CASE REPORT

Hepatopulmonary fusion: a rare and potentially lethal association with right congenital diaphragmatic hernia

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SUMMARY

Hepatopulmonary fusion is an extremely rare accompaniment of right congenital diaphragmatic hernia. It is associated with abnormal systemic arterial supply and venous drainage of the right lung along with congenital heart disease. Children with this condition have a comparatively poor prognosis. We report a case of right congenital diaphragmatic hernia with hepatopulmonary fusion along with review of the literature with stress on diagnosis and management.

BACKGROUND

Congenital diaphragmatic hernia (CDH) is a relatively common congenital malformation which affects one in 2500–3000 live births. Left-sided lesions are more common, with the right side being involved in just 15–20% of the cases.¹ Outcomes of right-sided lesions are in general poor as compared with the left-sided lesions.² Hepatopulmonary fusion is an extremely rare abnormality, which, if associated with right-sided CDH, often has a fatal outcome. We present a case of right-sided CDH with hepatopulmonary fusion along with extensive review of the literature.

CASE PRESENTATION

A 2-month-old girl presented to us with symptoms of respiratory distress and two episodes of lower respiratory tract infection. No antenatal ultrasound scan had been performed. The antenatal period was uneventful. The child was a normal vaginal delivery at home with no significant perinatal event at birth. The child had increased respiratory rate for which she was admitted twice elsewhere and managed with antibiotics and inhalational oxygen with temporary relief. Owing to persistence of symptoms the child was referred to us. On examination the child was afebrile and haemodynamically stable. The child had respiratory rate of 60/min with mild chest retractions. On auscultation the breath sounds were absent in the right lower chest and were normal on the left side. Heart sounds were normal. The child was maintaining an oxygen saturation of 93% on room air and the arterial blood gas analysis was within normal limits.

INVESTIGATIONS

The chest radiograph and a non-contrast CT had been performed which suggested presence of right eventration of diaphragm with elevation of her

liver into the right hemithorax with ipsilateral deviation of the mediastinum (figure 1). CT also revealed the presence of small opacities in the right lung suggestive of consolidation. A preoperative diagnosis of eventration was made and no further investigations were performed.

TREATMENT

With a diagnosis of right eventration of diaphragm the child was taken up for thoracoscopic repair. A 5 mm port was inserted into the 5th intercostal space in the midaxillary line. On inserting the telescope, it was noticed that extrapleural space had been entered and the pleura was adherent to the underlying lung. Thoracoscopy was abandoned and a right posterolateral thoracotomy was performed. A 5 cm×4 cm defect in the right hemidiaphragm was noted. Owing to this defect there was hepatopulmonary fusion between the right lower lobe of the lung and the superior surface of the liver. The lower part of the lung was not aerated and was receiving blood supply from multiple vessels which were coming through the diaphragmatic defect. Therefore, a diagnosis of intrapulmonary sequestration was considered. This suspected sequestration was also draining into a prominent vein at the medial end of the defect which was continuing inferiorly through the diaphragmatic defect (figure 2). There were dense adhesions in between the lung and the liver which were carefully separated. The arteries supplying the sequestration were ligated and divided. The vein at the medial end was also receiving tributaries from the upper lobe and was preserved. Only the tributary from the sequestration to this vein was divided. A bronchial communication was also noticed which was divided and lower lobe of right lung was resected. The diaphragm was repaired with interrupted 3–0 Prolene sutures. There was high insertion of the diaphragm and medially the edges of the diaphragm reached the level of 5th thoracic vertebrae. The medial end of the diaphragmatic defect was not repaired so as not to constrict the vein, which was passing through the defect.

OUTCOME AND FOLLOW-UP

The child has intermittent episodes of hypoxia after the surgery, which was initially responsive to rapid ventilation with a manual resuscitator. The episodes of hypoxia became more severe and did not respond to resuscitation. Investigations were

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Figure 1 (A) Plain chest radiograph reveals opacity in right lower thorax continuous with the hepatic shadow suggestive of right eventration/diaphragmatic hernia. White arrowhead denotes the presence of ipsilateral mediastinal deviation. (B) Non-contrast CT confirms the finding of the chest radiograph.

Figure 2 Intraoperative photograph following separation of the lung and the liver. Presence of aberrant artery (white arrowhead) and aberrant vein (black arrowhead) is also noted. The right lung has been retracted.

suggestive of respiratory failure with normal renal function test and bilirubin. The child expired within 12 hours after surgery. The parents refused consent for an autopsy.

