Case Report

Access this article online
Website: www.afrjpaedsurg.org
DOI: 10.4103/0189-6725.181708
Quick Response Code:

Congenital diaphragmatic hernia, Meckel's diverticulum and malrotation in a 3-month-old infant

Laxman Basani, Roja Aepala, B. Madhu Mohan Reddy[1]

ABSTRACT

Congenital diaphragmatic hernia (CDH) is a common developmental anomaly that usually presents in the neonatal period. It is known to be associated with cardiac, renal, genital and chromosomal anomalies. Late presentation of CDH (beyond 1-month of age) is seen in 13% of the cases. Malrotation is reported in 42% of CDH cases. We report a case of a 3-month-old infant with concurrent CDH, Meckel's diverticulum and malrotation. This is the first case report of such an association in an infant.

Key words: Congenital diaphragmatic hernia, infant, malrotation, Meckel's diverticulum

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a relatively common anomaly with an estimated incidence of 1 in 2000-3000 newborns.[1,2] The outcome of patients with CDH depends on pulmonary hypoplasia, pulmonary hypertension and associated malformations. Associated anomalies are reported in 46% of cases of CDH.[3] Meckel's diverticulum (MD), a developmental anomaly of omphalomesenteric or vitelline duct is the most common congenital anomaly of gastrointestinal (GI) tract with an incidence of 2% in the general population.[4] Malrotation in children is uncommon but is seen in 42% of cases of CDH.[5]

The association between CDH and MD, and also CDH with malrotation, have been reported earlier.[5,6] However, an association of CDH, MD and malrotation has not been reported previously.

Departments of Neonatology, [1]Pediatric Surgery, Dolphin Children's Hospital, Hyderabad, Telangana, India

Address for correspondence:
Dr. Laxman Basani,
Dolphin Childrens Hospital, 17-87, Road No 1,
Kamalanagar, Chaitanyapuri, Hyderabad - 500 060,
Telangana, India.
E-mail: laxmanbasani@yahoo.co.in

CASE REPORT

A 3-month-old male infant weighing 6.7 kg was referred to our hospital with respiratory distress. He had fever, vomiting and rapid breathing for 3 days prior to admission and was treated at a peripheral hospital.

He was born to a 28-year-old primigravida mother by caesarean section (Ind: Cephalopelvic disproportion) at 39 weeks of gestation and weighed 3460 g at birth. Antenatal ultrasound was normal. Apgar scores were 8 and 9 at 1 and 5 min of age. He was exclusively breastfed. Except for occasional vomiting he was well and weight gain was adequate.

On examination, the baby had respiratory distress with respiratory rate of 72/min and subcostal retractions. On auscultation, air entry was decreased on the left side and bowel sounds were heard. He was febrile with temperature of 100.9°F, heart rate 142/min, and SpO_2 89% in room air. Chest X-ray [Figure 1] showed bowel loops in left hemithorax with mediastinal shift to the right. A provisional diagnosis of CDH was made, and computed tomography (CT) scan of chest and abdomen was done.

CT scan showed bowel loops and spleen in left hemithorax with mediastinal shift to the right side suggestive of CDH on the left side.

Laparotomy done through left subcostal incision revealed classical posterolateral defect in the diaphragm with

Cite this article as: Basani L, Aepala R, Reddy BM. Congenital diaphragmatic hernia, Meckel's diverticulum and malrotation in a 3-month-old infant. Afr J Paediatr Surg 2016;13:47-9.

herniation of the small bowel, colon and spleen into left hemithorax [Figure 2]. Malrotation of small bowel and MD were noticed [Figures 3 and 4]. Examination showed MD located at 38 cm from the ileocecal junction, measuring 6 cm in length and 1.5 cm in width along the antimesenteric border of the ileum. Palpation of MD did not show any nodule or mass within its lumen.

The diaphragmatic defect was closed primarily after reducing the contents from left hemithorax. Ladd's procedure (excision of the Ladd bands, widening of the mesentery, appendectomy, derotation and intestinal repositioning) was done, and MD was excised. Baby was fed from the 3rd day and was discharged uneventfully on the 8th post-operative day.

DISCUSSION

The development of diaphragm starts at 4th week of gestation and involves fusion of septum transversum and pleuroperitoneal membranes.[1] During 4th-5th week of gestation, midgut herniates through the umbilical cord and returns back to the abdominal cavity at 9-10 weeks of gestation. Failure of the closure of pleuroperitoneal canal at 8 weeks of gestation results in herniation of abdominal contents into thorax. CDH is usually associated with pulmonary hypoplasia on the affected side causing severe respiratory distress soon after birth. Minor defects may be asymptomatic until abdominal contents herniate into the thoracic cavity. Several intra-abdominal organs like stomach, small bowel, spleen, omentum, colon and kidney can migrate through the diaphragmatic defect.

