Gastroenterol Hepatol. 2016;39(4):301–304

Gastroenterología y Hepatología

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LETTERS TO THE EDITOR

Cholestatic hepatitis in immunoglobulin-resistant Kawasaki disease☆

Hepatitis colestásica en la enfermedad de Kawasaki resistente a inmunoglobulinas

To the Editor,

Kawasaki disease (KD) is an acute vasculitis of unknown aetiology, and is the leading cause of acquired heart disease in children in developed countries. The highest incidence occurs in children aged between 6 months and 5 years (15.1/100,000 in Spain), although it can present at any age, even in adults.[1] Clinical symptoms can include cholestatic hepatitis and hepatosplenomegaly, parameters which, while they do not fall within classical diagnostic criteria, have been reported as predictors of poor response to immunoglobulins, as in our case.

A 6-year-old schoolboy was admitted from the emergency department for a 3-day history of high fever associated with a 4 cm × 5 cm right lateral-cervical lymphadenopathy, which did not improve after prescribing the appropriate doses of oral amoxicillin–clavulanic acid. Laboratory tests revealed C-reactive protein (CRP) 259 mg/L (normal CRP < 5 mg/L) and activated lymphocytes, for which he was prescribed intravenous cefotaxime; virus serology was requested. On the third day of admission (day 5 of fever) the patient deteriorated, with conjunctival injection, cracked lips, generalised exanthema, swollen hands, hepatosplenomegaly and jaundice of the skin and mucous membranes. Blood tests found glutamic–oxaloacetic transaminase (GOT): 79 U/L; glutamic–pyruvic transaminase (GPT): 192 U/L; gamma-glutamyl transferase (GGT): 241 U/L; alkaline phosphatase (ALP): 354 U/L; total bilirubin: 5.75 mg/dL; direct bilirubin: 5.07 mg/dL; albumin: 2.6 g/dL; N-terminal pro b-type natriuretic peptide (NT-pro-BNP): 14,700 pg/mL (normal <150 pg/ml); coagulation: normal; and virus serology (Epstein–Barr virus, cytomegalovirus, toxoplasma, hepatitis A, B and C, herpes simplex): negative. Abdominal ultrasound showed increased periportal echogenicity and intrahepatic bile duct dilatation. The presence of a moderate amount of free fluid was observed in the perihepatic space and pelvic floor (Fig. 1, showing the double-duct sign, characteristic of intrahepatic bile duct dilatation). Suspecting Kawasaki disease, treatment was initiated with 2 g/kg immunoglobulins and 80 mg/kg acetylsalicylic acid, but the patient remained febrile, so a second dose of gammaglobulin was administered at 72 h. Due to persistence of the fever, he required 30–15 mg/kg megadose methylprednisolone for 2 days, with an excellent response: the fever and hepatosplenomegaly resolved, and the cholestasis and CRP and cardiac inflammatory parameters (pro-BNP) improved. To complete the clinical picture of KD, on day 12 of admission the patient presented desquamation of the fingertips and reactive thrombocytosis (18,000,000), while cardiac ultrasound showed ectasia of the coronary arteries with a 4.2 mm aneurysm in the anterior descending artery. He remained in hospital for 15 days; laboratory tests 2 weeks after discharge were normal, as was the liver ultrasound.

Hepatomegaly is described in approximately 14% of patients with KD, according to the series[2]; of these, 20–30% will present hepatic dysfunction.[3,4] The aetiology of hepatic dysfunction in KD is unknown,[2] although some authors hypothesise that this liver damage could be caused by an increase in natural killer cells in the endothelium and hepatic sinusoids.[5] There is little information on liver histology in these patients, as hepatic dysfunction is rare, usually

