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Adult Hirschsprung's Disease: A Case Report of 27-Year-Old Female Patient in Hawassa University Comprehensive Specialized Hospital, Hawassa, Sidama, Ethiopia

Wondmagegn Gizaw[1]; Murtii Teressa Obolu[1]*; Kibrom Legesse[1]; Eyerusalem Fissehatsion[1]; Dereje Berhenu[1]; Gebril Ahmed[1]; Mishame Damtew[2]; Fekade Yerakly[2]

[1]Hawassa University College of Medicine & Health Science Department of Surgery Hawassa University Comprehensive Specialized Hospital, Sidama, Hawassa, Ethiopia.

[2]Hawassa University College of Medicine & Health Science Department of Pathology, Hawassa University Comprehensive Specialized Hospital, Sidama, Hawassa, Ethiopia.

***Corresponding Author(s): Murtii Teressa Obolu**
Hawassa University College of Medicine & Health Science Department of Surgery Hawassa University Comprehensive Specialized Hospital, Sidama, Hawassa, Ethiopia.
Email: Murtyko1984@gmail.com

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Keywords: Adult hirschsprung's disease; Refractory constipation; Swenson's pull through technique.

Abbreviations: HSD: Hirschsprung's Disease; SAM: Severe Acute Malnutrition.

Abstract

Background: Adult Hirschsprung's disease is a functional intestinal obstruction whenever it's diagnosed in adolescent older than ten years and adult age. The primary pathology of Hirschsprung's disease is a congenital gut motor neuron development disorder. The disease usually diagnosed in over 94% of cases before the age of five years. However, beyond children age the disease is often overlooked and misdiagnosed as refractory functional constipation, which delay in diagnosis and management.

Case presentation: In this study, we report the case of a 27-year-old female patient presented with prolonged difficulty of defecation, progressive abdominal distension, intermittent vomiting and appetite loss. She had received frequent laxatives and rectal enema for temporary relief of her symptoms. Subsequently, from clinical and imaging evidence, with an impression Adult Hirschsprung's disease, Rectal biopsy was taken and the biopsy result confirmed Hirschsprung's disease.

Result: Initially, diversion of the sigmoid loop colostomy was done. Which resulted in a significant reduction in colonic calibre and patient was improved from secondary malnutrition morbidity. Despite, some difficulties from anatomic change and deep female pelvis, Swenson pull through technique was successful applied for definitive management. Subsequently, post operative outcome was satisfactory.

Conclusion: Thus, adults with a refractory functional constipation, Adult Hirschsprung's disease should be considered as a differential diagnosis. Those patients need a gold standard diagnostic histopathologic study. Even though, the management sequence and choice of surgical techniques depends on different factors, Swenson's pull-through technique is the most convenient to apply with or without modification.

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Introduction

Background: Adult Hirschsprung's disease" is a functional intestinal obstruction, whenever it's diagnosed in adolescent older than ten years and adult age. Which is very rare.

The primary pathology of Hirschsprung's disease is congenital gut motor neuron development disorder. Either from absence/incomplete of caudal migration of neural crest cell or survival and maturation problems of ganglion cell after normal migration. Which result in abnormal peristaltic movement of affected segment.

In children the disease is usually presented with delayed meconium passage at birth, early neonatal obstruction, progressive abdominal distension, infrequent passage of stool and difficulty, body wasting, failure to thrive. However, in adults the disease has no peculiar clinical features and the rarity of clinical condition mislead physicians to consider refractory functional constipation. In this case we are describing Adult Hirschsprung's disease diagnosed at 27-year-old-female.

Diagnosis of Hirschsprung's disease generally needs rectal biopsy confirmation. However, imaging such as rectal barium enema, abdominal CT-scan are helpful in providing a hint to consider a diagnostic gold standard histopathologic study.

Overall, gaol of surgical management is resection of aganglionated segment and anastomosis of ganglionated bowel segment down to the anus. The choice of surgical techniques depends on the actual case scenario, surgeon preference and different related complications.

