fever by the Paediatric Infectology Society and the Paediatric Rheumatology Society. An Pediatr (Barc). 2011;74:194.e1–16.
2. Uthman I, Hajj-Ali RA, Arayssi T, Masri AF, Nasr F. Arthritis in familial Mediterranean fever. Rheumatol Int. 2001;20:145–8.
3. Conca W, Ghatasheh G, Al-Salam S, Neidl Van Gorkom K. The medical odyssey of a boy with arthritis of familial Mediterranean fever. Int J Rheum Dis. 2012;15:37–9.
4. Lidar M, Kedem R, Mor A, Levartovsky D, Langevitz P, Livneh A. Arthritis as the sole episodic manifestation of familial Mediterranean fever. J Rheumatol. 2005;32:859–62.
5. Majeed HA, Rawashdeh M. The clinical patterns of arthritis in children with familial Mediterranean fever. QJM. 1997;90:37–43.
6. Jarjour RA, Dodaki R. Arthritis patterns in familial Mediterranean fever patients and association with M694V mutation. Mol Biol Rep. 2011;38:2033–6.

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).

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

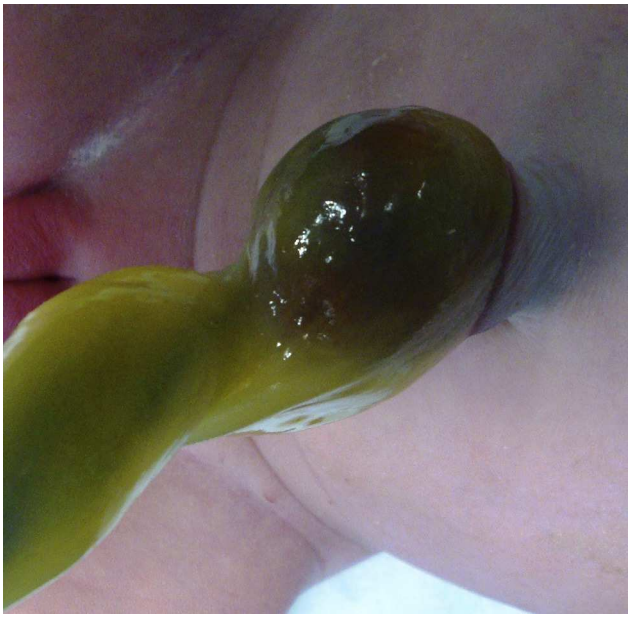

Figure 1 Transillumination of the wine-red mass in the umbilical cord.

☆ Please cite this article as: González Ruiz Y, Cotaina Gracia L, Ruiz de Temiño M, González Esgueda AJ, Delgado Alvira MR. Revisión de casos publicados de coristoma hepático. Diagnóstico diferencial de masas de cordón umbilical. An Pediatr (Barc). 2015;83:132–134.

Table 1 Literature review of published hepatic choristoma cases.

	Shaw and Pierog	Park et al.	Preminger et al.[3]	Wax et al.[4]	Vaideeswar et al.[5]	Go and Cho	Lee et al.[6]	Horn et al.[2]	Our case
Maternal age	19	29	ND	28	ND	ND	ND	ND	33
Delivery	Vaginal	Vaginal	Caesarean	Caesarean	Vaginal	Death	ND	Vaginal	Vaginal
Suggestive manifestations	Yes	Yes	RDS	RDS	Yes	–	ND	Yes	Yes
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	Female
Prenatal diagnosis	ND	ND	No	19 week US: UC mass, 6.3 mm × 3.5 mm × 7 mm, no blood flow	32 seek US: hyperechoic mass on base of UC, 2 cm × 2 cm	No	No	No	28 week US: UC mass, de 28 mm × 17 mm, UC diameter 16 mm
Location	Attached to navel by a stalk	CU insertion site	1.5 cm from the end of the UC	Proximal to UC and navel	2.8 cm from UC insertion	ND	Central at the navel	ND	1 cm from navel, proximal to UC
Size	2	7.5 cm × 6 cm × 3 cm	3 mm × 3 mm	2.5 cm × 3.5 cm	Very small	ND	1.8 cm × 1.2 cm × 1.2 cm	ND	4.5 cm × 2 cm × 1.8 cm
Colour	Red-purple	Yellow-green, brown interior	Red	Dark red	Pale brown	Brown	Yellowish grey	Purplish brown	Wine red
Consistency	ND	Firm, rubbery	Firm	ND	Firm	Soft	Firm, polyp-like	ND	Firm
Pathological anatomy	ND	ND	Normal hepatic tissue	Urachus cyst with immature hepatic tissue	Hepatic tissue with numerous portal areas	Hepatic cords without bile ducts	Hepatocytes and fibrous connective tissue stroma, blood vessels and nerves	ND	Hepatic tissue surrounded a small cyst-like structure corresponding to gallbladder wall tissue
Peritoneal communication	Uncertain	No	No	No	No	ND	No	Yes	Yes, with the round ligament of the liver
Associated anomalies	ND	Biliary atresia + ectopic pancreas in the jejunum	No	Atretic segment of the urachus	Double outlet RV + right lung agenesis + HMD in left lung	ND	Absence of fourth toe in left foot	No	No

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, angiomyomyxoma, patent urachus, ectopic liver, as well as the most common diseases of the umbilical cord, which are umbilical cord hernia, gastroschisis and omphalocoele.

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[2–6] (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 or biliary atresia, ectopic pancreas and heart and lung malformations. In our case, as happened in the one described by Horn et al.,[2] 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

1. Collan Y, Hakkiluoto A, Hästbacka J. Ectopic liver. Ann Chir Gynaecol. 1978;67:27–9.
2. Horn F, Kubikova E, Dubravova D, Babal P, El Falougy H, Selmeciova P, et al. Omphalocele with ectopic liver forming a mesodermal cyst. Bratisl Lek Listy. 2013;114:100–3.
3. Preminger A, Udassin R, Pappo O, Arad I. Ectopic liver tissue within the umbilical cord. J Pediatr Surg. 2001;36:1085–6.
4. Wax JR, Pinette MG, Cartin A, Blackstone J. Ectopic liver: a unique prenatally diagnosed solid umbilical cord mass. J Ultrasound Med. 2007;26:377–9.
5. Vaideeswar P, Yewatkar D, Nanavati R, Bhuiyan P. Ectopic liver tissue in umbilical cord. J Postgrad Med. 2011;57:229–30.
6. Lee SY, Nam CW, Suh JH. Heterotopic liver in the umbilicus: a case report. Basic Appl Pathol. 2011;4:30–2.

Y. González Ruiz[a,*], L. Cotaina Gracia[b], M. Ruiz de Temiño[a], A. Joana González Esgueda[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*

* Corresponding author.
E-mail address: yuremagr@gmail.com (Y. González Ruiz).

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 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/mm^3) 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 Pisón S, Martin 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.