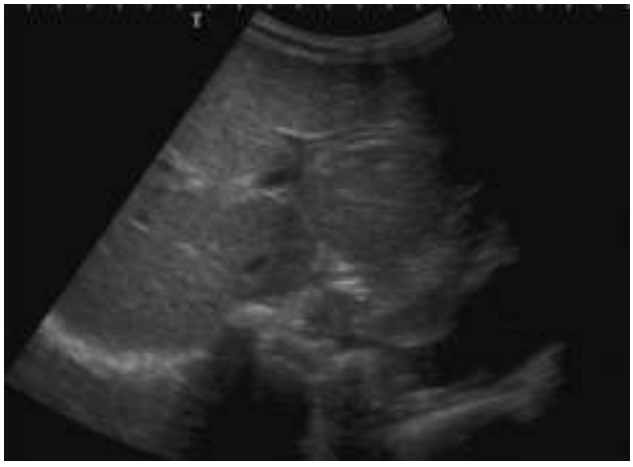

Figure 1 Double-duct sign, characteristic of intrahepatic biliary dilatation.

☆ Please cite this article as: Vázquez Gomis R, Izquierdo Fos I, López Yañez A, Mendoza Durán M, Serrano Robles MI, Vázquez Gomis C, et al. Hepatitis colestásica en la enfermedad de Kawasaki resistente a inmunoglobulinas. Gastroenterol Hepatol. 2016;39:301–302.

mild, and reverts with treatment, so liver biopsies are not routinely performed. The finding of vasculitis in liver tissue has been described in autopsies (in patients who died from this disease),[6] and a case has been reported in which a liver biopsy revealed periportal inflammation with increased polymorphonuclear cells.[7] With proper treatment, the liver symptoms return to normal. Treatment of KD is 2 g/kg non-specific gammaglobulin, with the possibility of administering a second dose after 24–72 h if the fever persists. About 15% of patients do not respond to this treatment,[8,9] so intravenous corticosteroids and even infliximab are necessary in non-responders. The presence of hepatic dysfunction is associated with a poor response to gammaglobulin and the appearance of coronary aneurysms,[3–5,10] as occurred in our patient. For this reason, the latest clinical guidelines published in Japan and the United Kingdom recommend the use of corticosteroids concomitantly with the gammaglobulin as a first option in these cases,[11] in order to avoid the appearance of coronary aneurysms, which is the major morbidity. It is important to know that KD can be the cause of febrile hepatic dysfunction of unclear aetiology, and is a prognostic factor of a poor responder to gammaglobulins.

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Sister Mary Joseph's nodule: Umbilical metastasis from gallbladder cancer☆

Nódulo de la hermana María José: metástasis umbilical de cáncer de vesícula

Dear Editor,

Sister Mary Joseph nodule (SMJN) is an umbilical metastasis of any primary tumour.[1–4] It can be the first sign of an undiagnosed neoplasm, or can appear as a recurrence or progression of an already known tumour.[1,5] We present a new case caused by a gallbladder cancer, and review the topic.

A 76-year-old man presented with a 3-month history of right upper quadrant pain and dyspepsia, accompanied by acholia, choluria and pruritus in the previous 10 days. He reported that he noticed umbilical discomfort that he related with a previous herniorrhaphy. His medical history included atrial fibrillation and umbilical herniorrhaphy. Physical examination found right upper quadrant pain, a petrous tumour in the umbilical region and jaundice. Blood tests revealed gamma glutamyl transferase (GGT): 718 IU/L, alkaline phosphatase (ALP): 391 IU/L, total bilirubin: 9.1; carcinoembryonic antigen (CEA): 92.7 mg/dL and CA19.9: 206 IU/L. Abdominal ultrasound and computed tomography (CT) scans showed a large heterogeneous solid lesion with poorly defined borders in the perihilar region, emerging from

DOI of refers to article:
http://dx.doi.org/10.1016/j.gastrohep.2015.05.010

☆ Please cite this article as: Ramia JM, de la Plaza R, López-Marcano A, Kuhnhardt A, David Gonzales J. Nódulo de la hermana María José: metástasis umbilical de cáncer de vesícula. Gastroenterol Hepatol. 2016;39:302–304.