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Review of published cases of hepatic choristoma. Differential diagnosis of umbilical cord masses

Revisión de casos publicados de coristoma hepático. Diagnóstico diferencial de masas de cordón umbilical

Dear Editor:

We present the case of a primigravida, 33 years of age, with no medical or surgical history of interest. Ultrasound examination at 28 weeks of gestation confirmed the presence of a 28 mm × 17 mm mass in the umbilical cord, with an umbilical cord diameter of 16 mm, and a small anechoic area with thin walls suggestive of hernial oedema.

The patient had a normal delivery at 40 + 1 weeks of gestation, giving birth to a girl that weighed 3290 g and had an Apgar score of 9/10.

At birth, we observed an umbilical cord with a 4.5 cm × 2 cm × 1.8 cm bulge protruding from its normal insertion site at the abdomen, lined with amniotic membrane through which could be seen a firm, wine-red mass located 1 cm away from the navel that was irreducible, with no accompanying symptoms (Fig. 1). Based on the examination findings, we considered the differential diagnosis of abdominal wall defect and umbilical cord mass.

The surgery involved the opening of the amniotic membrane in layers, revealing a solid mass in direct contact with the umbilical vein and with an intraperitoneal communication with the round ligament of the liver. The vascular structures and remnants of the umbilical cord were ligated, the mass fully resected, and the umbilical defect closed. There were no postoperative complications and the patient was discharged 5 days after the surgery.

The mass was submitted to the anatomical pathology department for investigation, and gross examination showed a well-defined brownish nodule measuring 2.5 cm, with a microgranular appearance upon sectioning that corresponded to hepatic tissue with preserved architecture at the histological level. The tissue surrounded a cyst-like structure consisting of gallbladder wall tissue that was compatible with a hepatobiliary choristoma.

Ectopic liver is a rare condition described as the presence of hepatic tissue outside the liver and with no hepatic connection.

The literature has reported the gallbladder as the most common location of ectopic liver, and it can also be found in the thorax, pancreas, spleen, hepatic ligaments, pylorus, greater omentum, oesophagus, gastric mucosa, adrenal cortex, retroperitoneum, pericardium, placenta and umbilical cord.

Several theories attempt to explain the appearance of ectopic liver in locations other than the gallbladder, such as the development of an accessory lobe that loses its connection with the main liver body, the migration of part of the pars hepatica to other sites where ectopic tissue then develops, or the trapping of hepatocytes by the adjacent mesenchyma during the formation of the liver

Figure 1 Transillumination of the wine-red mass in the umbilical cord.

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<table>
<thead>
<tr>
<th>Maternal age Delivery</th>
<th>Suggestive manifestations</th>
<th>Gestational age</th>
<th>Weight (g)</th>
<th>Sex</th>
<th>Prenatal diagnosis</th>
<th>Location</th>
<th>Size</th>
<th>Colour</th>
<th>Consistency</th>
<th>Pathological anatomy</th>
<th>Peritoneal communication</th>
<th>Associated anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shaw and Pierog 19</td>
<td>Vaginal Yes</td>
<td>28</td>
<td>1247</td>
<td>Male</td>
<td>ND</td>
<td>Attached to navel by a stalk 2</td>
<td>7.5 cm × 6 cm</td>
<td>Red-purple</td>
<td>ND</td>
<td>Urachus cyst with immature hepatic tissue</td>
<td>Fimbly, atresia + ectopic pancreas in the jejunum</td>
<td>No</td>
</tr>
<tr>
<td>Park et al. 29</td>
<td>Vaginal</td>
<td>40</td>
<td>3240</td>
<td>Female</td>
<td>ND</td>
<td>CU insertion site</td>
<td>1.5 cm from the end of the UC</td>
<td>Yellow-green, brown interior</td>
<td>Firm, rubbery</td>
<td>Firm Normal hepatic tissue</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Preminger et al.</td>
<td>Caesarean RDS</td>
<td>35</td>
<td>2180</td>
<td>Female</td>
<td>No</td>
<td>1.5 cm from the end of the UC</td>
<td>2.5 cm × 3.5 cm</td>
<td>Dark red</td>
<td>Urachus cyst with numerous portal areas</td>
<td>Fimbly, atresia + ectopic pancreas in the jejunum</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Wax et al. 28</td>
<td>Caesarean RDS</td>
<td>38</td>
<td>3314</td>
<td>Female</td>
<td>19 week US: UC</td>
<td>Proximal to UC and navel</td>
<td>2.5 cm × 3.5 cm</td>
<td>Urachus</td>
<td>Hepatic tissue with numerous portal areas</td>
<td>Firm, polyp-like Hepatocytes and fibrous connective tissue stroma, blood vessels and nerves</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Vaideswar et al.</td>
<td>ND</td>
<td>39</td>
<td>2460</td>
<td>Female</td>
<td>32 seek US: UC</td>
<td>ND</td>
<td>Very small</td>
<td>Pale brown</td>
<td>Soft Hepatic cords without bile ducts</td>
<td>ND</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Go and Cho 38</td>
<td>ND</td>
<td>28</td>
<td>3000</td>
<td>Male</td>
<td>No</td>
<td>ND</td>
<td>ND</td>
<td>Brown</td>
<td>ND</td>
<td>Firm Hepatic tissue surrounded by a small cyst-like structure corresponding to gallbladder wall tissue</td>
<td>ND</td>
<td></td>
</tr>
<tr>
<td>Lee et al. 38</td>
<td>ND</td>
<td>38</td>
<td>3290</td>
<td>Female</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>Purplish brown</td>
<td>ND</td>
<td>Firm, polyp-like Hepatocytes and fibrous connective tissue stroma, blood vessels and nerves</td>
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<td></td>
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<tr>
<td>Horn et al. 33</td>
<td>Vaginal</td>
<td>39</td>
<td>40</td>
<td>Male</td>
<td>No</td>
<td>Central at the navel</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>Firm Hepatic tissue surrounded by a small cyst-like structure corresponding to gallbladder wall tissue</td>
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<td></td>
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<tr>
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<td></td>
<td></td>
<td>3290</td>
<td>Female</td>
<td>28 week US: UC</td>
<td>16 mm</td>
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<td>ND</td>
<td>ND</td>
<td>Firm Hepatic tissue surrounded by a small cyst-like structure corresponding to gallbladder wall tissue</td>
<td>ND</td>
<td></td>
</tr>
</tbody>
</table>

