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Case Report

Ectopic Liver Tissue in Hernia of Umbilical Cord

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Abstract

A mass in umbilical cord hernia is a rare clinical presentation in neonates. Here a case of heterotopic liver within the hernia of umbilical cord in a one day old newborn. The baby was born in 38 weeks gestation, and weighed 3.200 kg at birth.

He presented with a congenital protruding umbilical cord containing a solid mass measuring 2.8 cm in its greatest dimension. Abdominal ultrasonography found no connection with the visceral organ.

The histology of the solid lesion consisted of epithelial cells, which were laid as plates or cords and separated by a thin vascular channel. This has given it a similar morphology of hepatocytes.

This is extremely rare and we are reporting this case of ectopic liver as new presentation. The aim of our case presentation is to draw medical society attention to this possible pathological variation and the surgical treatment of this condition

Introduction

Heterotopia is a misplacement or displacement of an organ or the formation of tissue in a place where its present location is abnormal.

Umbilical cord hernia usually occupied by small or large bowel. If there is a mass we should suspect a differential diagnoses of such lesion to be either hemangioma, hematoma, or teratoma (1).

We report a case of an heterotopic liver within the umbilical cord hernia of an 1 day old neonate.

Patient

A newborn baby was born at 38 weeks and 3 days of gestation and weighed 3.200kg at birth. He was presented with a congenital umbilical lesion, which appeared as a umbilical cord hernia (Figure 1), but no other anomalies was noted. The lesion was locally excised. Under general anesthesia with endotracheal tube we explore the content of hernia which was mainly a solid piece of tissue associated with a cystic like lesion

Specimen

Macroscopically

On gross examination, the lesion was a 2x 2.2x 1.2 cm located centrally in the umbilicus and associated with small cystic lesion connected to a long mesenteric vessels. The cut surface of that solid part was yellowish - grayish in color after formalin fixation.

Microscopically

The lesion consisted of epithelial cell nests and stroma composed of fibrous connective tissue, blood vessels, and nerve bundles. The epithelial cells were organized into plates or cords separated by thin vascular channels .The epithelial cells were polygonal in shape and contained eosinophilic cytoplasm with round and prominent nucleoli, morphologic features consistent with those of hepatocytes. Ducts and ductular structures compatible with a bile duct system which were well developed. While the cut surface of the cystic lesion was gray in color and measures 1 x1cm, histologically this lesion consisted of bowel.

Discussion

A mass involving the base of umbilical cord is rare. Because of recent developments in prenatal ultrasonography, early detection of such lesions has become relatively easy. The differential diagnoses

Table 1: Characteristics of reported cases of ectopic liver in the umbilicus (4).

Characteristics	Go and Cho	Wax *et al*	Preminger *et al.*	Park *et al*	Shaw and Pierog	Nora and Carr	So-Young Lee	This case
Associated clinical manifestations	Yes (intrauterine	no	no	Yes (bile duct obstruction	Yes (infection)	yes(death, hepatic necrosis)	no	no
Time of diagnosis	Prenatal (autopsy case)	Prenatal	After birth	After birth	After birth	After birth	After birth	After birth
Size (cm)	2	3.5	3	7.5	2	unknown	1.8	2.8

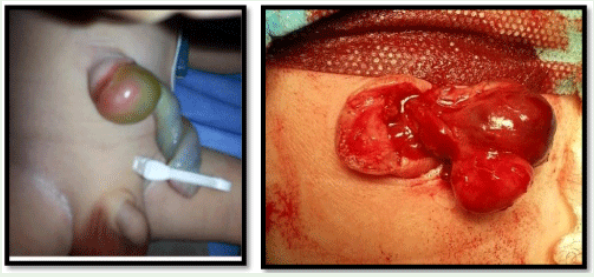

Figure 1: Saw scaled viper. Licensed under the Creative Commons Attribution-Share Alike 4.0 International license from Wikimedia [16].

of this mass include omphalocele hemangioma, and teratoma (1). Interestingly, the histology of the lesion in our patient was composed of hepatocytes.

Abnormalities in the position or number of liver parts are considered rare developmental anomalies; they are typically asymptomatic, and incidentally detected.

Although it is extremely rare, it may occur during an operation or autopsy. The liver develops from an endodermal bud from the most caudal part of the foregut around the fourth week of gestation. This hepatic bud divides into a larger cranial portion, the pars hepatica (forming the hepatic parenchyma), and a smaller caudal portion, the pars cystica (forming the extrabiliary system) (2).

The heterotopic hepatic tissue may have been differentiated from pluripotent endoderm, the urachal progenitor, in an ectopic location (1). Heterotopic liver tissue might have risen because of entrapment of liver cell nests in the foregut area after closure of the diaphragmatic or umbilical ring (3). Alternatively, liver tissue can migrate to various organs during embryogenesis. A review of the literature shows six other cases of heterotopic liver in umbilicus (Table 1).

Heterotopic liver has been reported in the esophagus, thorax, pericardium, testicle, and retro peritoneum (1-5). The most frequent organs in the abdominal cavity are gallbladder, gastrohepatic and umbilical ligaments, omentum and stomach. Above the diaphragm, it has been reported in the pleural cavity, mediastinum, lungs and heart. However, the gallbladder seems to be most frequently affected organ in the abdominal cavity (6).

Generally, the heterotopic liver present in the abdomen is usually asymptomatic. Our patient presented with no other intra-abdominal anomalies and was treated by simple surgical excision of the lesion without any complications.

The underlying basis for ectopic liver in cord is not exactly known. It is possible that a part of normal liver becomes entrapped outside the abdomen by the umbilical ring closure or there may be aberrant differentiation of the endoderm, which is common to the allantois, urachal progenitor and liver (7).

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