

Congenital Morgagni hernia: misleading images and delayed diagnosis

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ABSTRACT

Diaphragmatic hernia of the Morgagni type is relatively rare. We report the case of a term 10-day-old female infant admitted through the emergency department with symptoms of lethargy, poor feeding, and vomiting. Her general physical examination was normal except for weight loss of 450gm and minimal sub-costal indrawing, whilst normal breath sounds and heart sounds on auscultation. Sepsis markers were negative. Chest radiography revealed non-specific opacities in the right middle and lower zones. A diagnosis of pneumonia was made and the patient was admitted for parenteral antibiotics. The patient remained symptomatic after seven days of intravenous antibiotics and a repeat chest radiograph showed a suspicious radiolucent shadow in the right lower lobe. An upper gastrointestinal contrast study confirmed the diagnosis of right-sided congenital Morgagni hernia. Subsequently the diaphragmatic hernia was repaired and the infant made a quick recovery.

Keywords: Congenital hernia, diaphragmatic hernia, upper gastrointestinal contrast study

INTRODUCTION

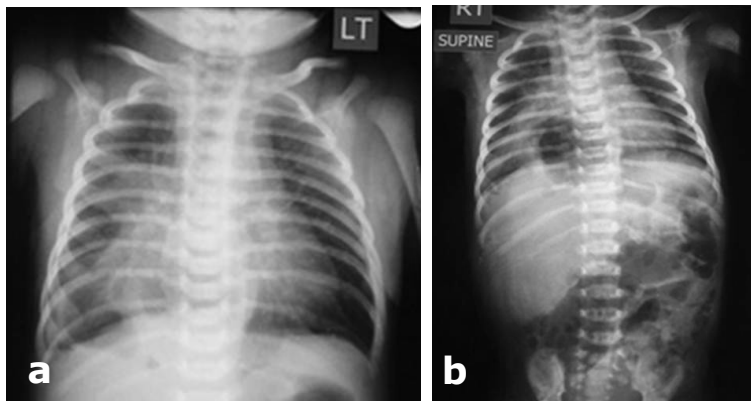
Right-sided congenital diaphragmatic hernia (R-CDH) is uncommon. Less than 20% of CDH present beyond the neonatal period. In most neonates with left-sided CDH (L-CDH), the clinical and radiological features are classical and the management present a major therapeutic challenge. R-CDH is difficult to diagnose in foetal life as well as in the neonatal period because the liver seals the defect in the diaphragm, restricting the thoracic migration of the gut. This case highlights the need

to consider CDH in the differential diagnosis of radiographic opacities resembling pneumonia affecting the right lung lower lobes. Clinicians can be misled by right lung opacities and the patient may be treated for pneumonia. ¹ The importance of discussing this case lies in recognising the varied signs and symptoms of CDH, and to consider it in the differential diagnosis of such presentations in clinical practice, especially when CDH presents as an isolated anomaly. Timely diagnosis and treatment can reduce the morbidities related to CDH.

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CASE REPORT

A female term baby weighing 2900 gm was



Figs. 1: a) A chest radiograph showing a non-specific opacity in the right middle and lower zone of the chest suggesting pneumonic changes, and b) a supine radiograph demonstrating lucency at the medial aspect of the opacity in the right lower chest. No definite

born by normal vaginal delivery to a primigravida mother following an uneventful antenatal period. The parents were non-consanguineous and of Chinese origin. Antenatal scans were normal. Maternal serologies were unremarkable and group B streptococcus status was unknown. There was no clinical or biochemical evidence of chorioamnionitis. At birth, the Apgar scores were 9 @1 and 5 minutes of life. The baby remained well in room air and was breast-fed. She was discharged home on day two of life.

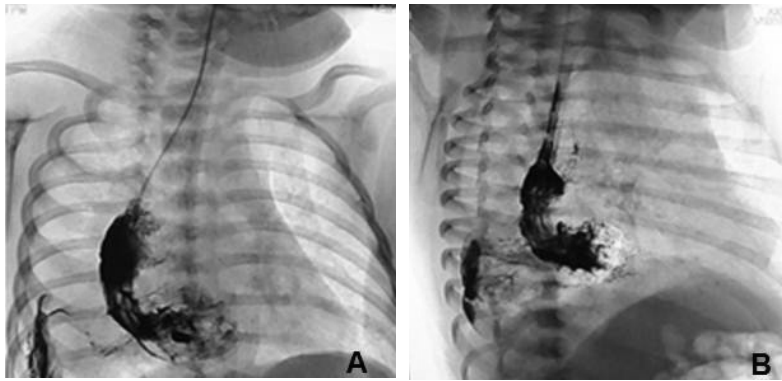
On the tenth day of life she was brought to the emergency department with lethargy, poor feeding and non-bilious vomiting. On examination she had a weight loss of 15%. She was non-dysmorphic, afebrile and pink with good peripheral pulses. Her heart rate and blood pressure were within normal limits and was saturating 96-98% in room air. Systemic examination revealed bilaterally equal air entry over the lung fields and no audible murmurs. She had reduced bowel and bladder output and abdominal examination was unremarkable.