DISCUSSION

Right diaphragmatic hernia is responsible for ~15% of all cases of CDH. Rarely this condition is associated with hepatopulmonary fusion. Hepatopulmonary fusion is an extremely rare condition with only 15 cases reported in the literature.³ Hepatopulmonary fusion is usually associated with abnormal systemic vascular supply and venous drainage from the right lung through the diaphragmatic defect.⁴ It is often associated with congenital heart defects.

This condition can be considered as a continuation of spectrum of other similar conditions like congenital pulmonary venolobar syndrome⁵ and pseudosequestration syndrome.⁶ The term 'Pseudosequestration syndrome' was used by Macpherson *et al* to describe three cases of abnormal systemic circulation to the right lower lobe of the lung along with right diaphragmatic defect, but since there was normal bronchial communication in these cases the term 'Pseudosequestration' was used. In one such patient they also noticed adhesions between the right lower lobe and liver. Congenital pulmonary venolobar syndrome is a syndrome in which presence of a hypogenetic lung and partial anomalous pulmonary venous return are the two most consistent findings.⁵ However, other associated findings may include systemic arterial supply to the lung, diaphragmatic defects, pulmonary sequestration etc.

Hepatopulmonary fusion can markedly compromise the prognosis of the child. Of the 16 cases of hepatopulmonary fusion described in the literature including the index case, 6 of them died.^{7 8} High mortality and morbidity in these cases can be attributed to hypoplastic lung and associated cardiac abnormalities. It is therefore imperative that this condition should be suspected in the preoperative period. The most characteristic feature mentioned for suspecting this condition is ipsilateral or absence of deviation of the mediastinum in patients with right congenital diaphragmatic hernia. In such patients if on further investigation left-sided congenital heart defects are detected, then the possibility of hepatopulmonary syndrome should seriously be considered.⁴ The absence of mediastinal deviation in these cases is because of hypoplasia of the right lung; however, in few patients with large diaphragmatic defect, mediastinal deviation was noted along with hepatopulmonary fusion.⁴ The definite diagnosis of this rare condition can only be made intraoperatively. Most of the times this condition is an intraoperative surprise for surgeons while operating for right diaphragmatic hernia. The differential diagnosis of this condition may include presence of ectopic liver tissue in thorax which is often misdiagnosed as a pulmonary mass.⁹ Another more common differential diagnosis can be pulmonary sequestration which is known to be associated with diaphragmatic hernia, where the collapsed lower lobe appears similar to liver tissue and is adherent to the diaphragm secondary to infection. There are two things unique about the index case, the child presented after 1 month of age whereas most cases described in the literature presented in the neonatal age. A unique finding which we noted in our case was high insertion of the diaphragm medially until the 5th vertebrae. Similar finding of high insertion of diaphragm has been reported in one case with hepatopulmonary fusion.⁷

Management of this condition is still not clear as very few cases have been reported in the literature. It is generally agreed that an attempt should be made to separate the thoracic and abdominal cavities. If a plan can be developed between the liver and the lung then they should be separated and the diaphragm should be repaired. It is postulated that the negative intrathoracic pressure will aid in the growth of the hypoplastic lung. However, if the separation between the lung and the liver becomes difficult some authors have recommended to suture the diaphragm to the margins of the remaining fusion.¹⁰ Others have recommended sacrificing the abnormal lower lobe of the lung to achieve complete separation.⁴ The management of the aberrant vessels that cross the diaphragm is also unclear with most of the published reports failing to describe this aspect of management. A recent report in which a right-sided CDH was

associated with abnormal systemic supply and venous drainage to the right lung, the authors divided these aberrant vessels.¹¹ The child survived after the surgery even though most of the venous drainage and the systemic supply to the right lung was divided. If suspected before the surgery, the case should be evaluated and discussed with cardiologist and pulmonologist. If the diagnosis is made intraoperatively, separation of the hepatopulmonary fusion can be attempted to the extent possible, the diaphragm attached to the margin of the fusion with a plan to further investigate the child with echocardiography, MR angiography, etc. Thereafter, a collective decision can be made after consulting other relevant specialists. This staged approach has been described previously.^{8 11}

Hepatopulmonary fusion is a rare abnormality and should be suspected if there is presence of right diaphragmatic hernia with absence of mediastinal deviation. Such cases should be investigated with echocardiography and if need angiography to confirm the diagnosis.

Learning points

- ▶ Hepatopulmonary fusion is an extremely rare condition associated with right congenital diaphragmatic hernia.
- ▶ It should be suspected in patients with right congenital diaphragmatic hernia with absence or ipsilateral deviation of mediastinum.
- ▶ These patients need detailed cardiac evaluation before surgery.
- ▶ Presence of this condition further worsens the prognosis of the patient.

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