

Short Clinical Report

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Submitted: 13-09-2020

Accepted: 27-10-2020

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DOI: <https://doi.org/10.47338/jns.v9.923>

Congenital paraesophageal hernia presenting as intrathoracic gastric volvulus in a neonate

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CASE PRESENTATION

A 5-day-old male term baby (with dysmorphic features) weighing 3.4 kilograms was evaluated for an antenatally diagnosed double aortic arch on fetal echocardiography. A postnatal echocardiogram also suggested the same. CT scan thorax was done which showed a large posterior diaphragmatic hernia on the left side with herniation of the abnormally rotated stomach into the left hemithorax. Mild dilatation of the esophagus in its entire extent was also noted. Great vessels in the thorax were torturous but had normal branching pattern. A double aortic arch was not appreciated. The patient was not tolerating breastfeed, hence further workup was planned. The upper gastrointestinal contrast study showed a part of the stomach in the thorax along with tortuous and dilated esophagus (Fig.1). At operation, the stomach was completely herniated into the thorax with associated organoaxial volvulus. The stomach was viable, therefore, a reduced back and detwisted. Two cm defect was noted at the esophageal hiatus on the left side of the spine, covered by a lax peritoneal sac. The paraesophageal defect was closed. Intraoperatively a small eventration of the left diaphragm was noted which was also plicated during the procedure. Antireflux procedure and gastropexy were not carried out. The baby withstood the procedure well.

The postoperative period in the NICU was uneventful and the baby was discharged on the eighth postoperative day. The patient required one more hospital admission at the age of 7 months for respiratory distress. Chest radiograph revealed multiple emphysematous spaces in the bilateral lung parenchyma with bilateral pneumothorax needing chest tube drainage. The previous CT scan of the thorax was reviewed again which showed a small air-filled cavitary lesion in the left lung. The patient was

managed conservatively. The baby is on our regular follow-up and doing fine.



Figure 1: Contrast study showing intrathoracic rotated stomach

DISCUSSION

Esophageal hiatal hernias are classified into 4 types.[1] Type 1 involves herniation of cardia only (sliding hernia); type 2 denotes herniation of the fundus of the stomach (rolling hernia); type 3 is the mixed variety where both the fundus and cardia herniate (our case); and type 4 is the herniation of other abdominal organs like colon, spleen.[2,3]

The exact cause of PEHH is not known. It is postulated that congenital form most likely occurs due to weakness of gastroesophageal attachments, deficiency of gastrocolic or gastrosplenic ligaments which combined with positive intraabdominal and

negative intrathoracic pressure facilitate the abdominal content (most commonly stomach) to migrate cranially.[4,5]

PEHH can be asymptomatic in 8% of patients but can present with gastroesophageal reflux or respiratory symptoms.[3] Vomiting and recurrent respiratory tract infection are seen in most of the cases. Other features are feed intolerance and failure to thrive; hematemesis should lead to the suspicion of gastric volvulus.[3,6] Our case had consequent feed intolerance.

Genetic factors may play a role in this condition such as Marfan's syndrome, collagen 3 disorder, right isomerism, etc.[4] Our case had features that would suggest collagen defects such as dysmorphic facies (dolichocephaly), torturous great vessels of thorax, development of cystic lung space, and pneumothorax. Further clinical and genetic evaluation is planned for the diagnosis.

Management of the PEHH is always surgical in the pediatric age group as most of the patients are symptomatic. Steps of surgery involved the reduction of hernial contents, complete excision of the sac, preservation of the vagus nerve, closure of the hiatal

defect with crural approximation, and gastropexy. Resection of the sac helps in the prevention of recurrence.[5] An abdominal approach is preferred. Antireflux surgery is recommended such as fundoplication. According to Karpelowsky et al., 60% of the postoperative patients who did not have additional antireflux procedure, developed symptomatic reflux.[7] Our patient didn't undergo antireflux procedure but doesn't have any symptoms related to the reflux in 13 months of follow up. The recurrence rate is 11 % but most of them may not require any further surgical management.[6] Laparoscopic repair of paraesophageal hernia has decreased the complications and recurrence rates, although it is challenging in young patients due to the small working space.[8]

Acknowledgements: Nil

Conflict of Interest: None declared

Source of Support: Nil

Consent to Publication: Author(s) declared taking informed written consent for the publication of clinical photographs/material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

Author Contributions: Author(s) declared to fulfil authorship criteria as devised by ICMJE and approved the final version.

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