On admission, her full blood count and C-reactive protein (CRP) were unremark-

able. Chest radiograph was reported as showing a non-specific opacity of the middle and lower lobes of the right lung (Figure 1a), suggestive of pneumonia, possibly following milk aspiration. Blood culture was negative and a repeat screening for septic markers was unremarkable. The parents were concerned about the persistent poor suck and lethargy even after a week of parenteral antibiotics. A repeat radiograph was done and this raised the suspicion of a R-CDH (Figure 1b).

An ultrasound scan of the chest showed intestinal loops in the right hemithorax. An upper gastrointestinal contrast study showed that the stomach and bowel loops located in the right chest, confirming the diagnosis of a R-CDH (Figures 2). Echocardiography showed a structurally normal heart with a small patent ductus arteriosus. Chromosome study was reported as normal 46XX karyotype.

Following confirmation of a R-CDH, surgical repair of the diaphragmatic hernia was performed without any complication. The patient recovered well and by the seventh day of diaphragmatic repair was breast-feeding successfully. She was reviewed two weeks



Figs. 2: An Upper gastrointestinal contrast study; A) AP view B) right lateral view showing the outlines the stomach in the right chest. The antrum of the stomach corresponded to the opacity seen in Figure 1 and the duodenum is also seen in the chest.

after discharge and was found to be breast-feeding well and gaining weight.

DISCUSSION

CDH is a neonatal emergency. R-CDH is uncommon and physicians are more familiar with L-CDH and its classical presentation. The Morgagni type of R-CDH in infants is challenging because of its rarity, non-specific symptomatology, subtle clinical signs and radiological misdiagnosis due to intra-thoracic herniation of the liver.^{2,3} Late-presenting CDH may present with varied gastrointestinal or respiratory symptoms with onset of symptoms delayed even up to adulthood.² Right lung opacities may masquerade as pneumonia, pleural effusion, pneumothorax or asymptomatic intra-thoracic mass.¹

CDH is characterised by the presence of an orifice in the diaphragm through which the abdominal contents herniate in to the chest. This herniating mass includes stomach, loops of intestine, spleen, omentum with fat and often liver in infants with R-CDH (80%).^{2,3} Congenital diaphragmatic hernia occurs with a frequency of 1 in 3000 live births, 85% on the left (90% - Bochdalek defect), 10% on the right side (90% - Morgagni defect) and approximately 5% are bilateral.²

Morgagni type of CDH, first described by Giovanni Morgagni and has a male preponderance.³ Morgagni hernia occurs in the anterior midline through the sternocostal hiatus of the diaphragm whereas Bochdalek hernia is a postero-lateral defect due to either absent migration of the diaphragmatic musculature or failure of the pleuroperitoneal folds to develop. Associated congenital anomalies in the Morgagni type of CDH was reported to be 71.7%, of which 39.6% had congenital heart disease (ventricular and atrial septal defects) and 28.3% had Down syndrome.³

Prenatal diagnosis of CDH are made more often on the left than right.⁴ Fetal diagnosis of L-CDH is based on the deviation of the cardiac axis and the presence of stomach bubble in the chest. In sliding hernias, the diagnosis may be impossible in foetal scan. Above all, in R-CDH, ultrasound scan image of the herniated intra-thoracic liver may resemble pulmonary tissue.⁵ At birth, the clinical presentation of large L-CDH is usually one of respiratory distress and poor oxygenation due to pulmonary hypoplasia, severe persistent pulmonary hypertension and less often with gastrointestinal symptoms like vomiting or intestinal obstruction. The incidence of delayed presentation varies from

from 5% to as high as 45% of all reported cases of CDH. ⁶ Kitano *et al.* observed a higher incidence of late-presenting CDH on the left (78.4%) than on the right side (21.6%). ⁶ A delayed diagnosis of R-CDH was made in infancy in 65%, 1-5 years in 20% and above five years in 15% cases. ⁷ L-CDH, more often presented with gastrointestinal symptoms whereas respiratory symptoms (81% cases) dominated in R-CDH. ^{3, 7} On the contrary, gastrointestinal symptoms were the only presenting feature in this reported case. Postnatal diagnosis is based on chest x-ray, ultrasound scan, computed tomography scan and upper gastrointestinal contrast study. Pneumonia and pneumothorax were diagnosed due to misleading symptoms and misinterpreted imaging findings, in few published case reports ⁷ similar to our case. Once diagnosis is confirmed, stabilisation of cardio-respiratory status will be followed by repair of the diaphragm. Outcome of children and adults following surgical repair of Morgagni defects were reported to be good except in cases with higher intrathoracic hepatic herniation. ⁷⁻⁹ Extracorporeal membrane oxygenation increased the survival of babies with R-CDH than L-CDH. Higher morbidities including chronic lung disease, gastro-oesophageal reflux disease and neuro-developmental deficits were reported with R- CDH. ¹⁰

In summary, this case report is an example of congenital right-sided Morgagni hernia presenting in the 2nd week of life with gastrointestinal symptoms. Chest x-ray lead to misdiagnosis of pneumonia and further imaging studies following unresponsiveness to medical treatment resulted in confirmation of the diagnosis. Early diagnosis and timely re-

pair of the diaphragmatic defect will reduce the long-term respiratory and gastrointestinal morbidities.

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