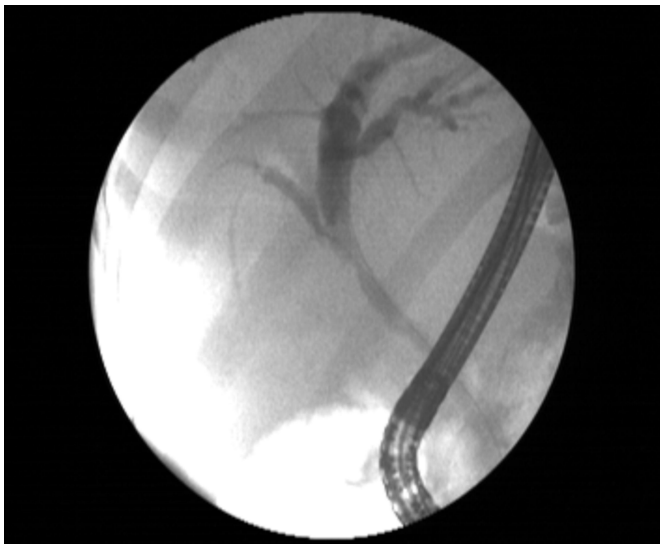

Figure 4: ERCP showing biliary stricture at the upper end with disproportionate left side biliary dilatation and disappearance of cyanoacrylate

Discussion

Duodenal varices result from retroperitoneal shunts caused by increased blood flow between the cystic branch of the superior mesenteric vein, superior and inferior pancreatic veins, and retroduodenal veins. Endoscopic cyanoacrylate injection has been shown to be a promising treatment option for bleeding duodenal varices.[1]Several complications have been reported with the injection of cyanoacrylate into gastric varices, especially embolization of the glue into the portal, pulmonary, coronary and cerebral veins in up to 5% of cases.[2,3] The risk of embolism appears to be related to the diameter of the vessel involved, pattern of blood flow, the volume injected and the rate of injection.[2,3] Injecting cyanoacrylate without diluting it with lipiodol—thus using a lesser volume—might have decreased the spillage into paracholedochal varices.

The paracholedochal veins course parallel to the CBD and are connected to the gastric veins, the pancreaticoduodenal vein, the portal vein and to the liver. The postulated mechanisms of biliary stricture in portal biliopathy are mechanical compression of the biliary tree by the veins and ischaemic injury to the CBD.[4,5]

In our patient, biliopathy possibly resulted from ischaemic changes secondary to cyanoacrylate casts in the paracholedochal veins and collaterals. Another possibility is mechanical compression of the CBD by these casts. A third possibility is spillage of a small amount of cyanoacrylate directly into the CBD. The possibility of asymptomatic portal biliopathy predating the injection of cyanoacrylate cannot be ruled out, since a majority of patients with portal biliopathy reported in the literature remain asymptomatic.[6] However, the absence of biochemical and sonographic evidence of the biliary obstruction before the injection therapy and sudden appearance of these after, along with the demonstration of radio-opaque casts, suggests a cause–effect relationship between the two. As biliary strictures persisted in our patient, even after 1½ years, by which time the casts had disappeared, ischaemia is the most likely mechanism involved. The possibility of spillage of cyanoacrylate into the CBD is less likely as the strictures are at the upper end of the CBD and they persist even after 1½ years of follow-up.

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Neuromuscular and vascular hamartoma of small bowel with prominent inflammatory changes

Introduction

Neuromuscular and vascular hamartoma (NMVH) is a rare, benign, non-neoplastic lesion mostly affecting the small intestine. It consists of aberrant arrangement of non-epithelial tissue normally occurring in the intestine and has been considered as a hamartoma.[1] The hamartomatous nature of this disorder has been questioned by many authors because identical features may be seen in Crohn's disease,[2] non-steroidal anti-inflammatory drugs (NSAIDs)-associated small intestinal diaphragm disease,[3,4] ischaemic enteritis and radiation enteritis.[1]

