Muscular Hamartoma of Intestine Causing Intestinal Obstruction

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ABSTRACT

Hamartomatous causes of small bowel obstructionare uncommon and of them, most are attributed to inflammatory bowel diseases and also certain medications such as NSAIDs. We describe a case of muscular hamartoma in a patient without prior chronic medical condition with brief review of literature.

Keywords: *bowel; hamartoma; muscle; obstruction*

INTRODUCTION

Mesenchymal hamartomas of Intestine are rare causes of Intestinal obstruction, first described by Fernando in 1982.[1] Histologically, the unusual appearance and the presence of the mixed vascular, lymphatic, smooth muscular, and adipose tissue components in the identified lesion meets the principles implied in the concept of hamartoma.[2] Rarity of the condition can cause a diagnostic challenge.

We present a case of hamartomatous lesion composed of muscular elements causing intestinal obstruction.

CASE REPORT

A 58 years old male patient was admitted to the surgical ward with complaints of intractable pain abdomen, recurrent vomiting and weight loss with recent abdominal distension since three months with recurrent hospital visits due to his condition. There was no history of per rectal bleeding or prolonged use of NSAIDs. Previous upper gastrointestinal endoscopic examination had revealed multiple erosions with nodular mucosa in antrum of stomach with mild narrowing of pylorus with reactive changes in histopathological examination.

Laparotomy was conducted for "Subacute bowel obstruction". Multiple strictures or mass lesion were identified intraoperatively as has been mentioned on the requisition form.

Specimen was received in 10% formalin in pathology laboratory. The specimen was identified as a segment of small intestine measuring 76cm in length. Breadth varied from 2.5 cm to 6 cm. Serosal surface was smooth with two strictures causing constriction. Mucosa showed focal edema. Mucosa corresponding to the stricture showed narrowed lumen.

Specimen of "gastric mucosa" was also submitted which measured 04x0.4x0.4cm^3, which was grossly unremarkable.

Histopathologically, sections from the strictures of the bowel segments revealed muscularization of submucosa replacing the normal loose connective tissue. The disorganized muscle bundles appeared to be arising from the muscularis mucosa. Prominence of vascular, neural component was not seen. Overlying mucosa

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showed chronic inflammation. Sections adjacent to the strictures showed mucosal inflammation and submucosal edema. Fissuring ulcers, skip lesions, crypt distortion, granulomas was not identified. Sections examined from the gastric mucosal tissue revealed features of Chronic gastritis with muscularization of the submucosa as well.

Masson trichrome and Smooth muscle actin confirmed the submucosal muscularization. S-100 was negative for any neural elements. A diagnosis of Muscular Hamartoma of intestine was made.

DISCUSSION

Fernando first described the hamartomatous condition of the small intestine consisting fascicles of smooth muscle derived from the muscularis mucosae, bundles of unmyelinated nerve fibres with scattered ganglion cells, and haemangiomatous vessels, occurring focally within a segment of the intestine, and causing stenosis.[1] It was termed "Neuromuscular hamartoma of intestine". Some authors have questioned their existence as a distinct entity suggesting their possible relation to Crohn's disease, non-steroidal anti-inflammatory drugs (NSAIDs)-associated small intestinal diaphragm disease, ischaemic enteritis and radiation enteritis.[3–6] A spectrum of histomorphological appearance have been described. The previously reported types of hamartomas of the ileal part of small intestine were neuromuscular and vascular hamartoma (NMVH), neuromesenchymal hamartoma (NMH), myoepithelial hamartoma (MEH), and Cowden hamartomatous syndrome. In present case, haphazardly arranged muscle bundles were seen within the submucosa replacing the normal loose connective tissue. Immunohistochemistry (SMA) showed evidence of muscular elements only and S-100 failed to demonstrate prominent neural component, thus, suggesting a diagnosis of "Muscular Hamartoma". The present case did not show any histological features of Crohn's disease such as transmural inflammation, fissuring ulcers, granulomas and did not have a history of prolonged intake of anti-inflammatory agents.

Patients usually present with complaints of recurrent episodes of abdominal pain, vomiting, constipation consistent with our case.[7–9] There is no age or sex predilection.

Muscular Hamartoma as a distinct entity is underemphasized. With acknowledgement of the diagnosis, its actual incidence rate, etiology and risk factors can be elucidated. Despite its rarity, it is fortunate that "Muscular hamartoma" is benign and remains a reassuring diagnosis for patients, pathologists and clinicians alike.

