Contrast-enhanced magnetic resonance cholangiography with gadoxetic-acid-disodium for the detection of biliary-cyst communication in Caroli disease

Contraste colangiografía RM con gadoxetic-ácido disódico para la detección de la comunicación biliares-quistes en la enfermedad de Caroli

A 49-year-old woman presented with longstanding, intermittent episodes of mild abdominal pain in the right upper quadrant. Clinical examination revealed a mild hepatomegaly, without splenomegaly or signs of portal hypertension. An abdominal ultrasound performed in the diagnostic work-up identified multiple liver cysts (Fig. 1A). She was further referred to our institution for a magnetic resonance (MR) examination, which confirmed the findings of multiple intrahepatic cysts in close relation to the biliary tree, some of them with the central dot sign, which represents a central fibrovascular bundle within the lesion (Fig. 1B,C). Contrast-enhanced MR cholangiography using hepatospecific contrast agent (gadoxetic-acid-disodium, Gd-EOB-DTPA), in the late hepatobiliary phase (biliary excretion of contrast agent, 2 h after contrast agent injection), showed filling of the cystic spaces by the contrast medium (Fig. 1D), allowing to prove the communication between these cysts and the intrahepatic biliary tree and, consequently, a confident diagnosis of Caroli disease (CD).

Caroli disease is a rare congenital disorder characterized by non-obstructive, saccular or fusiform dilatation (focal or diffuse) of the intrahepatic biliary ducts.[1] The conventional MR cholangiography findings of multiple intrahepatic cysts in close relation to the biliary tree in diffuse CD, as in our case, have to be differentiated from autosomal-dominant polycystic liver disease and peribiliary cysts, among other alternative diagnoses.[1] Although the central dot sign is another clue for the diagnosis of CD on conventional MR cholangiography,[1] it has also been described in other diseases.[3] In the last years, the use of contrast-enhanced MR cholangiography with Gd-EOB-DTPA has suggested to be effective in identifying such communications, ultimately allowing a confident, non-invasive diagnosis of CD.[1] Nonetheless, the use of this technique in the diagnosis of CD has been very rarely reported in the literature.[1,2] Although in our case late hepatobiliary phase images obtained 2 h after contrast medium injection were sufficient, delayed images obtained after 24 h[1] may be necessary to demonstrate biliary-cysts communication in some cases of slow-filling cavities.

With this report we aim at emphasizing the role of modern hepatobiliary MR imaging with gadolinium based contrast agents as a noninvasive tool of demonstrating

