Congenital focal eventration of the left hemidiaphragm: diagnostic dilemma resolved on prenatal MRI

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DESCRIPTION

A 28-year-old woman, G2A1, was referred to our institute at 24 weeks of gestation with a prenatal ultrasound diagnosis of left-sided congenital diaphragmatic hernia (CDH) in the fetus. Combined first trimester screen revealed a low risk for aneuploidy. A repeat ultrasound at 26 weeks of gestation at our institute confirmed intrathoracic herniation of the liver on the left side; however, stomach bubble was noted within the abdomen (figure 1A,B). The right hemidiaphragm was identified on ultrasound; however, the left hemidiaphragm was not clearly seen. Mediastinal shift to the right was noted with a structurally normal heart. There was no ascites, pleural effusion or polyhydramnios. As the stomach, which is the most common structure to herniate in a left-sided CDH, was intra-abdominal in position with no cranial migration of abdominal viscera into the thoracic cavity during fetal breathing movements, a possibility of a very thinned-out diaphragm as seen in congenital diaphragmatic eventration (CDE) was also considered. A prenatal MRI revealed a focal bulge in an intact left hemidiaphragm with superior displacement of the left lobe of liver (figure 1C,D). The sagittal image showed the intact T2 hypointense left hemidiaphragm with focal anterior bulge, confirming the diagnosis of left-sided CDE.

The antenatal period was uneventful and a male baby weighing 3510 g was delivered at 37 weeks 4 days of gestation with an Apgar score of 8 and 9 at 1 and 5 min, respectively. The baby was shifted on oxygen to neonatal intensive care unit in view of respiratory distress at birth; however, he did not require any other type of respiratory support. A contrast-enhanced CT of the chest done on postnatal day 3 confirmed left-sided CDE (figure 2). The baby was discharged on day 7 and kept on regular follow-up. Left posterolateral thoracotomy and plication of left hemidiaphragm was performed at the age of 1 year and 4 months. Per-operative findings revealed thinned-out left hemidiaphragm with large bowel extending into left hemithorax with adequate expansion of left lung in both lobes. The baby was discharged in a stable condition.

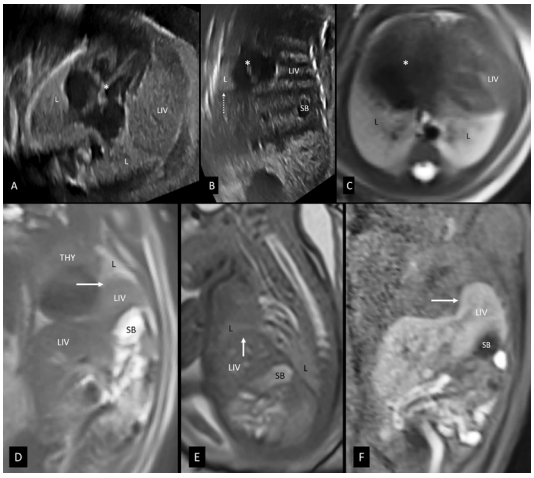

Figure 1 Prenatal ultrasound axial image (A) at the level of heart (*) reveals intrathoracic herniation of the liver with mediastinal shift to the right. Anterior coronal image (B) reveals intrathoracic herniation of the liver; stomach bubble appears intra-abdominal. There is mediastinal shift to the right. Right hemidiaphragm is visualised (dotted white arrow). Axial T2-weighted HASTE (Half-Fourier Acquisition Single-shot Turbo spin Echo imaging) image (C) of prenatal MRI shows intrathoracic liver and displacement of the heart (*) to the right side. Note the normal signal intensity of bilateral lungs. (D) Coronal T2-weighted HASTE (Half-Fourier Acquisition Single-shot Turbo spin Echo imaging) image shows a focal bulge in the left hemidiaphragm (arrow) with superior displacement of the left lobe of the liver. (E) Sagittal TRUFISP (True fast imaging with steady state precession) image shows complete T2 hypointense left hemidiaphragm with focal anterior bulge (arrow). (F) Coronal T1-weighted VIBE (Volumetric interpolated breath-hold examination) image clearly depicts the focal bulge in the intact left hemidiaphragm (arrow), confirming the diagnosis of focal eventration of the left hemidiaphragm. L, lungs; LIV, liver; SB, stomach bubble; THY, thymus.

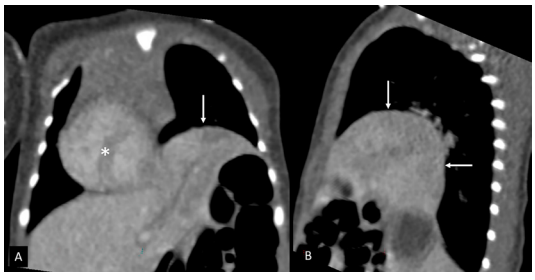

Figure 2 Postnatal contrast-enhanced CT. Coronal (A) and sagittal reformatted images (B) reveal focal elevation of left hemidiaphragm anteriorly (arrow in A and B) with mediastinal shift to the right (*). There was no focal discontinuity of diaphragm, confirming the diagnosis of focal eventration of the left hemidiaphragm.

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CDE is uncommon and accounts for 5% of all diaphragmatic abnormalities.[1] It results from partial muscularisation of diaphragm and replacement of muscle by a thin membranous sheet leading to protrusion of abdominal viscera into the thorax. Thus, it can mimic CDH which is more prevalent resulting in misdiagnosis.[2] Although both entities are clinically distinct, the ultrasound findings are almost similar. However, as fetal outcomes are much worse in CDH with a high postnatal mortality rate and need for immediate surgical repair in all cases, prenatal differentiation of these entities is critical.[3] For prenatal diagnosis of CDE, visualisation of a continuous diaphragm delineating the abdominal and thoracic cavity is necessary, which is often difficult on ultrasound, as seen in our case. Fetal MRI is more beneficial as it allows a three-dimensional evaluation of the diaphragm and is not limited by position of the fetus or maternal body habitus.[4] Moreover, multiplanar views provide a detailed picture of fetal anatomy, even in advanced pregnancy. In cases where a congenital diaphragmatic anomaly is suspected on ultrasound with visualisation of intra-abdominal contents in thoracic cavity, a prenatal MRI can help to differentiate between CDH and CDE.

Learning points

- Congenital eventration of the diaphragm is an abnormal elevation of diaphragm as a consequence of partial muscularisation of the diaphragm with a thin membranous sheet replacing the normal muscle of the diaphragm.
- It is difficult to differentiate congenital eventration of the diaphragm from congenital diaphragmatic hernia based on findings of prenatal ultrasound and fetal MRI may be useful in these cases.
- Prenatal differentiation between the entities is important as the perinatal management and prognosis for both is different.

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