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IMAGE

Segmental absence of intestinal musculature

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Segmental absence of the intestinal musculature (SAIM) is a rare clinical entity that presents usually in the neonatal period. Our patient was a 3-year-old male who was brought with the complaints of chronic constipation and a 2-day history of non-bilious vomiting, abdominal distension, and fever. He was subjected to exploratory laparotomy and found to have an intestinal perforation. Resection anastomosis of the bowel was done. The surgical pathology specimen consisted of a 30-cm-long piece of small intestine, with thin bowel wall and one area of healed perforation. Histopathological examination revealed a multifocal partial or complete absence of muscularis propria (Fig. 1). The mucosa and muscularis mucosa appeared unremarkable. The submucosa and serosa were edematous and infiltrated by lymphocytes and plasma cells. Foci of hyperplasia of the muscularis propria were seen. The myenteric plexus was not seen in areas of absence of muscle but present where muscular layer was present. Diagnosis of SAIM is made only after pathologic examination of the resected bowel [1]. The usual presentation is consistent with peritonitis, although obstruction may also be a presentation. SAIM may be either congenital or acquired, the latter possibly secondary to ischemic injury [2].

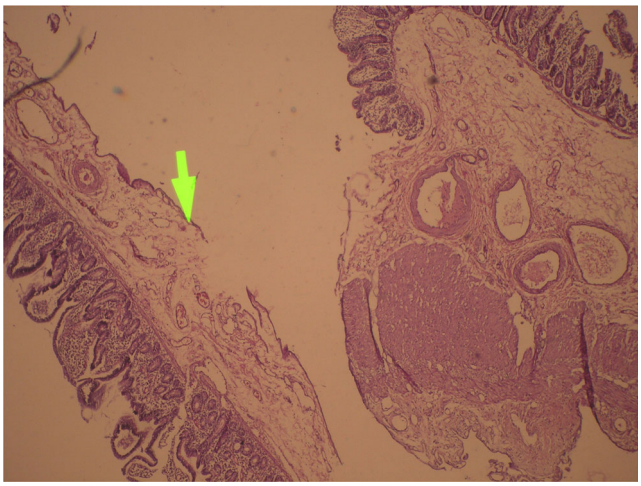

Fig. 1 Microphotograph of small intestine showing an absence of muscularis propria in one section (*arrow*); other sections are showing hypertrophy of muscularis propria (H&E, ×40)

References

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