



Case Report

Right sided congenital diaphragmatic defect with liver hernia: A case report

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Abstract

Congenital diaphragmatic hernia is a rare entity occurring in 1 in 2000-4000 live births and accounts for 8% of all major congenital anomalies. Congenital diaphragmatic hernia (CDH) is a major surgical emergency in new-borns because the key to survival depends on the prompt diagnosis and treatments. We reported here a case of right sided congenital diaphragmatic defect with liver hernia in a 7 month old baby, who came to seek medical care at a tertiary care centre in Navi Mumbai for on and off cold and cough with breathlessness after feeds. The baby was tachypneic on examination. Immediately chest X-ray and CT scan of thorax and abdomen was done to assess symptomatically and clinically suspected case of congenital diaphragmatic hernia. It was a case of right sided congenital diaphragmatic hernia.

Key words

Congenital diaphragmatic hernia, Right side, Rare, Liver herniation, 7 month female.

Introduction

Congenital diaphragmatic hernia is definitely a matter of concern due to its high incidence of morbidity and mortality. There is a male predominance with the ratio of 3:2 [1]. There are three types and these are posterolateral

Bockdalek hernia (usually occurring at approximately 6 weeks of gestation), the anterior Morgagni hernia and the hiatal hernia. Most common type of CDH is left-sided Bockdalek hernia (85%) through posterolateral defect in diaphragm (Foramen of Bockdalek). In left sided hernia the large and the small bowel

with or without intra abdominal solid organ may be herniated into the thorax. In right-sided hernia (incidence 13%) only the liver and portion of small bowel tend to be herniated into the thorax [2]. Chest x-ray and CT are considered as best diagnostic methods [3].

Case report

A 7 month old female, full term, normal vaginal delivery with no previous antenatal ultrasonography done presented to MGM Hospital, Kamothe, Navi Mumbai with chief complaints of on and off episodes of cough and cold, breathlessness especially after feeds since two months. Usually child presents with malnutrition after 1 month of birth. This is unlike the classic presentation of CDH as this patient was well developed and presented late i.e. 5 months of age. There were no gastrointestinal symptoms in this case. On examination, the child was found to be tachypneic with a respiratory rate of 60 breaths per min with mild intercostal retractions and decreased air entry on the right side. The most severely affected events develop respiratory distress at birth whereas large majority present respiratory symptoms within 24 hours. Of birth, only 2.6-10% of the cases may present after this period. It has been seen that the proportion of right sided CDH in late presenting cases is higher. In retrospective view of patients with right CDH, the mean age for diagnosis to be six months. Usually right sided CDH presents with gastrointestinal problems and left sided CDH presents with respiratory symptoms. Immediately chest x-ray showed an ill defined non-homogenous soft tissue opacity occupying lower half of the right hemi-thorax with shift of mediastinum and heart towards the left side. The right hemi-diaphragm silhouette is obscured with this opacity (**Photo - 1**) whereas the left hemi-diaphragm appears at its normal level without any diaphragmatic defect.

Photo - 1: Frontal chest radiograph: Right hemi-diaphragm is obscured with the well-defined opacity in right lower zone.



Tomogram showed soft tissue attenuation occupying lower half of right hemi thorax with non-visualization of right hemi diaphragm. There is evidence of mediastinal and cardiac shift towards the left side. (**Photo - 2**) NECT of thorax showed soft tissue attenuation occupying right lower hemi thorax part of the right lobe of liver. It showed at the level of origin of left lower lobe bronchus the liver can be seen, herniation of part of right lobe of liver in the right hemi thorax with non-visualization of right dome of diaphragm throughout its entire extent. There is evidence of shift of mediastinum and heart in left hemi thorax. (**Photo - 3, Photo - 4**) The contrast enhanced CT was not done on the view of increased radiation exposure to the child. Right diaphragmatic crura were found to be discontinuous on its progression. (**Photo - 5**) Kidney was found in normal position whereas liver was herniating into right hemi thorax.

(Photo - 6) Post-operatively X-ray showed repair of the hernia with absence of the opacity in the right lower zone, the right lung appears well expanded. **(Photo - 7)**

Photo - 2: Tomogram: Soft tissue attenuation occupying lower half of right hemi thorax with non-visualization of right hemi diaphragm.



Photo - 3: NECT of thorax: Soft tissue attenuation occupying right lower hemi thorax part of the right lobe of liver.



Photo - 4: NECT of thorax: Soft tissue attenuation occupying right lower hemi thorax part of the right lobe of liver.



Photo - 5: Arrow showing left crus of diaphragm (normal) and right and arrowhead showing right diaphragmatic crura which is discontinuous on its progression.