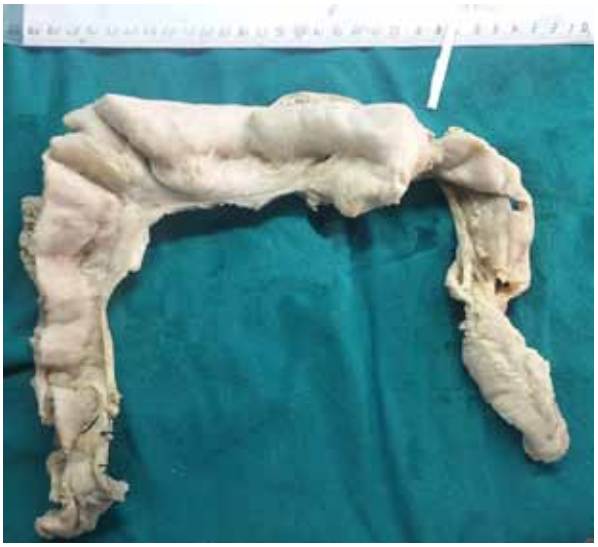

Figure 1. Macroscopic evaluation of Small intestinal segment shows stricture.

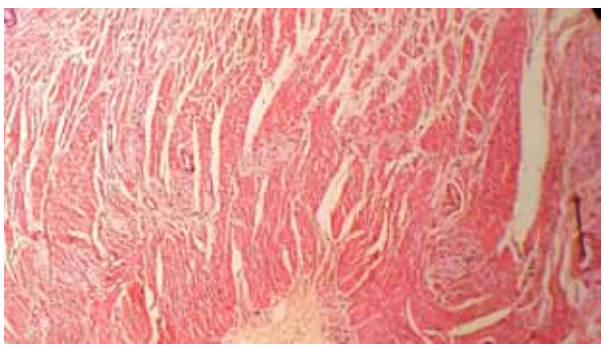

Figure 2. Section from Ileum revealing muscularization of submucosa and mucosal chronic inflammation (H&E; 4x).

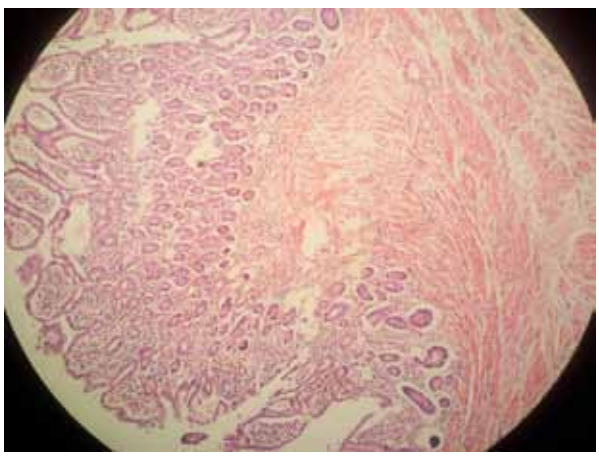

Figure 3. Section from Ileum revealing muscularization of submucosa and mucosal chronic inflammation (H&E; 10x).

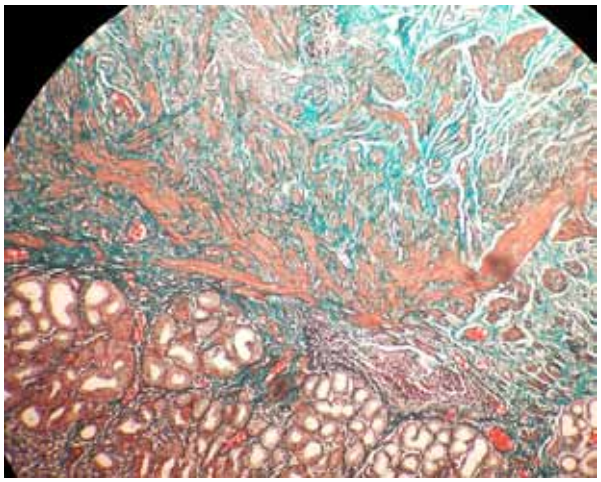

Figure 4.Gastric tissue. Masson trichrome showing positivity for the muscle bundles seen within the submucosa.

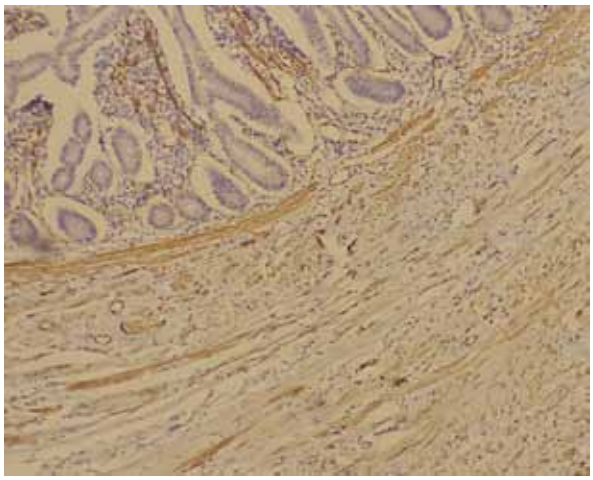

Figure 5. Smooth Muscle Actin positivity for the muscle fascicles within the submucosal layer.

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