Images in Surgery Challenge

A 23-year-old male undergraduate presented with a history of recurrent constipation since childhood. He observed that the duration of constipation had progressively increased over the past decade with no known relieving factors. He has always had abdominal distension, which had also increased significantly over the past couple of years. He has had no episodes of bleeding per rectum. Examination revealed a young man with gross abdominal distension with tympanic percussion note and no palpable masses or organomegaly. The rectum was empty on digital examination. Plain abdominal radiographs revealed a markedly distended large bowel.

An abdominal computed tomography (CT) scan was performed [Figure 1].

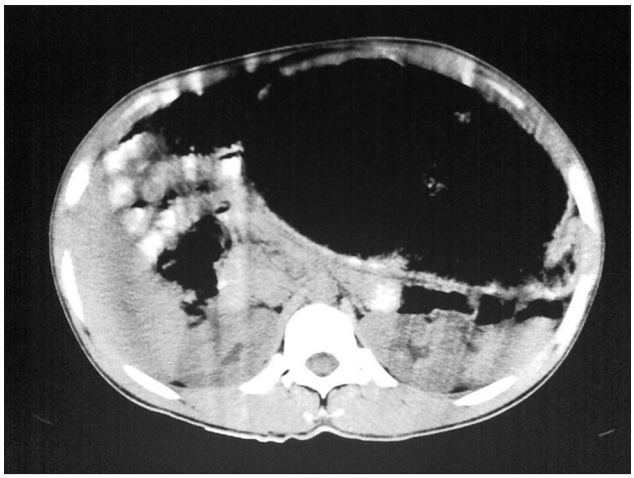

Figure 1: Abdominal CT scan of the patient

Question 1: Which features can be seen on the CT scan?

Question 2: List 3 possible differential diagnoses?

He had laparotomy, which revealed a hugely dilated sigmoid and descending colon with collapsed rectum. The transverse colon and the ascending colon were of normal calibre and exhibited normal peristalsis [Figure 2].

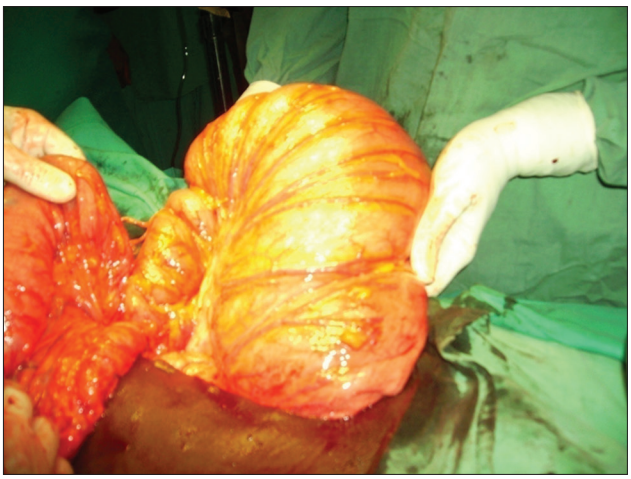

Figure 2: Intraoperative findings in the patient's colon

He had resection of the markedly dilated and redundant sigmoid colon and a divided colostomy as the first-stage operation. The remaining sigmoid colon and the rectum were resected at the second-stage operation 5 months later followed by colo-anal anastomosis. Histopathology of the resected rectum is shown in Figures 3 and 4.

Question 3: List two classical features in Figures 3 and 4.

Question 4: What is the final diagnosis?

He was followed up for three years in good condition with no constipation and good anal sphincteric tone.

DISCUSSION

Hirschsprung's disease is a congenital condition arising from absence of enteric ganglion cells of the myenteric plexus as well as the submucosal plexus. It may affect a variable length of the large intestine but typically presents with constipation from birth necessitating treatment in infancy or early childhood. Ignorance of the disease, particularly in ultra-short segment involvements, could lead to endurance of chronic constipation, leading to presentation in adulthood as in the index case. Treatment usually consists of resection of the affected segment of the rectum and colon, followed by restoration of intestinal continuity with a colo-anal anastomosis using varieties of techniques. A preliminary colostomy as in the index patient is often necessary in long-standing cases. Public health information on this condition is important in our setting.

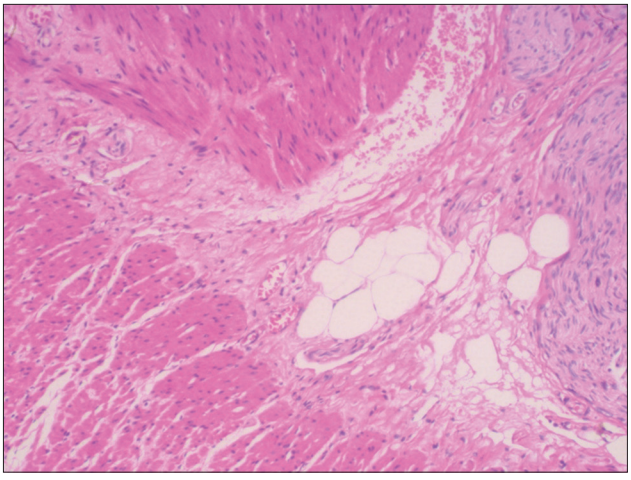

Figure 3: Photomicrograph of the rectal wall

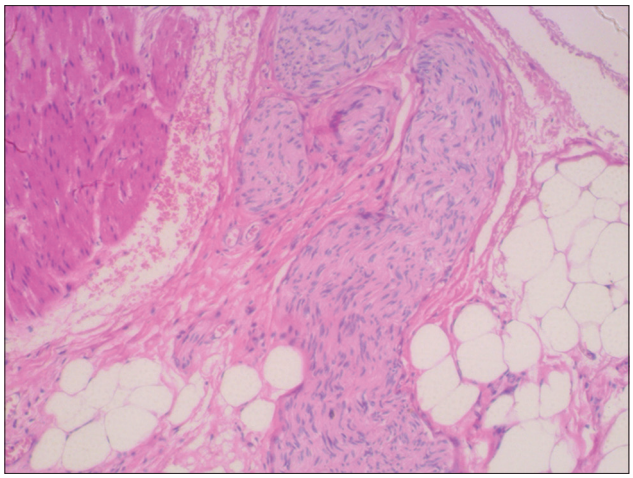

Figure 4: Photomicrograph of the rectal wall (×10)

ANSWERS TO QUESTIONS

Question 1:

a. Markedly dilated sigmoid colon
b. Displacement of the intra-abdominal organs to the right.

Question 2:

a. Adult Hirschsprung's disease
b. Colonic pseudo-obstruction
c. Sigmoid volvulus
d. Chagas disease
e. Colonic carcinoma.

Question 3:

1. Figure 3: Absent ganglion cells, muscle hypertrophy and adipocytes
2. Figure 4: Hypertrophied nerve bundle in the muscularis propria.

Question 4:

Hirschsprung's disease presenting in an adult.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 13-Aug-2020 **Revised:** 21-Aug-2020
Accepted: 18-Mar-2021 **Published:** 22-Sep-2022

Access this article online

Quick Response Code:

Website: www.chs-journal.com

DOI: 10.4103/njhs.njhs_36_20

How to cite this article: Adisa AO. Images in surgery challenge. Niger J Health Sci 2020;20:69-70.