The signs and symptoms of late presenting CDH are nonspecific and are frequently related to the digestive tract (abdominal pain, vomiting and dysphagia) than the respiratory system (dyspnoea) in contrast to the classic neonatal presentation.[2,5,7] This late presentation is more frequent in men (3:1) on the left side (70-90%) and is rarely bilateral. The basic investigation is the chest radiograph from which a diagnosis can be made often. Other diagnostic

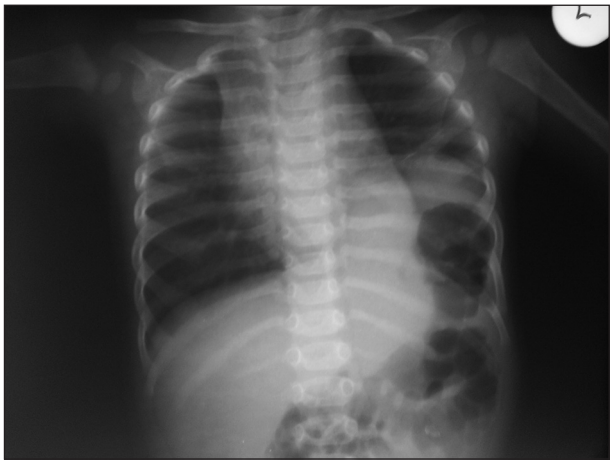

Figure 1: Chest X-ray shows bowel loops in left hemithorax and mediastinal shift to right side

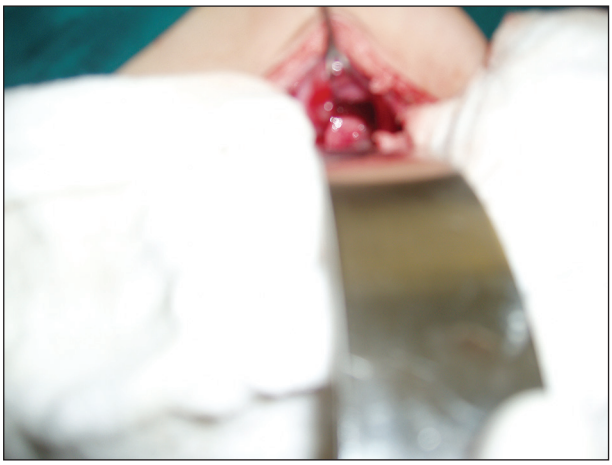

Figure 2: Laparotomy shows posterolateral defect in diaphragm

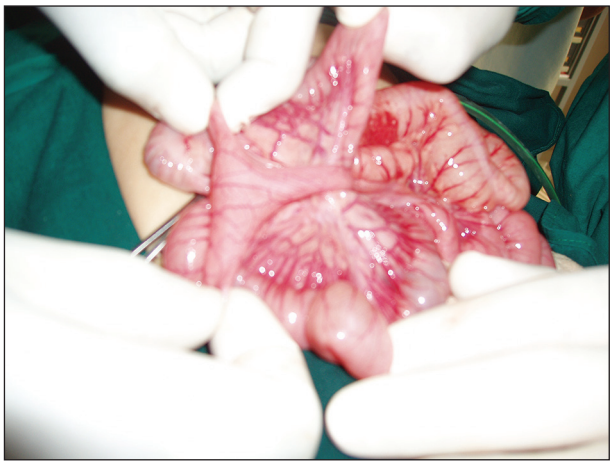

Figure 3: Malrotation of intestine

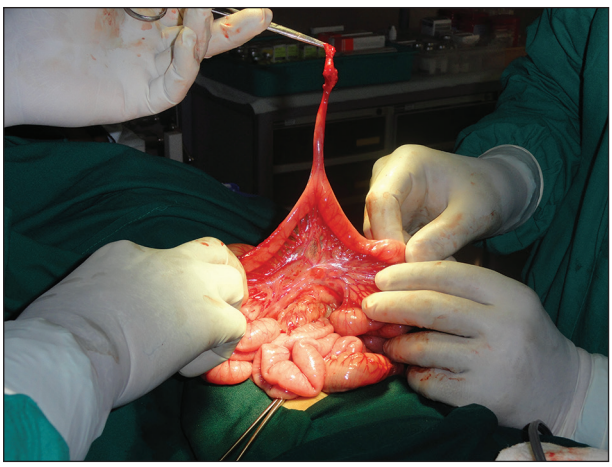

Figure 4: Meckel's diverticulum

modalities used for diagnosis are ultrasound chest, CT chest and GI contrast study.[8]

MD occurs due to incomplete obliteration of the vitelline duct at 7 weeks of gestation and is seen in 2% of the population.[4]

When midgut returns to the abdominal cavity during 9th-10th week of gestation, a 270° anticlockwise rotation around the upper mesenteric artery occurs, resulting in duodenal arch formation. Displacement of the abdominal viscera into the thoracic cavity distorts intestinal anatomy and fixation.[5]

The incidence of malrotation is estimated to be 1 in 500 births, but the true incidence is underestimated as many cases remain asymptomatic.[9] The incidence in adults seems to be increasing with the increasing use of diagnostic imaging.[10] A thickened band of mesentery (Ladd's bands) can join the cecum to the duodenum causing obstruction. Intestines suspended with only one point of fixation are prone to torsion and formation of volvulus causing acute intestinal obstruction.

