Olgu Sunumu Case Report

Jinekoloji - Obstetrik ve Neonatoloji Tıp Dergisi
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What Is Your Diagnosis?

Tanınız Nedır?

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A 29-year old multiparous pregnant woman was referred to our prenatal medicine outpatient clinic due to polyhydramnios. Ultrasonographic imageof fetal thorax is seen in Figure-1, 2 and 3. What is your diagnosis?

Figure-1: A solid mass (white arrow) is seen in fetal thorax next to the heart (red arrow).

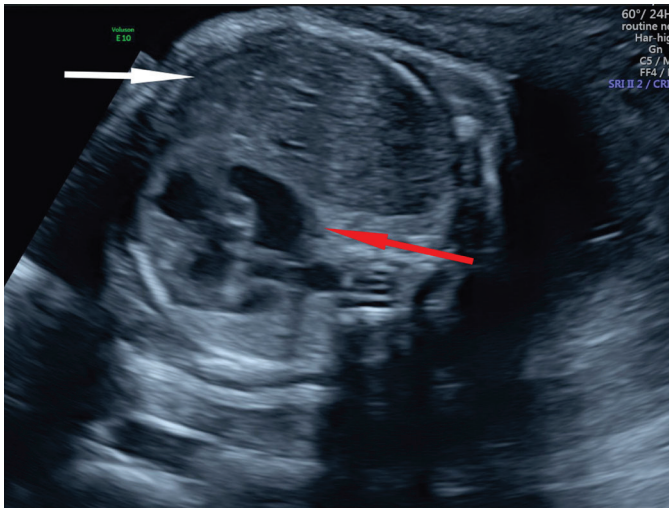
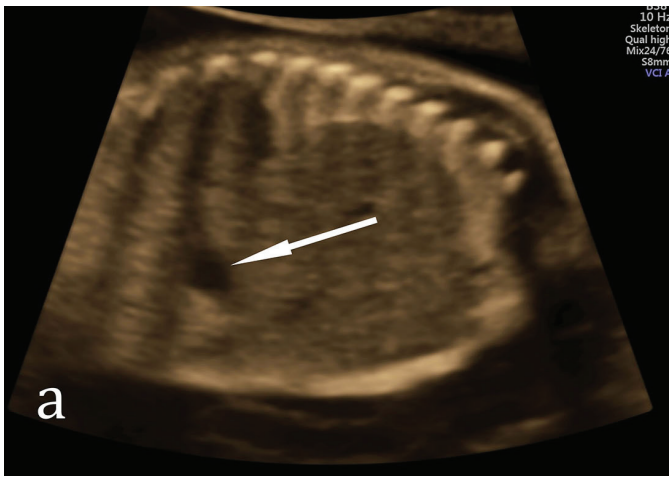


Figure-2: (a) the stomach; (b) the left mediastinal shift: the extremely left sided heart is seen just above the stomach. Next to the fetal heart, a right-sided solid mass is viewed.



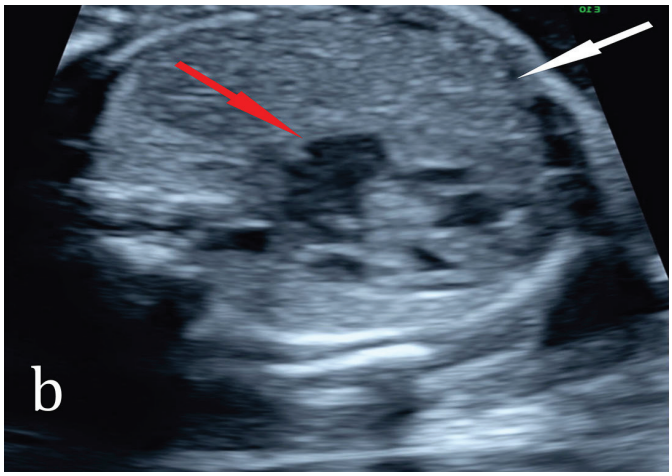
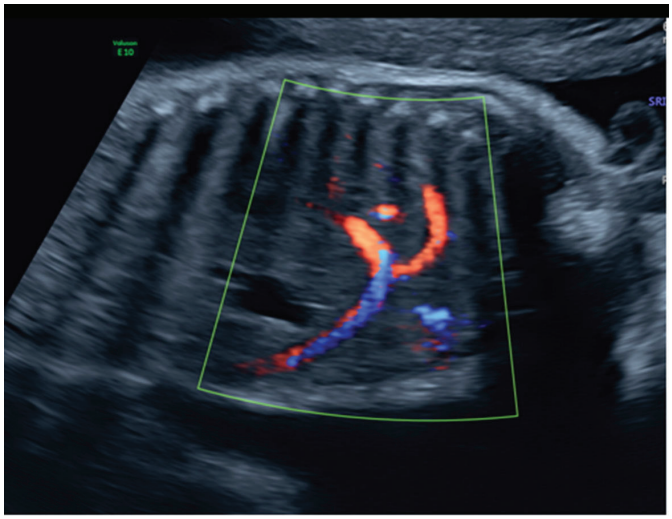


Figure-3: Intrahepatic vasculature is discovered with color mapping.



Answer

Figure-1 demonstrates the position of the liver inside the fetal thorax next to the fetal heart. As a consequence, there is left mediastinal shift and a displacement of the heart. Although Figure-1 focuses on the position of the liver and not on the structure of the heart, a large ventricular septum defect can be suspected.

In the right parasagittal view of Figure 2, the remaining part of the right diaph-

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ragm hernia can be seen. Due to the large hernia, the liver is placed inside the thorax.

Color Doppler mapping in Figure-3 demonstrates the intrahepatic vessels that are placed above the level of the diaphragm.

A right-sided diaphragmatic hernia was diagnosed.

The prevalence of congenital diaphragmatic hernia (CDH) is approximately 2-3 per 10.000 births (1). Right-sided herniation is very rare and covers about 10% to 15% of these cases. Prenatal diagnosis of right sided CDH is challenging, due to the echogenity of the liver that is similar to that of the lungs. The detection rate depends on gestational age (<50% detected before 24 weeks), associated anomalies and especially on the experience of the clinician (2).

A right-sided CDH should be suspected if there is left mediastinal shift with the gallbladder being inside the fetal thorax and the stomach inside the abdomen, respectively. Color Doppler flow may help to map the hepatic vasculature (4).

CDH is an isolated finding in about half of the cases. However, it can also be associated with other structural defects, such as cardiac defects, numerical and structural chromosomal abnormalities, such as trisomy 18 and mosaic tetrasomy 12p (Pallister-Killian syndrome) and some genetic syndromes, such as Fryns or Cornelia De Lange syndrome, respectively (2).

Postnatal mortality and morbidity depends on the associated anomalies, on the position of the liver and on theremaining size of the lungs. With decreasing volume of the lungs, the number of vessels and bronchioli decreases resulting in a hampered oxygenation and pulmonary hypertension.The chance of survival can be estimated by the lung to head ratio (LHR) and the respective observed to expected LHR (o/e LHR). To compute the LHR, the area of the contralateral lung is divided by head circumference. In general, the ratio between the observed and expected LHR is used for counseling as this marker is independent from gestational age.In a series of 161 fetuses with isolated left sided CDH and intrathoratic liver herniation, about half of the neonates were discharged from hospital alive. The chance of postnatal survival was (258 × (o/e LHR (%))−28,68)/100. In this registry there were also eight cases with isolated right sided CDH and none of the cases survived after birth. In cases with severe CDH (o/e LHR < 25%), fetal endoscopic tracheal occlusion can be offered which may increase the survival rate (5).

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