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A RARE CASE REPORT OF HIRSCHSPRUNG DISEASE PRESENTING AS ACUTE OBSTRUCTION IN ADULT

General Surgery

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ABSTRACT

Hirschsprungs disease is an aganglionataed disease of colon and rectum . Hirschsprungs disease is not commonly seen in adults as most patients are diagnosed early in life and are treated surgically. However, some patients with mild symptoms may go undiagnosed into adulthood.However, at some point, the dilated proximal colonic segment may decompensate secondary to the distal obstruction and patients may experience rapidly worsening constipation or even acute obstruction.

KEYWORDS

hirschsprung disease, adult onset, constipation, obstruction

INTRODUCTION

Hirschsprung's disease (HD) is a malformation of the embryological development of the large intestine (colon). It is characterized by an abnormality of the motor function of the colon. This is due to the absence of the ganglion cells of Meissner's plexus in the submucosa, and those of Auerbach's plexus in the muscularis [1]. Common symptoms include delayed passage of meconium, recurrent constipation and failure to thrive in early childhood1[2-3].Hirschsprungs disease is not commonly seen in adults as most patients are diagnosed early in life and are treated surgically. However, some patients with mild symptoms may go undiagnosed into adulthood. Adult HD by definition occurs when the diagnosis is made after the age of ten. Patients usually suffer for many years before the diagnosis is established. They have a long-standing history of constipation and due to progressive dilation of the upstream colon present with very characteristic radiologic findings[4].However, at some point, the dilated proximal colonic segment may decompensate secondary to the distal obstruction and patients may experience rapidly worsening constipation or even acute obstruction[5].

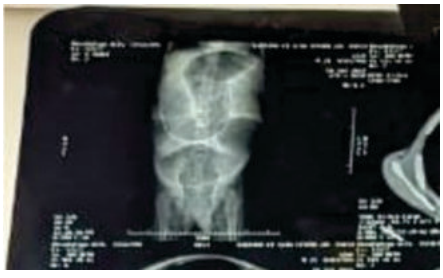
Case Study

A 17 year old male came to the emergency department with a 5 day history of pain abdomen,vomiting and abdominal distension and non passage of flatus.

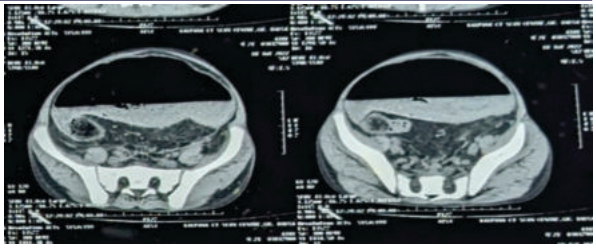
He had past history of constipation during the last 10 years with multiple consultations and was on laxatives and enema to pass motion requiring increasing doses since 4 months, He had no history of melena or bleeding per rectum.

On clinical examination patient was in shock and adequate fluid resuscitation was done. He was poorly built and malnourished with bmi-15.5 Abdomen was grossly distended with visible peristalsis Pr –rectum was empty. Investigations revealed sev anaemia with hb -7 g /dl, albumin levels -2.1 g/dl ,hyponatremia -128 mmol/dl,and hypokalemia 3.0 mmol/dl.

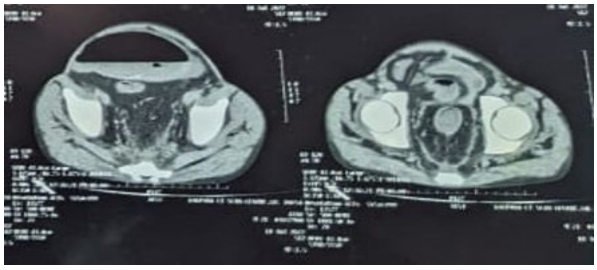
Patient underwent cect abdomen which was suggestive of large bowel obstruction with abrupt cutoff at the rectosigmoid junction.



Massively Dilated Large Bowel On Imaging

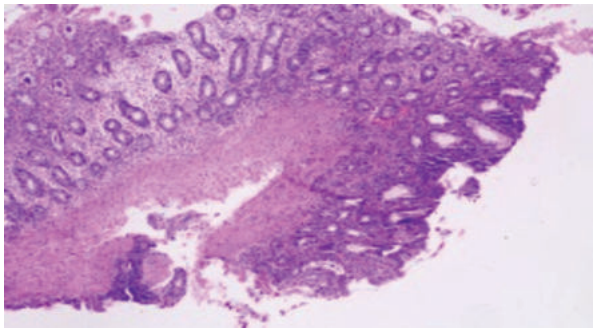


Transition Point Of Rectosigmoid Junction



Dilated Colon And Collapsed Rectum.

In view of non resolving obstruction patient underwent emergency laparotomy. Intra op Large bowel was grossly dialated and was collapsed distal to the recto sigmoid junction, a temporary diversion colostomy was made biopsy was taken from the rectum,which revealed hirschsprungs disease, a definitive colonic pull through surgery is planned at a later date.

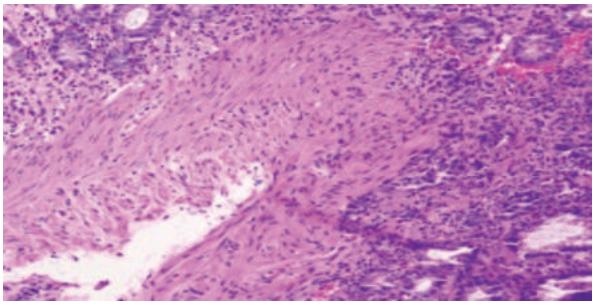


Histopathology slide showing hypertrophic nerve bundles and absence of ganglion cells.

CONCLUSIONS

Adult hirschprungs disease is a rare entity and is misdiagnosed or missed, high index of suspicion in a long standing chronic constipation

requiring laxatives and enemas on a regular basis with no identifyable cause should be evaluated. When the presentation is of acute intestinal obstruction an emergency diversion followed by diagnosis confirmation and definitive pull through surgery at a later date would be justified.



Absent ganglion cells with hypertrophic nerve bundles

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