Association between Visceral Myopathy and Hirschsprung's Disease: A case report. Authors: María Jesús Nally R. (1), Rodrigo Maluje J. (2). 1. Pediatric Surgery Resident Exequiel González Cortés Hospital, University of Chile, Santiago, Chile. 2. Pediatric Surgeon Exequiel González Cortés Hospital, University of Chile, Santiago, Chile.

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Abstract

Background: Visceral myopathy (VM) is an etiology of chronic intestinal pseudo-obstruction syndrome (CIPO) and is characterized by intestinal dysmotility secondary to vacuolar degeneration, atrophy and fibrosis of the muscular propria layer of the intestinal wall, without inflammatory cells. The diagnosis is suspected clinically and confirmed with a total wall biopsy. Among the differential diagnoses is Hirschsprung's Disease (HD), although its association has not been demonstrated in the literature.

Material and Methods: We present a case of a 4-day-old male patient who presented septic shock secondary to perforation of the transverse colon. The initial biopsies reported MV of the colon and absence of ganglion cells in the rectum. A year later, laparoscopic intestinal mapping was performed, which demonstrated the absence of ganglion cells in the rectum and hypoplasia of the muscularis propria from the sigmoid to the distal ileum, confirming concomitant HD and VM.

Discussion: No cases of association between both diseases were found. Our case could correspond to an incidental finding or be the expression of the spectrum in a common etiology for both alterations. Further studies are necessary to determine if there is an association between VM and HD.

Categories
Gastro Intestinal