Objective

To improve physicians' awareness in diagnosis of Adult Hirschsprung's disease.

Case presentation

History and physical examination

This is a 27-year- old female patient referred to our hospital who had compliant of prolonged difficulty in defecation since she known herself as old child. with current exacerbation of progressive abdominal distension, significant weight loss but unquantified, frequent abdominal pain, appetite loss. For this suffering she was on periodic laxative use and intermittent rectal enema for temporary relieving of her symptoms. On objective evaluation: the patient was chronically sick looking and wasted, hard palpable faecal mass abdominally, loose stool component on digital rectal examination.

Investigations

On labs exam moderate aneamia of chronic illness on complete blood count profile, electrolyte results; K^+-(1.7mEq/L), Na^+-(120mEq/L), Liver function; Albumin-(1.21g/dl). Imaging; Colon was full faeces and gaseous, with hugely distended entire colon on contrast enema (Figure 2) and computed tomography (Figure 1) both showed classic evidence of HSD that proximally distended and distally collapsed with clear demarcation of transition zone.

Management Outcomes and follow ups

With all the above evidences adult-onset severe acute malnutrition and Adult Hirschsprung's disease were diagnosed. For this initially patient was managed for acute secondary complications of severe acute malnutrition. Simultaneously, Rectal biopsy was taken and biopsy result confirmed Adult Hirschsprung's disease. Subsequently, on initial intraoperative finding colon was significantly dilated having typical transition zone at recto sigmoid region (Figure 3), with multiple faecalomas in the colon. Faecaloma removal and diversion loop colostomy was done. Post operatively patient was discharged home after advised colostomy care. later, biopsy result confirmed aganglionated Rectum.

In long run, Patient was significantly improved from her malnutrition associated and abdominal symptoms, she was admitted to ward for definitive management. Despite, intraopera-

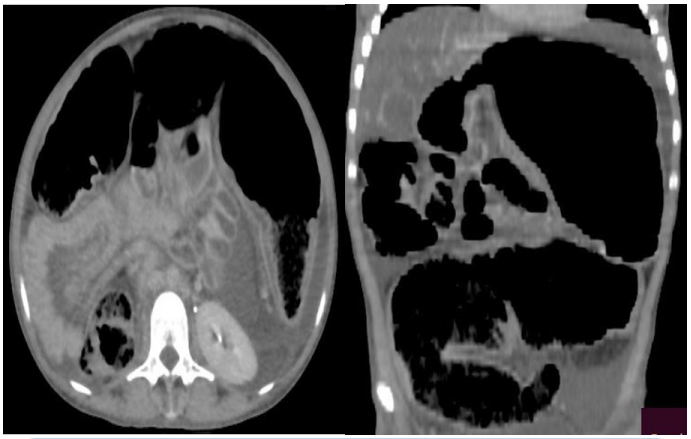

Figure 1: Axial and coronal section of abdominopelvic CT-scan shows significant colonic dilatation above transition zone and collapsed rectum below and the entire colon filled with gas.

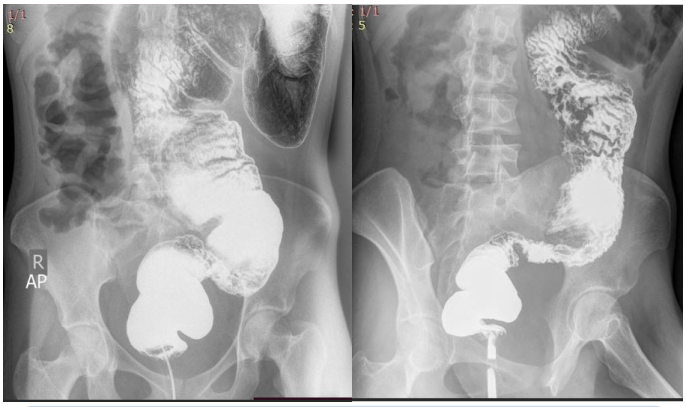

Figure 2: Contrast barium enema showing rectal collapse and significant Colon dilatation and the transition point at rectosigmoid and faecal loaded entire colon.

