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Aganglionic megacolon in the adult. Urgent and surprising cause of intestinal occlusion

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Key words: Hirschsprung's disease. Intestinal obstruction. Anastomosis stenosis.

Dear Editor,

We have read the interesting article published recently in your journal titled "Hirschsprung disease with debut in adult age as acute intestinal obstruction: a case report" (1). In this regard, we would like to report an exceptional case of a combined evolution of Hirschsprung's disease and an infrequent surgical complication.

Case report

A 56-year-old male with a previous Duhamel's intervention and chronic constipation presented to the Emergency Room due to abdominal pain and cessation of stools of a ten-day duration. The examination revealed abdominal distension, pain and tympany. The computed tomography (CT) scan showed a colon dilation of 16 cm in diameter (Fig. 1) that produced a displacement of the viscera such as the liver. Stenosis of the colon adjacent to the previous colorectal anastomosis was also observed. An urgent total colectomy with ileostomy was performed and the pathological anatomy confirmed agangliosis in the distal segment of the colon (Fig. 1), as well as hypertrophy of the pre-aganglionic muscle itself. Thus, confirming a diagnosis of Hirschsprung's disease and stenosis of the previous colorectal anastomosis.



Discussion

Hirschsprung's disease or aganglionic megacolon is a congenital disease that usually occurs during the neonatal period and also in the adult. It is characterized by the absence of ganglion cells in the submucosal and myenteric plexuses, producing a functional obstruction and proximal dilation of the affected intestinal segment (1). Previously, the initial surgical treatment was subtotal colectomy, which was abandoned due to its high mortality and gave way to different techniques (2,3). In 1956, Duhamel described the widely accepted technique with the objective to preserve the pelvic innervation by means of a posterior colorectal anastomosis forming a sensitive but aganglionar neorectum in front, with the ganglionic propellant colon behind (4). However, this procedure also has complications (5) such as constipation, fecalomas, fistulas, incontinence, pelvic infection, rectal hemorrhage and even stenosis of the surgical anastomosis. In some cases, this can cause complicated intestinal occlusion.

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Figura 1: A) Corte coronal de TC abdominal donde se objetiva una severa dilatación del colon de hasta 16 cm de diámetro. B) Corte axial de TC abdominal donde se señala el colon transverso desplazando el hígado C) Imagen histológica al microscopia óptico con tinción Hematoxilina-eosina(HE) 200X donde se observa el plexo mientérico que muestra fascículos nerviosos hipertróficos (señalado) con ausencia de células ganglionares (aganglionosis).

Fig. 1. A. Coronal section of an abdominal CT where a severe dilation of the colon of up to 16 cm in diameter is observed. B. Axial section of an abdominal CT showing the transverse colon displacing the liver. C. Histological image under an optical microscope with hematoxylin-eosin (HE) 200x staining showing the myenteric plexus with hypertrophic nerve fascicles (arrow); an absence of cells is observed in the ganglion (aganglionosis).