INTERNAL MEDICINE & PATIENT CARE

http://elynspublishing.com

Research Article | Journal of Internal Medicine and Patient Care | Open Access

Neonatal Short-segment Hirschsprung Disease: A Dilemma in the Emergency Room

Avani Hegde[1], Amanpreet Kaur[1], Abhinandan Sood[1], Manoj Dhanorkar[1], Kushaljit S. Sodhi[2], Muneer A. Malik[3], Arushi Gahlot Saini[1]*, and Praveen Kumar[1]

[1]Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India
[2]Department of Radiodiagnosis, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India
[3]Department of Pediatric Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

Received Date: : July 18, 2018, **Accepted Date:** August 20, 2018, **Published Date:** August 28, 2018.

Corresponding author: Arushi Gahlot Saini, Advanced Pediatric Centre, Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India, Tel: 91-172-275-5359; Fax: 91-172-274-4401; E-mail: doc.arushi@gmail.com; doctorpratibhasinghi@gmail.com

Abstract

Abdominal distention is a common disorder in newborns. Intestinal obstruction leading to abdominal distention often presents as an emergency. Hirschsprung disease should be suspected in neonates presenting with signs and symptoms of large bowel obstruction. Abdominal radiograph and contrast enema provide important clues to support the clinical diagnosis in the emergency room. Early diagnosis is important to prevent complications. This case sensitises the pediatricians regarding diagnosis of such cases.

Keywords: Abdominal distention; Intestinal obstruction; Hirschsprung disease

Introduction

Hirschsprung disease (congenital aganglionic megacolon) is a neurocristopathy affecting the enteric nervous system. It is characterized by absence of ganglion cells in the submucosal and myenteric plexus, most commonly in the rectosigmoid region (80% cases) followed by long-segment (10-15%) and complete bowel aganglionosis (5%) [1]. Most cases of Hirschsprung disease are diagnosed in the newborn period. It should be suspected in neonates presenting with features of distal bowel obstruction. Early recognition of Hirschsprung disease is essential in reducing morbidity and mortality.

Case

A two-day-old male neonate presented with progressive abdomen distention. He was born by normal vaginal delivery at 40 weeks gestation, weighed 3.25 kg at birth and was initiated on breastfeeds. He first passed meconium at 30 hours of life followed by 1 episode regurgitation of milk and generalised abdominal distention. On examination, the baby had good state-to-state variability, alertness, diffuse gaseous abdomen distention, bilious aspirates, normal bowel sounds and anal tone. He passed meconium every time following rectal enema. There was no fever, lethargy, features of peritonitis or gut-associated sepsis. Clinical diagnosis of septic ileus, meconium plug syndrome, Hirschsprung disease was considered. Repeated plain abdominal radiograph showed persistently dilated large bowel loop and absence of rectal gas shadow (Figure 1A). Blood and cerebrospinal fluid examination for sepsis were not contributory. Thyroid function test was normal. Contrast enema study showed dilated colon with a narrow transition zone (Figure 1B) suggesting Hirschsprung disease. He underwent colostomy and is awaiting definitive repair.

Discussion

In this case, diagnosis of Hirschsprung disease was made based on clinical presentation and radiological features. Hirschsprung disease is a common cause of neonatal large gut obstruction. Neonatal presentation includes progressive abdomen distention, delayed passage of meconium (>24 hours after birth), bilious vomiting or aspirates with feed intolerance. It is commonly confused with septic ileus, necrotizing enterocolitis or meconium plug syndrome. Absence of clinical and laboratory features of sepsis, absence of rectal gas shadow and presence of fixed dilated bowel loops on abdominal radiograph should raise suspicion of Hirschsprung disease. Contrast enema helps in early confirmation and initiating definitive management [2]. Surgical intervention is the definitive treatment modality. It includes excision of the aganglionic and transition zones followed by surgical apposition of normally innervated bowel [3].

In conclusion, Hirschsprung disease should be suspected in neonates presenting with signs and symptoms of large bowel obstruction and absence of sepsis. Abdominal radiograph and contrast enema provide important clues to support the clinical diagnosis in the emergency room.

Conflict of Interest

There is no conflict of interest between authors.

Funding

There was no funding agency/source involved in this study. No honorarium, grant or any other form of payment was given to anyone to produce the manuscript.

Author's Contributions

AH: Substantial contributions to analysis of data, drafting, final approval of the version to be published and agreement to be accountable for all aspects of the work. AK: Substantial contributions to acquisition of data, drafting, final approval of the version to be published and agreement to be accountable for all aspects of the work. AS: Substantial contributions to acquisition of data, drafting, final approval of the version to be published and agreement to be accountable for all aspects of the work. MD: Substantial contributions to analysis and interpretation of data, drafting, final approval of the version to be published and agreement to be accountable for all aspects of the work. KSS: Analysis and interpretation of radiological data, final approval of the version to be published and agreement to be accountable for all aspects of the work. MAM: Chief operating surgeon, final approval

