Segmental absence of intestinal musculature in infant with Hirschprung disease

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BACKGROUND & OBJECTIVES

Segmental absence of intestinal musculature (SAIM) is a rare histopathologic entity with a few published cases in neonatal pathology and still has uncertain pathogenesis. It is classified as primary congenital or secondary acquired. Clinical manifestations usually are intestinal obstruction and perforation. Majority of reported cases are neonates, children, but it can affect adults also.

Figure 1. Gross image of small intestine will thinned wall and peritonitis

METHODS

We present a case of male, 6 months old baby, with previously diagnosed Hirschprung disease when multiple colorectal biopsies were taken and colostomy was made. After 1months he develops fever and vomiting with poor clinic evolution. Intraoperativelly there was diffuse peritonitis and intestinal adhesions due to perforation. A 75cm long segment of small intestine was resected. The wall of intestine was thinned to 1mm (paper-like) with massive hemorrhage and peritonitis. Many specimens were taken for pathohistologic analysis along the whole length of the intestinal segment. The tissue sections were formalin fixed and paraffin embedded and routinely stained with HE. Additional immunohistochemical analysis was performed with SMA and S-100.

RESULTS

All specimens revealed absence of intestinal muscular layer with intact mucosa, muscularis mucosae, submucosa and serosa. There was transmural diffuse acute inflammatory infiltrate and hemorrhage with fibrinopurulent peritonitis. Immunohistochemically SMA confirms the absence of muscularis propria and marks muscularis mucosae. S-100 showed no neuronal plexuses. Resection of the affected specimen is curative. Up to now the baby is in good condition.

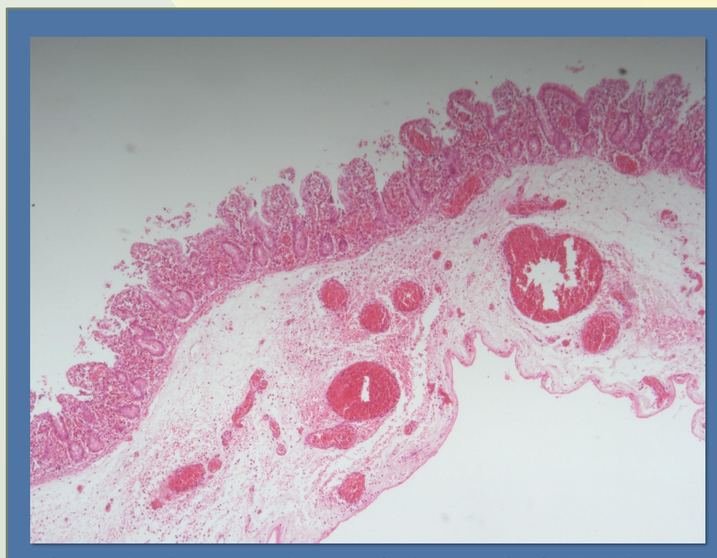

Figure 2. Absence of muscularis propria (HEx50)

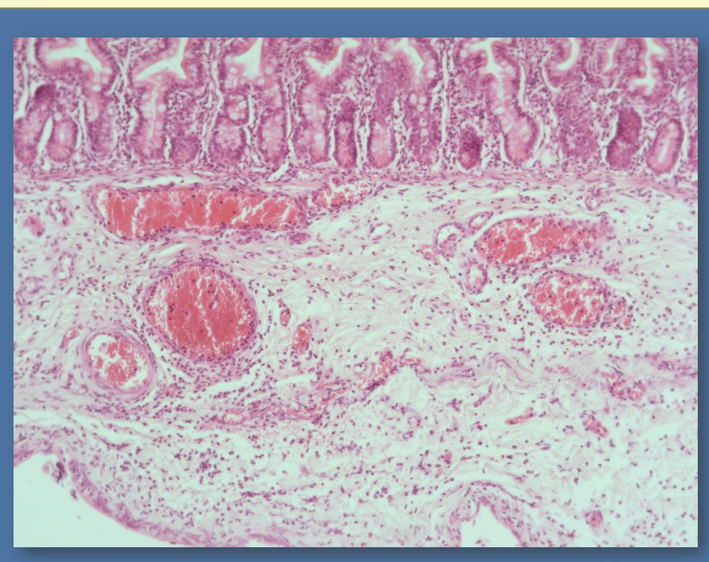

Figure 3. Absence of muscularis propria (HEx100)

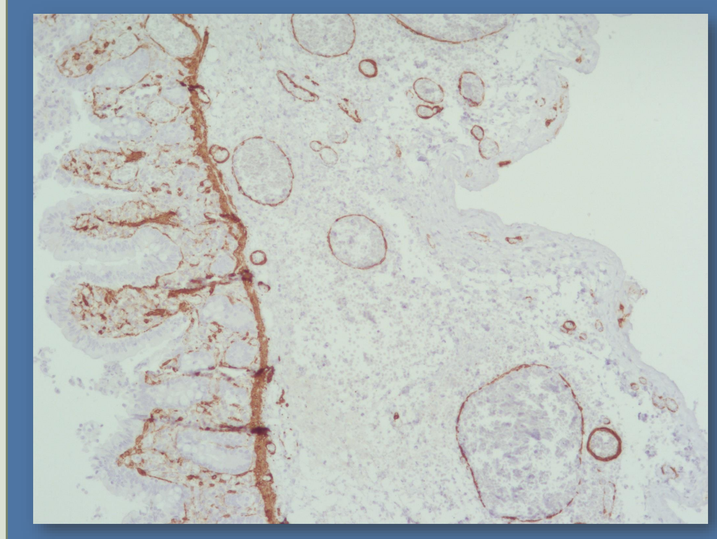

Figure 4. Muscularis mucosae and vessels(SMAx100)

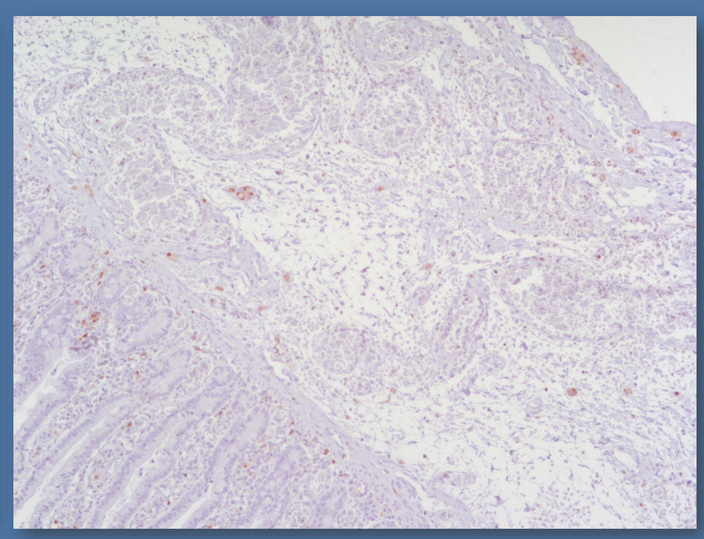

Figure 5.Loss of neural plexuses (S-100x100)

CONCLUSIONS

There is no published case of this condition associated with Hirschprung disease. As etiopathogenesis remains unclear and there is a spectrum of published histopathologic morphologic elements there is a need of homogenous classification for further comparative studies.

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Contact