In 70% of patients, malrotation presents with chronic symptoms like vague or intermittent abdominal pain, nausea, vomiting, diarrhoea and abdominal distension for 6 months or more before the diagnosis is made. Contrast studies show a vertical duodenum, lack of duodenojejunal flexure (80% of cases) and abnormal location of cecum or colon. CT scan allows the evaluation of superior mesenteric vessels, position of the duodenum and 'whirlpool appearance' of the small bowel due to volvulus of the midgut around the vascular pedicle.[10]

Definitive treatment is surgery with reduction of the herniated contents and closure of the diaphragmatic defect. Prognosis is good in late presenting CDH due to normal lung development. The surgical treatment for malrotation is the universally accepted Ladd procedure.

The management of incidentally found MD is controversial.[11] The reported morbidity rates after removal of incidentally found asymptomatic MD are much lower than resection of symptomatic MD.[12-14]

CONCLUSION

CDH, MD and malrotation might accompany each other and need appropriate attention. This case of CDH, MD and malrotation in an infant is perhaps the first one to be reported so far.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Jen TK, Lally KP. Congenital diaphragmatic hernia and eventration. In: Holcombe GW 3rd, Murphy JP, editors. Ashcraft's Pediatric Surgery. 5th ed. Philadelphia, Pa, USA: Elsevier Saunders; 2010. p. 304-23.
2. Wright JC, Budd JL, Field DJ, Draper ES. Epidemiology and outcome of congenital diaphragmatic hernia: A 9-year experience. Paediatr Perinat Epidemiol 2011;25:144-9.
3. Sweed Y, Puri P. Congenital diaphragmatic hernia: Influence of associated malformations on survival. Arch Dis Child 1993;69: 68-70.
4. Hosgor M, Karaca I, Karkiner A, Ucan B, Temir G, Erdag G, *et al*. Associated malformations in delayed presentation of congenital diaphragmatic hernia. J Pediatr Surg 2004;39:1073-6.
5. Schropp KP, Garey CL. Meckel's diverticulum. In: Holcombe GW 3rd, Murphy JP, editors. Ashcraft's Pediatric Surgery. 5th ed. Philadelphia, USA: Elsevier Saunders; 2010. p. 526-31.
6. Sehgal A, Chandra J, Singh V, Dutta AK, Bagga D. Congenital diaphragmatic hernia: Delayed presentation with asymptomatic spleenic herniation. Indian J Chest Dis Allied Sci 2002;44:57-60.
7. Kang IS, Ahn SM, Han A, Oh JT, Han SJ, Choi SH, *et al*. Giant Meckel's diverticulum associated with a congenital diaphragmatic hernia. Yonsei Med J 2004;45:177-9.
8. Abubakar AM, Bello MA, Chinda JY, Danladi K, Umar IM. Challenges in the management of early versus late presenting congenital diaphragmatic hernia in a poor resource setting. Afr J Paediatr Surg 2011;8:29-33.
9. Bösenberg AT, Brown RA. Management of congenital diaphragmatic hernia. Curr Opin Anaesthesiol 2008;21:323-31.
10. El-Chammas K, Malcolm W, Gaca AM, Fieselman K, Cotten CM. Intestinal malrotation in neonates with nonbilious emesis. J Perinatol 2006;26:375-7.
11. Salústio R, Nabais C, Paredes B, Sousa FV, Porto E, Fradique C. Association of intestinal malrotation and Bochdalek hernia in an adult: A case report. BMC Res Notes 2014;7:296.
12. Robijn J, Sebrechts E, Miserez M. Management of incidentally found Meckel's diverticulum a new approach: Resection based on a risk score. Acta Chir Belg 2006;106:467-70.
13. Soltero MJ, Bill AH. The natural history of Meckel's diverticulum and its relation to incidental removal. A study of 202 cases of diseased Meckel's diverticulum found in King County, Washington, over a fifteen year period. Am J Surg 1976;132:168-73.
14. Cullen JJ, Kelly KA, Moir CR, Hodge DO, Zinsmeister AR, Melton LJ 3rd. Surgical management of Meckel's diverticulum. An epidemiologic, population-based study. Ann Surg 1994;220:564-8.