

Short Clinical Report

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A case of hernia of the umbilical cord with an accessory liver

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CASE PRESENTATION

A female neonate was born at 38 weeks, 4 days gestation via normal vaginal delivery with a birth weight of 2.984 kg; both 1- and 5-min APGAR scores were 9. She had been suspected of having a small omphalocele with an umbilical cord cyst on prenatal ultrasound at 25 weeks of gestation. Repeated ultrasonography every two weeks did not change the findings of umbilical cord cysts and small omphalocele. After birth, her umbilical cord base was 20 mm in diameter, with an elastic rigid tumor-like structure instead of an umbilical cord (Fig. 1A), and a definitive diagnosis of hernia of the umbilical cord with accessory liver was confirmed following computed tomography scan (Fig. 1B).

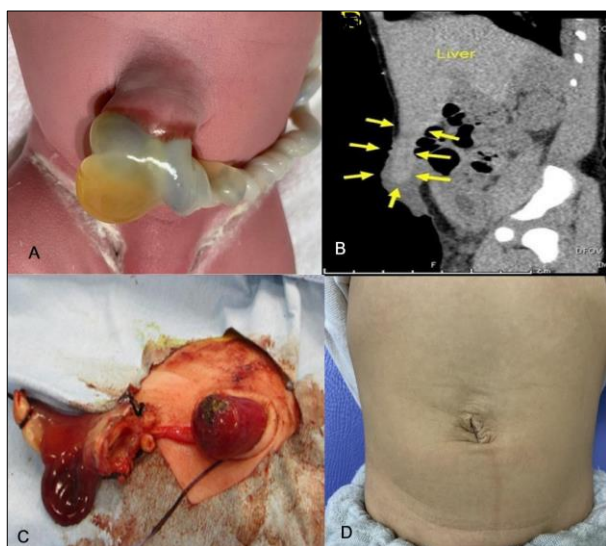


Figure 1: A) Pre-operative picture showing hernia of umbilical cord. B) CT scan showing accessory liver herniating through abdominal wall defect (Arrows). C) Peroperative figure showing dissected accessory liver tissue. D) Follow up after one year of umbilicoplasty.

As the herniation was not reducible. Blood examination after birth showed AST/ALT 394/10U/L, LDH

1291U/L, and CK 610U/L, there was a possibility of reduced blood flow to the liver. Her postnatal condition was stable, she was operated on the day of her birth. After making a circular skin incision around the umbilical cord and a longitudinal opening of the linea alba, the round ligament of the liver and medial and lateral umbilical ligaments were ligated and cut simultaneously, and the abdominal cavity was accessed. The accessory liver protruded from the edge of the lateral segment of the liver and firmly adhered to the umbilical cord. It was approximately 15 mm in size and appeared congestive and dark red compared to the main liver. Immediately after a careful adhesiolysis using an electric cautery, the invaginated tissue was restored to its normal condition and then preserved and returned into the abdominal cavity (Fig. 1C). The postoperative course was uneventful, without signs of torsion of the accessory liver. Currently, the patient is a 1-year-old healthy girl with a satisfactory navel appearance (Fig. 1D).

DISCUSSION

Hernia of the umbilical cord (HUC) is a failure of the umbilical ring's morphogenesis that occurs around 4 weeks of gestation, it is thought to develop due to abnormalities in the reducing process of physiological umbilical herniation at ≥ 10 weeks of gestation.[1–3] Glenisson et al. recommended classifying accessory liver defects according to their morphological presentation: A) hepatic parenchymal tissue is interposed between the hepatic parenchymal and primary liver tissue, B) the main liver organ and hepatic parenchymal tissue are connected by a peduncle, and C) the hepatic parenchymal tissue mass is completely separated from the liver.[4] Our case met type A according to this classification.

A review of the literature revealed that the number of HUC cases with an accessory liver is small.[1–3,6,7] Consistent with our findings, the firmly adhered accessory liver to the hernial sac of the umbilical cord

has been reported previously. This finding might suggest morphogenetic involvement of the adhesion in the liver bud to the umbilical coelom because the liver bud and umbilical cord are very close to each other at around 4 weeks of gestation when the umbilical ring begins to form. In early reports, the accessory liver had been aggressively resected [1,5] although it has been avoided in recent reports.[2,3,6] According to descriptions of the torsion of the accessory liver,[7] the accessory liver's size is unlikely to develop a torsion. Thus, considering the risk of sequelae such as bleeding from the cut surface and post-surgical adhesion, preserving the invaginated accessory liver and

leaving it in the abdominal cavity would be the best choice.

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