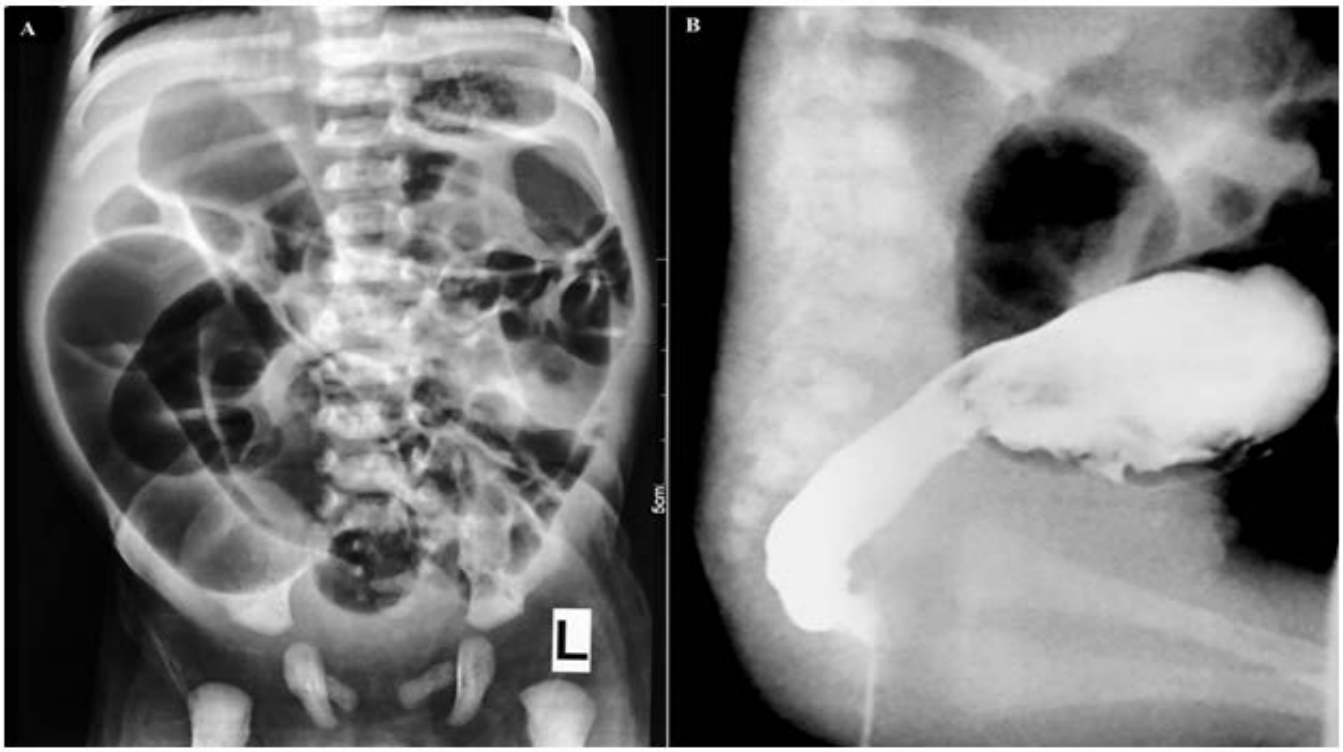

Figure 1: Radiological features of Hirschsprung disease. (A) Plain abdominal radiograph showing dilated large bowel loop and absence of rectal gas (B) Contrast enema study done by per rectal administration of dilute urograffin solution shows transition zone at the recto-sigmoid region (arrow), with dilated sigmoid colon.

of the version to be published and agreement to be accountable for all aspects of the work. AGS: Substantial contributions to the conception of work and interpretation of data, revising it critically for important intellectual content drafting, final approval of the version to be published and agreement to be accountable for all aspects of the work. PK: Substantial contributions to the conception of work and interpretation of data, revising it critically for important intellectual content drafting, final approval of the version to be published and agreement to be accountable for all aspects of the work.

References

1.Parisi MA. Hirschsprung Disease Overview. 2002 Jul 12 [Updated 2011 Nov 10]. In: Pagon RA, Adam MP, Ardinger HH, et al. (eds). Gene Reviews® [Internet]. Seattle (WA): University of Washington, Seattle; 19932015. Available from: http://www.ncbi.nlm.nih.gov/books/NBK1439/. [Accessed on: March 24, 2015].

2.Putnam LR, John SD, Greenfield SA, Kellagher CM, Austin MT, Lally KP, et al. The utility of the contrast enema in neonates with suspected Hirschsprung disease. J Pediatr Surg. 2015;50(6):963-6. doi: 10.1016/j.jpedsurg.2015.03.019.

3.Das K, Mohanty S. Hirschsprung Disease - Current Diagnosis and Management. Indian J Pediatr. 2017;84(8):618-623. doi: 10.1007/s12098-017-2371-8.

Corresponding author: Arushi Gahlot Saini, Advanced Pediatric Centre, Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India, Tel: 91-172-275-5359; Fax: 91-172-274-4401; E-mail: doc.arushi@gmail.com; doctorpratibhasinghi@gmail.com

Received Date: July 18, 2018, **Accepted Date:** August 20, 2018, **Published Date:** August 28, 2018.

Citation: Hegde A, Kaur A, Sood A, Dhanorkar M, Sodhi KS, Malik MA, Saini AG, Kumar P (2018) Neonatal Short-segment Hirschsprung Disease: A Dilemma in the Emergency Room. J Int Med Pat Care 1(1):101.