Case report

A 28-year-old woman presented with abdominal pain and recurrent vomiting for 1 month duration. On enteroscopy, a single stricture was identified in the jejunum. Barium meal follow-through revealed a stricture causing compression in the mid jejunum. The rest of the bowel was unremarkable. No significant past medical or surgical history was present. The patient underwent a segmental resection with anastomosis. Gross examination revealed a 16 cm segment of the small bowel with a single, well-delineated stricture measuring 2.5 cm × 2 cm. The overlying serosa on the stricture was dusky blue with congestion, haemorrhage and prominent vessels (**Figure 1**). On opening, there was a circumferential stricture with ulcerated and congested overlying mucosa. Cut section through the stricture revealed thickened and fibrotic wall. Microscopically, focal superficial erosion and ulceration of the mucosa was seen with single fissure formation extending into the submucosa (**Figure 2A**). The lamina propria was widened with dense lymphoplasmacytic inflammation admixed with sparse eosinophils and neutrophils (**Figure 2B**). Focal crypt abscesses were observed. However, no architectural disruption was evident. Transmural lymphoid hyperplasia was present without any granuloma formation. The most conspicuous findings were present within the submucosa and subserosa. The submucosa was expanded with neural hyperplasia and presence of large variably sized aggregates of ganglion cells (**Figure 2C**). Haphazard intertwined proliferating bundles of smooth muscle fibres appear to arise from muscularis mucosae were seen (**Figure 2D**). There were numerous disorganized ectatic blood vessels recognized within the submucosa and subserosa (**Figure 3A**). Blood vessels were of variable calibre and showed variable thickness of their walls. No vasculitis or

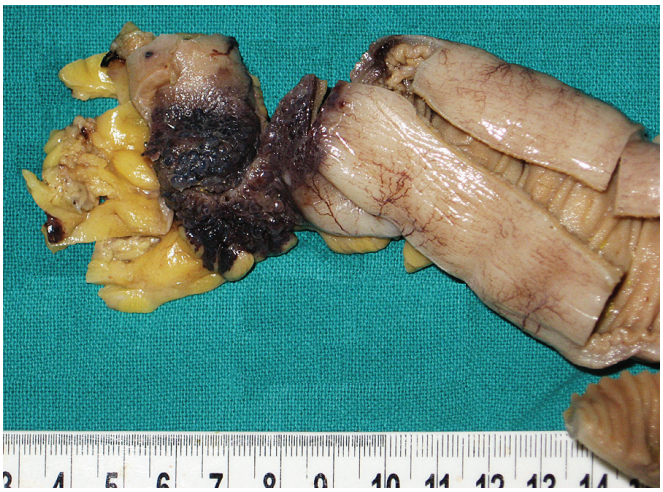

Figure 1: Segment of jejunum with a well-delineated stricture and prominent vessels on the serosal aspect

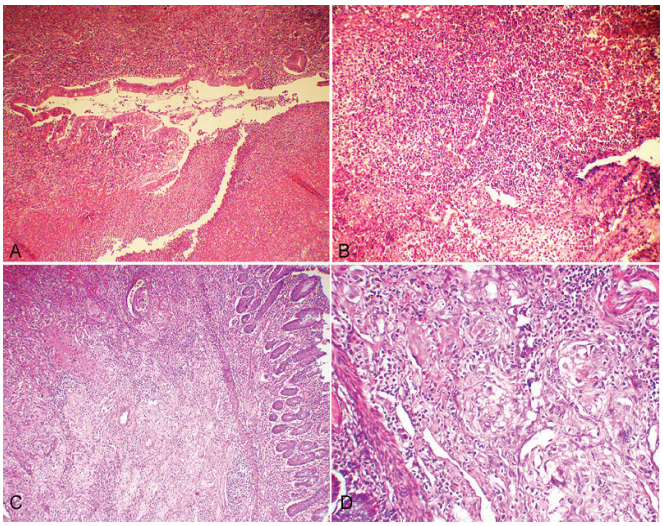

Figure 2: (A) Surface mucosal ulceration and fissure formation, H&E x40; (B) dense mixed inflammatory infiltrate is present in the lamina propria, H&E x200; (C, D) disorganized smooth muscle fibres, prominent nerve bundles and clusters of ganglion cells within the submucosa H&E x40 and H&E x400

