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.¹

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.

<table>
<thead>
<tr>
<th>Maternal age</th>
<th>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>19</td>
<td>Vaginal</td>
<td>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 × 3 mm</td>
<td>Red-purple</td>
<td>ND</td>
<td>Normal hepatic tissue</td>
<td>Urachus cyst with immature hepatic tissue</td>
<td>Uncertain</td>
</tr>
<tr>
<td>28</td>
<td>Vaginal</td>
<td>Yes</td>
<td>40</td>
<td>3240</td>
<td>Female</td>
<td>ND</td>
<td>CU insertion site</td>
<td>2.5 cm × 3.5 cm</td>
<td>Red</td>
<td>Firm</td>
<td>Firm, rubbery</td>
<td>Hepatic tissue with numerous portal areas</td>
<td>No</td>
</tr>
<tr>
<td>ND</td>
<td>Caesarean</td>
<td>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 mm</td>
<td>Dark red</td>
<td>Normal hepatic tissue</td>
<td>Firm, polyp-like Hepatocytes and fibrous connective tissue stroma, blood vessels and nerves</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>ND</td>
<td>Caesarean</td>
<td>RDS</td>
<td>38</td>
<td>3314</td>
<td>Female</td>
<td>19 week US: UC mass, 6.3 mm × 3.5 mm × 7 mm, no blood flow</td>
<td>Proximal to UC and navel</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>ND</td>
<td>Vaginal</td>
<td>Yes</td>
<td>39</td>
<td>2460</td>
<td>Female</td>
<td>19 week US: UC mass, 6.3 mm × 3.5 mm × 7 mm, no blood flow</td>
<td>Proximal to UC and navel</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>ND</td>
<td>Death</td>
<td>ND</td>
<td>28</td>
<td>3000</td>
<td>Male</td>
<td>No</td>
<td>Proximal to UC and navel</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>38</td>
<td>3290</td>
<td>Female</td>
<td>28 week US: UC mass, 28 mm × 17 mm, UC diameter 16 mm</td>
<td>Central at the navel</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>ND</td>
<td>Vaginal</td>
<td>Yes</td>
<td>39</td>
<td>40</td>
<td>Female</td>
<td>28 week US: UC mass, 28 mm × 17 mm, UC diameter 16 mm</td>
<td>Central at the navel</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
</tr>
</tbody>
</table>

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, haematoma, umbilical artery aneurysm, haemangioma, teratoma, angiomoyxoma, 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 literature3-6 (Table 1), and the diagnosis of the umbilical cord mass was made prenatally 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 utrachal cysts, umbilical hernia, and portal hypertension. In our case, as happened in the one described by Horn et al.,7 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

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

Toxicidad medular secundaria a 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).1

We present the case of a 14-year-old male patient with Crohn’s disease on thiopurines, with the diagnosis of EBV, and the development of HLH. The patient was started on IFX and azathioprine (AZA) due to the presence of febrile neutropenia (500 cells/mm³) and was suspened after 72 h following a negative blood culture and a positive Paul–Bunnell test. Epstein–Barr virus was detected by polymerase chain reac-

---

134 SCIENTIFIC LETTERS

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

Toxicidad medular secundaria a 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).1

We present the case of a 14-year-old male patient with Crohn’s disease on thiopurines, with the diagnosis of EBV, and the development of HLH. The patient was started on IFX and azathioprine (AZA) 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-

---

Please cite this article as: Ruiz Hernández C, Sánchez Hernández D, Vila Miravet V, Pinillos Písón S, Martín de Carpi J. Toxicidad medular secundaria a primoinfección por virus de Epstein-Barr en paciente con enfermedad de Crohn en tratamiento con tiopurínicos. An Pediatr (Barc). 2015;83:134-135.