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.


S. Camacho-Lovillo*, A.C. García-Martínez

Sección de Inmunopatología y Enfermedades Infecciosas, Unidad de Pediatría, Hospital Universitario Virgen del Rocío, Sevilla, Spain

*Corresponding author. E-mail address: marisolcl73@gmail.com (S. Camacho-Lovillo).

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

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| Maternal age | 19 Vaginal | 29 Vaginal | ND Caesarean | 28 Caesarean | ND Vaginal | ND Vaginal | ND Vaginal | 33 Vaginal |
| Delivery | Yes | Yes | RDS | RDS | Yes | RDS | Yes |
| manifestations | | | | | | | | | |
| Gestational age | 28 | 40 | 35 | 38 | 39 | 28 | 38 | 39 | 40 |
| Weight (g) | 1247 | 3240 | 2180 | 3314 | 2460 | ND | 3000 | ND | 3290 |
| Sex | Male | Female | Female | Female | Female | Male | Male | Female | Male |
| Prenatal diagnosis | ND | ND | No | 19 week US: UC mass, 6.3 mm × 3.5 mm × 7 mm, no blood flow | US: hyperechoic mass on base of UC, 2 cm × 2 cm | No | No | 28 week US: UC mass, 28 mm × 17 mm, UC diameter 16 mm | ND |
| Suggestive | Yes | Yes | RDS | RDS | No | No | No | Yes | |
| manifestations | | | | | | | | | |
| Location | Attached to navel by a stalk 2 | CU insertion site | 1.5 cm from the end of the UC | Proximal to UC and navel | 2.8 cm from UC insertion | Central at the navel | ND | |
| Size | 7.5 cm × 6 cm × 3 cm | Yellow-green, brown interior | 2.5 cm × 3.5 mm | Firm, rubbery | Firm, normal hepatic tissue | ND | ND | 4.5 cm × 2 cm × 1.8 cm | |
| Colour | Red-purple | Dark red | Firm | ND | Firm | Firm, polyp-like | ND | Wine red |
| Consistency | ND | ND | ND | ND | ND | ND | ND | |
| Pathological anatomy | Biliary atresia + ectopic pancreas in the jejunum | Atretic segment of the urachus | Urachus cyst with immature hepatic tissue | Hepatic tissue with numerous portal areas | Hepatic cords without bile ducts | Firm, polyp-like Hepatocytes and fibrous connective tissue stroma, blood vessels and nerves | ND | |
| Peritoneal communication | Uncertain | No | No | No | No | No | Yes | Yes, with the round ligament of the liver |
| Associated anomalies | ND | Biliary atresia + ectopic pancreas in the jejunum | No | | | | No | No |

HMD, hyaline membrane disease; ND, not described; RDS, respiratory distress syndrome; RV, right ventricle; UC, umbilical cord; US, ultrasound.
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 tiopurinicos

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).

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 pancytopaenia and elevated levels of transaminases, triglycerides and ferritin (Table 1). Intravenous empirical antibiotic therapy was initiated due to the presence of febrile neutropaenia (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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Y. González Ruiz a,b, L. Cotaina Gracia a, M. Ruiz de Temiño a,c, A. Joana González Esqueda a, M.R. Delgado Alvira a

a Servicio de Cirugía Pediátrica, Hospital Universitario Miguel Servet, Zaragoza, Spain
b Servicio de Ginecología y Obstetricia, Hospital Universitario Miguel Servet, Zaragoza, Spain
c Corresponding author.
E-mail address: yuremagr@gmail.com (Y. González Ruiz).

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b Servicio de Ginecología y Obstetricia, Hospital Universitario Miguel Servet, Zaragoza, Spain
c Corresponding author.
E-mail address: yuremagr@gmail.com (Y. González Ruiz).