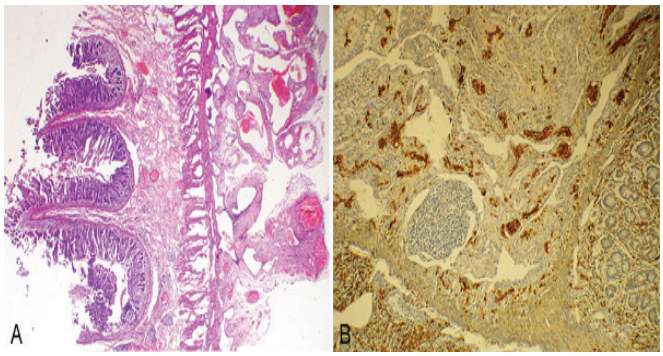

Figure 3: (A) Ectatic variably thickened vessels present within the submucosa and serosa H&E x20 (B) S-100 immunostain highlights aberrant non-myelinated nerve fibres within the wall of the intestine

thrombosis was seen. The non-strictured intestinal segment and resected edges were within normal limits. The mesenteric fat showed reactive lymph nodes. S-100 immunostain highlighted proliferating hypertrophied nerve bundles (**Figure 3B**), whereas CD34 and factor VIII antibodies showed vascular channels. Smooth muscle actin was present in the submucosa, blood vessels and hypertrophied muscularis mucosae. CD68 immunostain did not confirm the presence of either histiocytes or ill-formed granulomas.

A diagnosis of NMVH was made in view of the short history of upper gastrointestinal (GI) symptoms, a single isolated jejunal stricture, endoscopically normal distal bowel, absence of classical hallmarks of Crohn's disease, viz. skip lesions, prominent fissuring ulcers, granulomas, and crypt architectural disruption with conspicuous mural changes in the form of neuromuscular and vascular proliferation. A comment was made with regard to the prominent inflammatory changes and follow-up of the patient was advised. After 2 year of follow-up, the patient was doing well.

Discussion

NMVH was first described in two women patients by Fernando and Mcgovern in 1982 as a specific hamartomatous condition of the small intestine.[1] Since then, 9 more cases have been reported in which it is also described as a distinct entity. NMVH occurred in the small intestine in all except three cases—one in the cecum and other two in Meckel's diverticulum.[5–7] All the cases lacked manifestation of any multisystem disease such as neurofibromatosis or features of inflammatory bowel disease or prolonged intake of NSAIDs.

Patients with NMVH usually present with a history of recurrent episodes of abdominal pain with vomiting or rarely with constipation.[1,6,8] They may present with anaemia due to chronic blood loss from the GI tract.[1,9] There is no known age or sex predilection. Endoscopic, barium studies of the small bowel and CT studies show single or multiple strictures without any demonstrable mechanical cause.[8] Two of the 11 cases had a previous history of surgical procedure and one had a history of treated tuberculosis and radiotherapy, thus raising a question whether NMVH is a post-traumatic or post-inflammatory lesion?[1,6]

In agreement with Fernando and Mcgovern,[1] we diagnosed the present case as a hamartomatous lesion independent of any systemic disease. However, Shepherd and Jass[2] proposed NMVH as a part of inflammatory bowel disease and suggested that the pathological features may be those of a chronic burnt-out form of the disease. Although we observed prominent inflammatory changes with transmural lymphoid hyperplasia, all classical features of Crohn's disease were lacking. The exact pathogenesis of the disease is not known. The reported association with inflammatory bowel disease and NSAID-associated small intestinal diaphragm disease indicates that it could be a non-specific reactive lesion.[2–4]

