considered in the differential diagnosis of stricturing and mass-forming intestinal lesions. In the case presented here, there were no clinical, macroscopic or histological features that were indicative of active or ‘burnt out’ Crohn’s disease, or another reactive process, and we are therefore confident that this is an example of a neuromuscular and vascular hamartoma.

Conflicts of interest

All authors declare no conflict of interest.

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Ethical approval

Ethics committee approval was not required for this case report.

Consent

Written and signed informed consent from the patient has been obtained.

Author contribution

Dr. Kristen Elstner—primary author.
Dr. Rohan Geet—operating surgeon, supervision of project and editing of paper.
Dr. Louise Killen—pathology images, description of pathological process.
Dr. Mih Ru Qiu—supervision and editing of pathology images, written pathology descriptions.
Dr. Christopher Vickers—supervision and editing of paper.

Guarantor

Dr. Kristen Elstner.

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