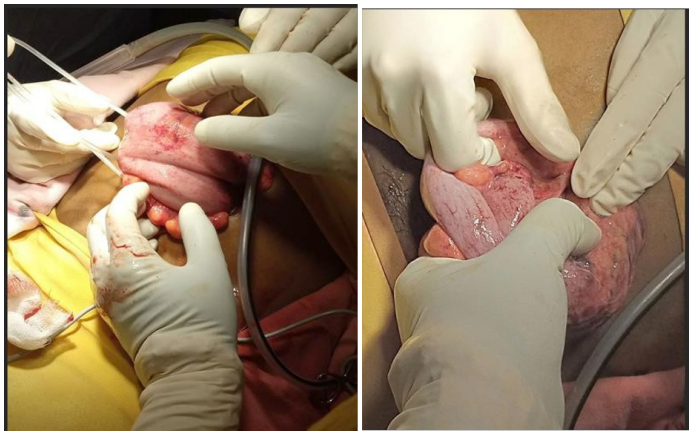

Figure 3: Intraoperative photograph shows hugly dilated sigmoid colon delivered to surgical incision for diversion.

tive challenge from anatomic change and deep female pelvis, Swenson pull through technique was successfully applied**.** The aganglionated segment was resected and gangilonated segment anastomosed to down to Anus. Resected aganglionated segment sent for histopathologic exam (Figure 4). Later biopsy revealed same result as previous (Figure 5,6). Post operative condition was smooth except colostomy site infection, that was treated with wound care. On Subsequent follow ups patient is doing well having regular bowel habit.

Discussions

Hirschsprung's illness in adults was first described in detail by Rosin in 1950. Since then, numerous instances and systematic review literature have been reported from all over the world, but the epidemiologic profile of adult Hirschsprung's illness is still unclear. According to the majority of literatures, the disease is prevalent in men and is diagnosed between the ages of 10 and 73, with a median age of 24. However, diagnoses are most frequently made in those under 30 years old [1-3].

Hirschsprung's disease is a congenital abnormality of the development of gastrointestinal motor neurons. Either because the neural crest cell's caudal migration was absent or partial, or because ganglion cells had problems with maturation and survival following normal migration. Which result in abnormal peristaltic movement of affected segment.

Hirschsprung's illness in adulthood rarely exhibits any peculiar symptoms; instead, it nearly always manifests as a constellation of refractory functional constipation [3-6].

Diagnosis of HSD in adults need high index clinical suspicion for patients with chronic constipation. A number of diagnostic techniques has been introduced for diagnosis including barium enema, rectal manometry and abdominopelvic CT-scan. Overall imaging, barium enema is found to be diagnostic in 83% of cases. However, absence of ganglion cell on Auerbach's and meissner's layer, and hypertrophied nerve trunk on histopathologic study remains the gold standard [7-11].

In addition to the diagnosis, there have been reports on several surgical therapy options for adult HSD; nevertheless, no particular surgical procedure or improved surgical technique has been recommended as a general solution. The Duhamel, Swenson, and Soave endorectal pull-through procedures, as well as anorectal myectomy with low anterior resection, are the most suitable surgical management options [12-14].

Adults with HSD who have severe acute malnutrition, considerable proximal dilatation, and acute obstruction should have rectal biopsies and protective diversion colostomies. This enables the patient to be prepared for more definitive surgical care. The general objective of surgical management is to remove an aganglionated intestinal segment and anastomose the ganglionated segment down to the anus [3,8,10,12,14,15].

Conclusion

Thus, adults with a refractory functional constipation, Adult Hirschsprung's disease should be considered as a differential diagnosis. Those patients need a gold standard diagnostic histopathologic study. Even though, the management sequence and choice of surgical techniques depends on different factors, Swenson's pull-through technique is the most convenient to apply with or without modification.