Grossly solitary or multiple strictures or a polypoidal mass have been reported. Microscopically, the abnormalities are seen mainly in the submucosa and subserosa. Mucosa over the narrowing reveals ulcerations or erosions. The lesion consists of bundles of non-myelinated nerve fibres with clusters of ganglion cells, fascicles of smooth muscle fibres merging with muscularis mucosae and hypertrophied muscularis propria. Ectatic variable sized, thick and thin-walled vessels were seen.[1,6,8] The vessels may extend both into the serosa and into the lamina propria. Histopathological changes need to be correlated with clinical and endoscopic findings.

The differential diagnosis of NMVH is discussed in **Table 1**. The salient features which distinguish NMVH from other overlapping disorders are absence of granuloma or fissure formation or transmural inflammation as seen in Crohn's disease; granulomas (fresh or hyalanized) in tuberculosis; dense fibrosis in radiation and ischaemic enteritis; clinical history of prolonged use of NSAIDs in diaphragm disease. Peutz–Jeghers hamartomas may enter into the differential diagnosis, but differ from NMVH by the characteristic branching core of a muscular tissue derived from the muscularis mucosae expanding toward the luminal surface.

As shown in the differential diagnoses, there are only subtle differences between Crohn's disease and NMVH. In the present case, in contrast to the previously reported cases, prominent inflammation was seen along with mucosal ulceration and focal fissure formation. Transmural inflammation was absent though reactive lymphoid follicles were dispersed throughout the wall of the intestine. Few lymphoid aggregates were present in the wall of ectatic vessels giving an appearance of the lymphangiomatous process. However, in this case, clinical and imaging findings were not in favour of inflammatory bowel disease. The patient had only one stricture and remained well after surgery on follow-up. Therefore, the presence of inflammatory changes cannot preclude the diagnosis of NMVH, especially if clinical and endoscopic findings are correlated

Table 1: Differential diagnoses of NMVH

	NMVH	Crohn's disease	Tuberculosis	NSAID (Diaphragm disease)	Ischaemia/Radiation	Neurofibromatosis / Sipple disease
Common site	Small bowel/ cecum	Small bowel	Ileocecal	Small bowel	Small and large bowel	Multiple organs
Gross	Single/Multiple strictures	Multiple strictures, cobblestone appearance	Transverse stricture	Superficial ulceration with stricture	Stricture	
Nerve fibres	++	++	+	++	–	++
Ganglion cells	++	++	+/–	++	–	+
Muscularization of submucosa	++	++	+/–	++	–	–
Granulomas	–	+/–	+	–	–	–
Vascular changes	++	+	+	++	+	–
Mucosal ulceration	+	+/–	+	+	+/–	–
Fissuring	+/–	+	+	–	+/–	–
Transmural inflammation	–	+	–	–	–	–
Fibrosis	–	+	++	+	++	–
Smooth muscle fibre drop out	–	–	–	–	+	–
Nuclear atypia	–	–	–	–	+	–
Drug intake	–	–	–	++	–	–

with pathological findings.

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Giant hydatid cyst of the liver

A 65-year-old woman presented with complaints of progressively increasing distension of the upper abdomen associated with mild abdominal discomfort for the past one year. There was no history of fever, jaundice, alteration of bowel habits or constitutional symptoms. Physical examination revealed a large cystic lump in the epigastrium and right hypochondrium, extending up to the right lumbar and umbilical region (**Figure 1**). She had a normal haemogram and the biochemical investigations revealed normal liver and renal function tests. Ultrasound abdomen and contrast-enhanced computed tomography (CECT) scan of the abdomen showed a 19×15×12.5 cm cystic mass in liver segments V, VI and VII of the right lobe, extending into the hepatorenal fossa and compressing the right kidney—suggestive of a large hydatid cyst (Gharbi type I) (**Figure 2**). The diagnosis was supported by a positive serology for antibodies (titre 1:800) to