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Hernia into the umbilical cord with incarceration of liver and gall bladder in a newborn

Nahidh W. Hasaniya^{a,d,e}, Shyamal Premaratne^{b,c,d,e,*}, Philip M. Varnes^c, David Shin^f, Walton Shim^f^a Department of Surgery, School of Medicine, Loma Linda University, Loma Linda, CA, USA^b Hunter Holmes McGuire Veterans Administration Medical Center, Richmond, VA, USA^c Virginia Union University, Richmond, VA, USA^d Department of Surgery, John A. Burns School of Medicine, University of Hawaii and, Honolulu, HI, USA^e Research Laboratory at the Queen's Medical Center, Honolulu, HI, USA^f Kapiolani Medical Center for Women and Children, Honolulu, HI, USA

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ABSTRACT

Incarceration of liver tissue in a hernia of the umbilical cord is a rare occurrence. An incarceration of the gall bladder is even rarer. We report such a case in a newborn that had incarceration of both liver and gall bladder in to the umbilical cord.

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Abdominal wall hernias are among the most common of all surgical problems. The umbilical cord is the critical connection between the embryo (and later, the fetus) and the placenta. It houses the blood vessels that are responsible for nourishing the fetus. Proper functioning of the umbilical cord is essential for the growth and development prior to birth. A review of the embryologic origin of the umbilical cord aids in the understanding of abdominal wall and umbilical cord defects that can present in the newborn period. In the delivery room, inspection surrounding the base of the cord may necessitate a delay in shortening the cord [1,2]. Surgical consultation may also be indicated. It is a rare phenomenon to describe an incarceration of a newborn liver in a hernia of the umbilical cord [3–11]. An incarceration of the gall bladder is even

more rare [5–8]. We report such a case of a newborn with incarceration of liver tissue and gall bladder in a herniation into the umbilical cord.

1. Case report

A day-old, full-term male newborn was transferred to Kapiolani Medical Center for women and children in Honolulu, HI due to the presence of a mass at the base of his umbilical cord. The child was asymptomatic. On examination, a 2 cm × 1.5 cm firm mass was found at the base of the cord which could not be reduced. The abdominal skin extended for approximately 1 cm up the umbilical cord.

On auscultation, the child had a grade II/IV systolic murmur. An echocardiogram led to the diagnosis of Tetralogy of Fallot, a common but serious cyanotic congenital cardiac defect. There was also a partial amputation of his right thumb. At surgery, a 1 cm nubbin of liver tissue and gall bladder were found adherent to and incarcerated in the 2 cm hernia sac with a 1 cm fascial defect. The gall bladder was rudimentary (Fig. 1). The incarcerated liver was connected to the main tissue mass of the liver by a narrow band of liver

* Corresponding author. Hunter Holmes McGuire Veterans Administration Medical Center, Mail Stop 151, 1201 Broad Rock Boulevard, Richmond, VA 23249-0001, USA. Tel.: +1 804 675 5000; fax: +1 804 675 5359.

E-mail address: spremaratne@vuu.edu (S. Premaratne).

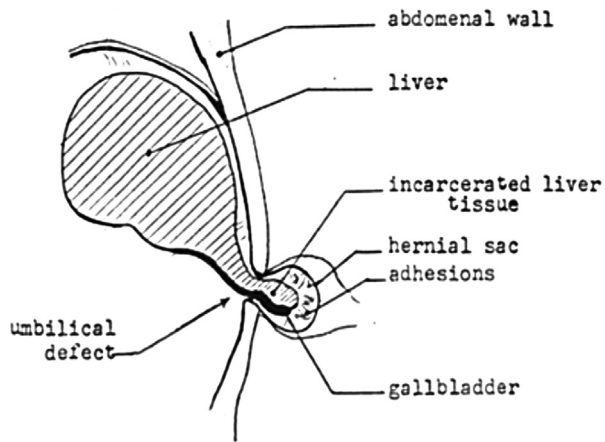


Fig. 1. Sagittal section, showing the incarcerated liver substance and rudimentary gall bladder in the umbilical hernia.

tissue, which accompanied the neck of the gall bladder and the cystic duct (Fig. 2). The patient was also found to have malrotation of the bowel. The cystic duct along with a narrow band of liver was ligated and amputated. The umbilical hernia was excised and closed. Recovery was uneventful and he was discharged home without any complications.

2. Discussion

Herniation of intestines into the extracelomic cavity of the umbilical cord is a natural developmental phase of the early embryo. The presence of liver in this normal herniation has not been documented, but could happen in a large proportion of cases since up to half of all omphaloceles contain liver tissue [5,7]. The adherence of the liver to the omphalocele lining could then result in a portion of the liver being incarcerated into the umbilical cord. Whether or not this represents an “accessory lobe” has not been proven by anatomical studies. Such accessory lobes have been reported including in a recent work up of a child with a respiratory infection [12]. Even so, it appears doubtful that this would be a true accessory lobe. The presence of the gall bladder with the incarcerated liver would tend to negate the accessory lobe theory as the gall bladder straddles the interlobular boundary. Its presence in the hernia probably means a portion of both the left and right lobes are caught in the hernia.

Some authors have used the term, ‘ectopic,’ which it most certainly is [13]. However, we feel that it should be more correctly designated as an ‘incarcerated liver substance’ [8–10]. The decision to excise the pedunculated liver tissue and gall bladder is supported by reports such as those of Azmy et al. [6], who reported torsion of a herniated gall bladder in a similar case.

A valuable point brought up by Tan and Tan is that there may be an association between liver incarceration and biliary atresia [10]. Feston and colleagues advise that cholangiography can be performed in cases where the gall bladder is incarcerated (to rule out biliary atresia) since it can be used to provide a connection between the liver and the intestines [8].



Fig. 2. Photomicrograph of rudimentary gall bladder embedded in an incarcerated liver.

3. Conclusions

It is important to investigate the presence of accessory liver tissue with or without a gall bladder in an umbilical hernia so as to not miss a diagnosis of biliary atresia and to preserve the gall bladder as a possible conduit if there is a biliary atresia as suggested by Feston and colleagues [8]. Cholangiography may be done to rule out biliary atresia in cases where this diagnosis is suspected. Although previous authors have considered this finding an accessory lobe or an ectopic liver, it may be more appropriate to designate it as an incarcerated liver substance.

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