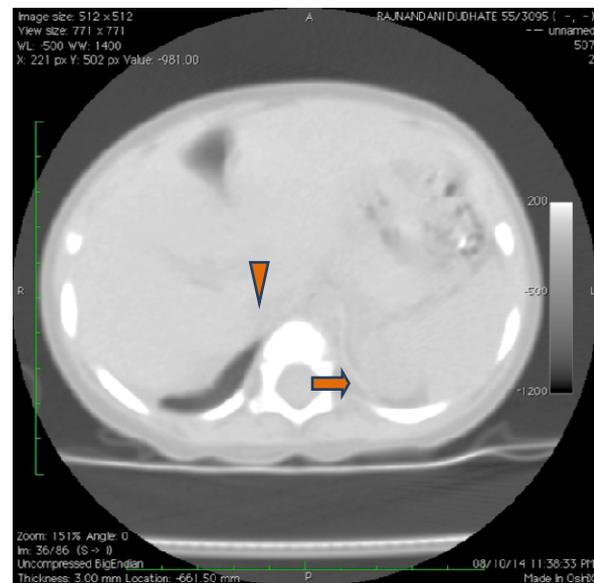


Photo - 6: Coronal image: Kidney in normal position and liver herniated into right hemithorax.

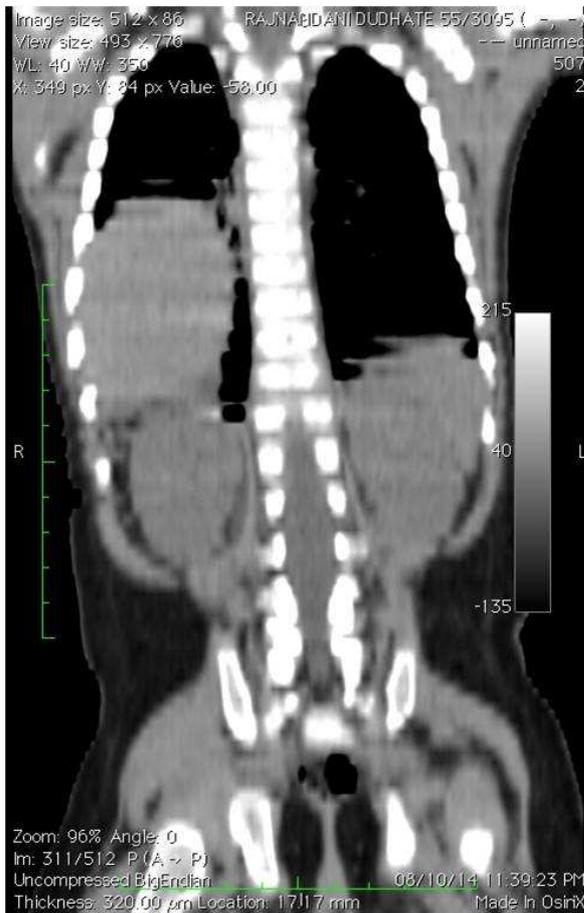


Photo - 7: Post-operative X-ray: Repair of the hernia with absence of the opacity in the right lower zone, the right lung appears well expanded.



Discussion

The diaphragm is the mesodermal partition in between thorax and abdomen develops at 3 to 8 week of intrauterine life from the following sources:

- 1) Septum transversum forms the central tendon of diaphragm. It is a sheet of mesoderm lying caudal to the pericardial sac and extends from the ventral body wall to the oesophageal segment of the foregut.
- 2) Dorsal mesentery of the oesophagus forms the crura.
- 3) Peripheral part is developed from the shelf-like projection of the body wall.
- 4) Pleuroperitoneal membranes: The openings are situated dorsal to the septum transversum and on each side of the dorsal mesentery of the oesophagus. Each opening is closed by a pleuroperitoneal membrane which is dissected off from the body wall by the caudal growth of the lung-bud. The membrane fuses with septum transversum and with the dorsal mesentery of the oesophagus [4, 5].

Here the defect is in the failure of the right sided pleuroperitoneal membrane to close the same sided pleuroperitoneal canal [6]. Pleuroperitoneal membranes are located dorsolateral to the pleuroperitoneal canals. The pleuroperitoneal canal is closed by the fusion of its edges.

The hernia most commonly occurs in the left side as it closes later but here in this reported case the hernia is in the right side which is very uncommon. The diaphragmatic defect allows the abdominal viscera to enter the thoracic cavity. The herniated mass prevents the growth of the right lung causing pulmonary hypoplasia [7].



Even though majority of such cases occur as isolated entity, it is important to look for other congenital malformations like cardiac anomalies, pulmonary agenesis or hypoplasia, renal agenesis, hydronephrosis, spina bifida and chromosomal anomalies such as Trisomy 21, 18 or 13 which can coexist along with CDH. Familial inheritance is reported to occur in 2% of the cases. The key to survival of these patients depends on the prompt diagnosis, prenatal management, delivery, stabilization and treatment. The site, size, contents of the hernia and associated anomalies also greatly influence the outcome of the disease.

CDH is also reported to occur in babies of the mothers who are exposed to quinine, thalidomide and antiepileptic drugs during pregnancy [8]. Liver herniation might be associated with poorer prognosis detected by radiological investigations in fetal congenital diaphragmatic hernia. Plain radiography sometimes may be helpful for presumptive diagnosis and contrast CT may help to arrive at a precise diagnosis.

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