**Table 1** Literature review of published hepatic choristoma cases.

HMD, hyaline membrane disease; ND, not described; RDS, respiratory distress syndrome; RV, right ventricle; UC, umbilical cord; US, ultrasound.
sinusoids and their subsequent migration to more distant regions, such as the umbilical cord, while the connection with the main liver may be maintained through the umbilical vein.

The differential diagnosis of umbilical cord masses is complex and must include cyst and pseudocyst, haematomatoma, umbilical artery aneurysm, haemangioma, teratoma, angiomyxoma, patent urachus, ectopic liver, as well as the most common diseases of the umbilical cord, which are umbilical cord hernia, gastrochisis and omphalocele.

Ectopic liver in the newborn is usually diagnosed by chance following imaging tests or surgical procedures performed for unrelated reasons. However, it may be diagnosed due to complications like torsion, which manifests with abdominal pain, gastric outlet obstruction and respiratory distress syndrome, caused by the presence of hepatic tissue in supradiaphragmatic locations.

Only eight other cases of hepatic tissue in the umbilical cord have been described in the literature (1–4) (Table 1), and the diagnosis of the umbilical cord mass was made postnatally in three of the nine cases, with the definitive diagnosis being made by anatomical pathology. On rare occasions it can be accompanied by symptoms of infection and be associated with other abnormalities, such as urothelial or biliary atresia, ectopic pancreas and heart and lung malformations. In our case, as happened in the one described by Horn et al. (5), we observed an intraperitoneal connection with the liver that may correspond to the round ligament, a vestige of the left umbilical vein.

To conclude, we would like to highlight that when ultrasound examination reveals a mass in the umbilical cord we should consider the possibility of rare conditions, like the one described here, along with more common diseases.

Doppler ultrasound of the mass can be helpful to this end, although as we mentioned above, in most cases the definitive diagnosis will be made postnatally. At any rate, the histological characteristics of the lesion should not change the obstetric approach in the absence of intestinal or vascular involvement in the foetus, and the surgical approach will depend on the suspected diagnosis after birth.

References


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Bone marrow toxicity secondary to a primary Epstein–Barr infection in a patient with Crohn's disease on thiopurines treatment

Toxicidad medular secundaria a la primoinfección por virus de Epstein-Barr en paciente con enfermedad de Crohn en tratamiento con tiopurínicos

Dear Editor:

The efficacy of thiopurine immunosuppressants in the treatment of inflammatory bowel disease (IBD) has been demonstrated, and thiopurines are the most commonly used drugs to maintain remission induced by exclusive enteral nutrition or steroids in paediatric patients with Crohn’s (EC) disease. Their long-term use may facilitate the development of opportunistic infections by viruses such as Epstein–Barr virus (EBV). Thiopurine blocking of regulatory T cells enhances the cytotoxicity of EBV, leading to B-cell lymphoproliferation. In immunosuppressed patients, the manifestation of EBV may range from an infectious mononucleosis to a haemophagocytic lymphohistiocytosis (HLH). 7

We present the case of a 14-year-old male patient with CD in clinical and laboratory remission following combined treatment with infliximab (IFX) and azathioprine (AZA) since diagnosis. In order to reduce the risk associated with dual immunosuppression, IFX was discontinued 10 months after initiating treatment, and the patient developed a high fever, odynophagia, submandibular lymphadenopathy and splenomegaly. Laboratory analysis revealed pancytopenia and elevated levels of transaminases, triglycerides and ferritin (Table 1). Intravenous empirical antibiotic therapy was initiated due to the presence of febrile neutropenia (500 cells/mm3) and was suspended after 72 h following a negative blood culture and a positive Paul-Bunnell test. Epstein–Barr virus was detected by polymerase chain reac-

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