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Intestinal Pseudo-obstruction Due to Abnormal Layering of Muscularis Propria: A Case Report of Two Cases and Review of Literature

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Abstract

Chronic Intestinal Pseudo-Obstruction (CIPO) is a rare gastrointestinal disorder which impairs gastrointestinal motility. It can affect adults as well as children. In this report a 3 year old child who presented with a history of mass at rectosigmoid junction is described, along with a 27 year old female who presented with intestinal obstruction. On gross examination, no tumor was identified in both cases. Microscopic examination revealed additional hypertrophic muscular layer; in first case in between inner circular and outer longitudinal layers and in second case external to normal muscularis externa.

Keywords

Gastrointestinal disorder • Chronic intestinal pseudo-obstruction • Tumor

Introduction

Peristalsis is dependent on the contraction and relaxation of the smooth muscle in muscularis propria. Normally there are two muscle layers, namely, inner circular and outer longitudinal [1,2]. Myenteric plexus and ganglion cells are present between the two layers. Any disorder or developmental aberration that effects these structures can lead to CIPO. Thus, these disorders are classified as myopathies, neuropathies and mixed disorders [1-3]. Visceral myopathy of the gut wall is a rare disorder that can present as pseudo-obstruction in infants, children and adults. The disorder mainly presents in the first year of life with symptoms of obstruction including vomiting, abdominal distention and constipation. The disorder usually affects males and is associated with other findings including mega cystitis, mega ureter, and skeletal deformities [1-4]. The disorder can affect both the large and small bowel. Histologically, the disorder can present with intrinsic myocyte defect and morphogenic abnormalities of muscularis propria. Abnormal layering of muscularis propria is extremely rare with only 8 previously reported cases [1].

Case Series

Case 1

A three year old male patient presented with a history of intestinal obstruction and was clinically diagnosed with a 6 cm diffuse mass at the rectosigmoid junction. The affected segment of the colon was resected and was sent to our Hospital. On gross examination, although, no tumor or polyp was found, there was mild diffuse thickening of the bowel wall. Other causes of obstruction including adhesions, volvulus and intussception were also ruled out on gross examination. Extensive sampling of the gut wall was carried out to rule out Hirshsprung disease. On microscopic examination, mucosa was unremarkable, while the submucosa revealed mild congestion. The muscularis propria was found abnormally thickened. Further examination revealed that there was an extra oblique layer present between inner circular and outer longitudinal layers (Figures 1A-1D).

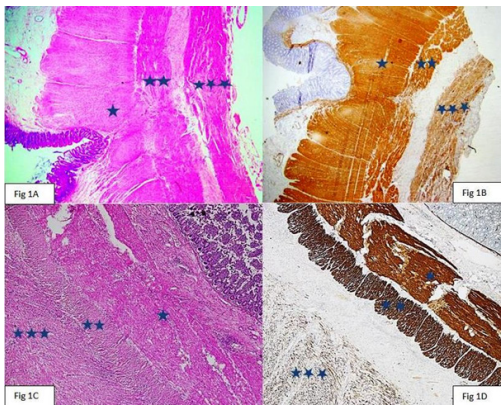

Figure 1. (A) : Demonstrating an irregular oblique muscle layer (*), between inner (*) and outer muscle layer (**), (B): Desmin immunostain highlighting additional irregular oblique layer (*), between inner (*) and outer muscle layer (**), (C): Showing aberrant muscular layer (***) external to normal muscularis propria (inner circular [*] and outer longitudinal layers [**]) and (D): Desmin immunostain highlighting additional muscle layer (**), external to outer longitudinal layer (**).

Case 2

A 27 year old female patient presented with intestinal obstruction. Operative findings were interloop adhesions and pelvic abscess. The affected segment of ileum was resected and sent to our hospital. On gross examination the bowel wall showed alternating thick and thin walled areas. The serosal surface showed fibrinous exudates and mucosa was unremarkable. No discrete tumor was identified grossly. Microscopic examination of thick walled areas showed aberrant oblique muscle layer external to normal muscularis externa. Intervening thinned out areas revealed edema, transmural inflammation and serositis.

Results and Discussion

Chronic Intestinal Pseudo Obstruction (CIPO) refers to impaired peristalsis resulting in intestinal obstruction in the absence of a mechanical cause [1,4]. It is a rare clinical disorder. Based on the cause, chronic intestinal pseudo obstruction falls into three categories, i.e , neurogenic , myopathic and mixed. Neurogenic causes include hypoganglionosis and intestinal neuronal dysplasia. Visceral myopathies may also be divided into two categories. The first category includes myopathies due to intrinsic myocyte defects which show vacuolar degeneration of smooth muscle fibres with thinning and interstitial fibrosis of muscularis propria [1,3,4]. The second category includes developmental abnormalities of muscularis

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propria. Both atrophic and hypertrophic patterns have been reported. Abnormal layering of muscularis propria is rare and only 8 cases have been reported till now. Full thickness biopsies are required for the diagnosis of either the myopathic or neuropathic form of chronic intestinal pseudo obstruction [1].

In the case reported by Angakathunyakul et al. full thickness biopsy of terminal ileum revealed marked thickening of muscularis propria and revealed abnormal layering into three layers 1) inner circular 2) additional oblique and 3) outer longitudinal layer. In one of our cases the affected segment of gut was colon and also showed the presence of additional oblique layer between inner circular and outer longitudinal layer highlighted. In the second case of 27 year old patient, the affected segment was ileum. On microscopic examination, an aberrant oblique muscle layer was present external to normal muscularis externa [1,5-7].

In two cases reported by Kapur and Correa, biopsy from the first patient who was a 19 year old male showed an abnormal layer of cytologically mature smooth muscle in the outer submucosa. The second patient was a six weeks old boy and full thickness sections from the jejunum showed unremarkable mucosa and submucosa but an additional circular layer was present outside the longitudinal layer 3. Smith and Mila presented the term "segmental additional circular muscle coat" in patients with similar findings. A to their study an additional smooth muscle layer was present on the inner surface of circular muscle layer along with an additional neural plexus between the inner two layers 5.

Yamagiwa, et al. analyzed the clinical and histopathological features of an 11 days old neonate who presented with inability to feed along with other congenital anomalies like cryptorchidism and scoliosis. Histopathologically, the small bowel exhibited marked thickening and there was an additional oblique layer outside the longitudinal muscle layer 6. May, et al. reported a case of a 5 month old patient who had an abnormal extra intestinal muscle layer in association with Mowat-Wilson syndrome and Hirschsprung disease [8]. Few of the above mentioned studies also found association with certain other congenital disorders for example, megaureter, megacystis, gut malrotation and congenital Varicella [5-7]. Other common causes of intestinal obstruction in childhood are intussusception, volvulus, bowel or obstruction due to constricting bands [6-8].

Conclusion

There is no specific treatment for atresia and stenosis in individuals with chronic intestinal pseudo obstruction. Treatment is directed to support adequate nutritional needs. Treatment options include dietary modification, pharmacotherapy such as use of pro kinetic drugs. Some individuals are treated by intestinal decompression and rarely in very specific and severe cases, surgical resection of affected portion of intestine is considered.

Conflicts of Interest

There are no conflicts of interest.

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