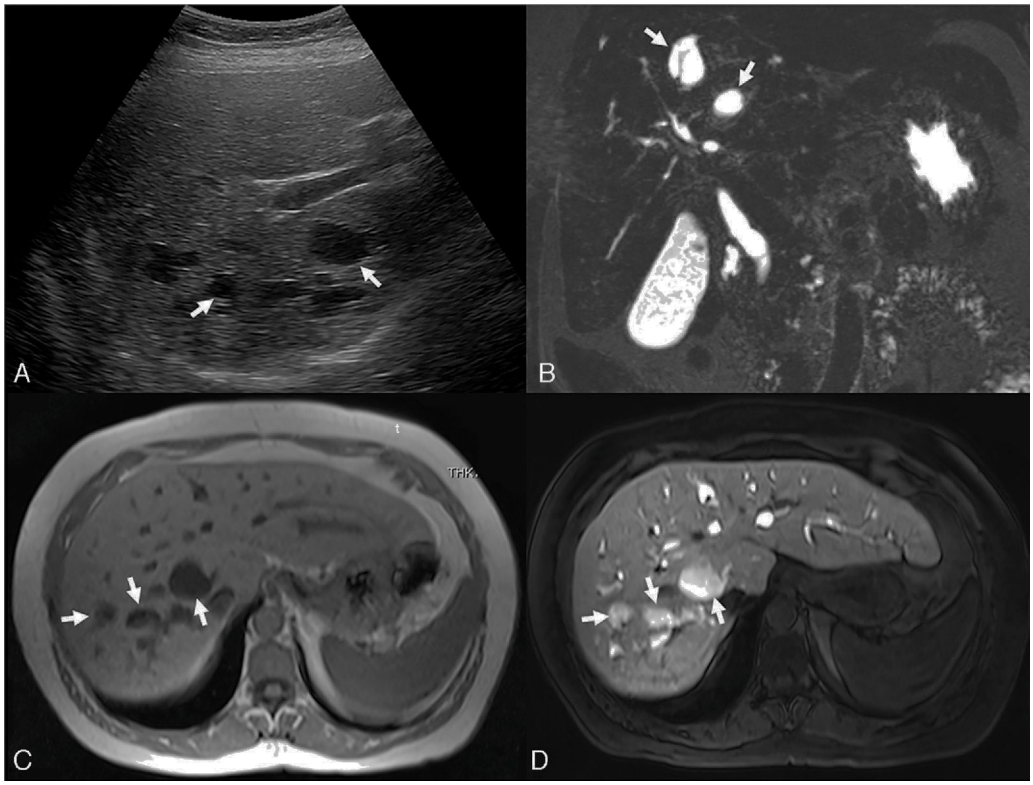

Figure 1 Ultrasound image (A) demonstrates multiple intrahepatic cysts (arrows). Coronal conventional T2-weighted MR cholangiography (B) and axial pre-contrast T1-weighted MR image (C) confirm the finding of multiple liver cysts in close relation to the biliary tree (arrows), some of them with the central dot sign (B). Late hepatobiliary phase of contrast-enhanced MR cholangiography using gadoxetic acid (D) demonstrates filling of cysts by contrast agent (arrows), proving their communication with the biliary tree and adding complementary information supportive of a diagnosis of Caroli disease.

biliary-cysts communication, adding complementary diagnostic information to conventional MR sequences and allowing a conclusive diagnosis of CD in the appropriate clinical setting, without the use of invasive methods (e.g., endoscopic retrograde cholangiography and percutaneous transhepatic cholangiography), which may be associated with complications such as bleeding, infection and pancreatitis.[4]

Financial support

None.

Conflicts of interest

The authors declare no conflict of interest.

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2444-3824/

Poor correlation between acute phase reactants and intestinal involvement in patients with onset of Crohn's disease under treatment with an interleukin-6 inhibitor due to seronegative arthropathy☆

Mala correlación entre reactantes de fase aguda y afectación intestinal en paciente con comienzo de la enfermedad de Crohn bajo tratamiento con inhibidor de IL-6 por artropatía seronegativa

This case concerned a 44-year-old woman, with no drug allergies or history of tobacco, alcohol or illicit drug use, with seronegative arthritis that commenced at age 18 in the form of asymmetric oligoarthritis, predominantly in the large joints. At age 22, she began to suffer rapidly destructive right coxitis, which required right hip replacement (RHR) at age 42. Rheumatoid factor, anti-citrullinated peptide antibodies, antinuclear antibodies and HLA-B27 tests were negative. She continued with remission induction therapy with hydroxychloroquine, gold salts (discontinued due to inefficacy) and methotrexate (MTX) (discontinued due to intolerance). In 2006, she initiated treatment with adalimumab (discontinued at 3 months due to skin rash) and then with etanercept (2008–2009), with partial response, and required glucocorticoids during the arthritis flares. She then received rituximab every 6 months (2009–2011, interrupted for the RHR surgery). She required infiltration of the right carpus for a new arthritis flare, and recommenced rituximab in June 2014, with a partial response.

In July 2014, the biological treatment was switched after the patient began to lose weight and became anaemic, with deterioration in her general health and arthritis, and elevated acute phase reactants (erythrocyte sedimentation rate [ESR] 64 mm/h and C-reactive protein [CRP] 56 mg/L (normal value: <5 mg/L). As a result of the marked systemic component, intravenous tocilizumab (TCZ) monthly monotherapy (due to intolerance to MTX) was initiated in October 2014.

In January 2015, she was admitted to the gastroenterology department for loose stools (5–7 day) with no blood, pus or mucus, abdominal pain and 10-kg weight loss, which had commenced 1 month previously. Physical examination was remarkable for abdominal pain predominantly in the left flank, with no palpable masses or visceromegaly. Laboratory tests showed neither leukocytosis nor anaemia (haemoglobin 14.2 g/dL), with CRP 0.24 mg/L and albumin 44 g/L. To complete the study, she underwent fibrogastroscopy, which was normal, and colonoscopy, which showed extensive, deep ulcers with healthy interlesional mucosa extending from 12 cm from the anal margin to the caecum, as well as the terminal ileum; 15 cm were affected with minute aphthae (Fig. 1). Histopathology findings were consistent with inflammatory bowel disease (IBD). She later presented high faecal calprotectin (FC) levels of 2714 mg/kg faeces (normal value <100 mg/kg). This was diagnosed as a severe flare-up of ileocolic Crohn disease (CD), with associated

☆ Please cite this article as: Zacarías Martínez L, Mateo Soria L, Mañosa Círia M, Clos Parals A, Cabré Gelada E, Domènech Morral E. Mala correlación entre reactantes de fase aguda y afectación intestinal en paciente con comienzo de la enfermedad de Crohn bajo tratamiento con inhibidor de IL-6 por artropatía seronegativa. Gastroenterol Hepatol. 2016;39:670–672.