EA060

TOTAL COLONIC AGANGLIONOSIS, A RARE CASE OF HIRSCHPRUNG'S DISEASE

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Introduction: The condition known as total colonic aganglionosis (TCA) is a rare variety on Hirschprung's (HSCR) disease. It affects the total colon which is aganglionic but may extend proximally into different lengths of the small bowel and is present in about 2-13% of HSCR cases.

Purpose: To present an uncommon case of megacolon leading to total colectomy in a 13-month-old patient.

Case report: Male patient 9 months old referred to our department diagnosed with HSCR for further treatment. From the patient's history, inability to feed, bilious vomiting and inability of meconium expulsion reported on the 1st day of life. On the 21st day of life exploratory laparotomy was performed during which, a stenosis was detected 6-7cm near the ileocecal valve. Full thickness biopsies from the ileum, transverse and descending colon were sent for histopathology exam. Appendectomy was performed and the appendix was also sent for exam. Loop ileostomy was performed. The histopathology report pointed out total lack of ganglion cells from each segment sent. At the age of 15 months the main surgical restoration was performed by total colectomy with ileorectal anastomosis using the Soave technique. Prophylactic loop ileostomy reconstruction took place after rapid biopsies sent during surgery

Conclusion: Patients with TCA are a challenging field of diagnosis and treatment. Diagnosis in the majority of the cases is based on the biopsies from the appendix and treatment is based on performing ileostomy. The final procedure aims for an accepted quantity and quality of daily stools with high mortality rates in these patients.