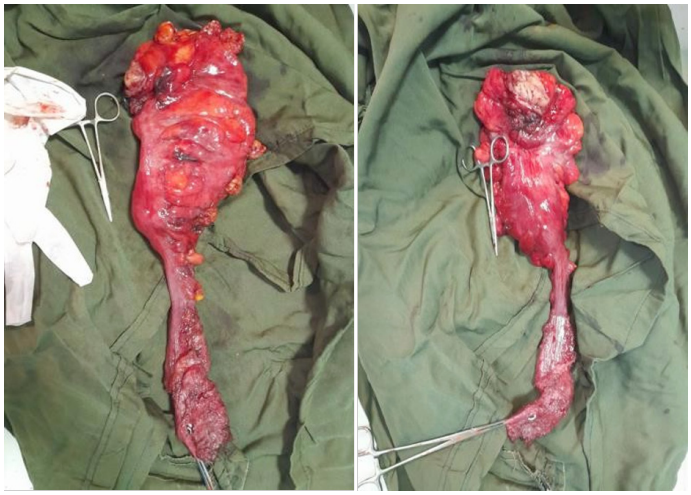

Figure 4: Intraoperative photograph of resected bowel segment shows collapsed rectum and dilated sigmoid colon.

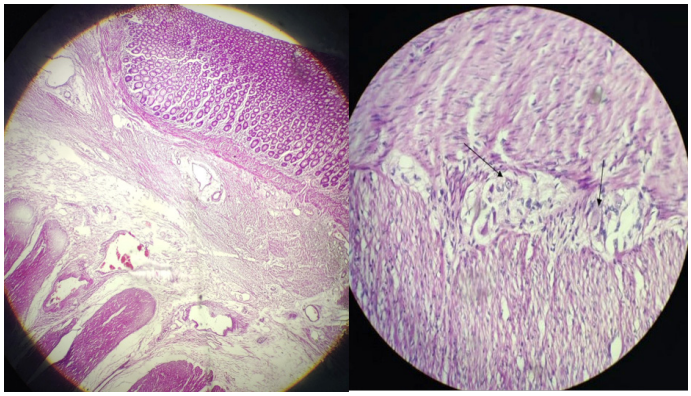

Figure 5: Low and high-power section shows sigmoid colon muscularis layer having ganglion cells as shown with black arrow.

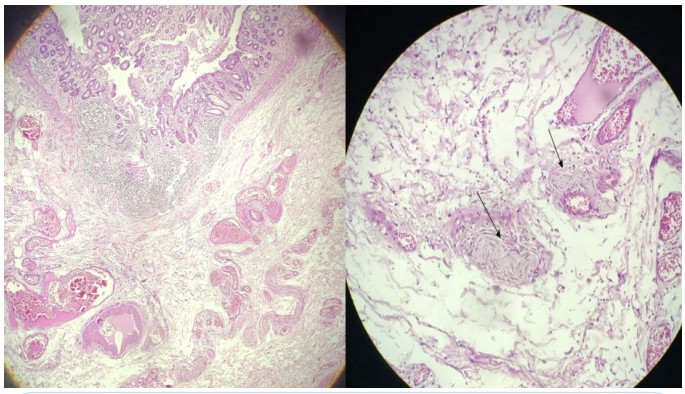

Figure 6: Low and high power-section show rectal tissue composed of bland mucosal glands and submucosa having hypertrophied nerve bundles.

Authors' contributions

Wondmegagn Gizaw

Conceptualization, supervision, validation and final manuscript approval

Murtii Teressa Obolu

Writing original draft, review editing , data curation

Other authors

Data curation and investigation

Informed consent: We declare that the patient has provided formal written informed consent for the publication of her medical case for educational purposes; the patient's name has been anonymised for privacy.

Institutional review board: Not applicable as this is case report.

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Disclosure of Conflict of interest: There is no conflict of interest in computation.

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