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.

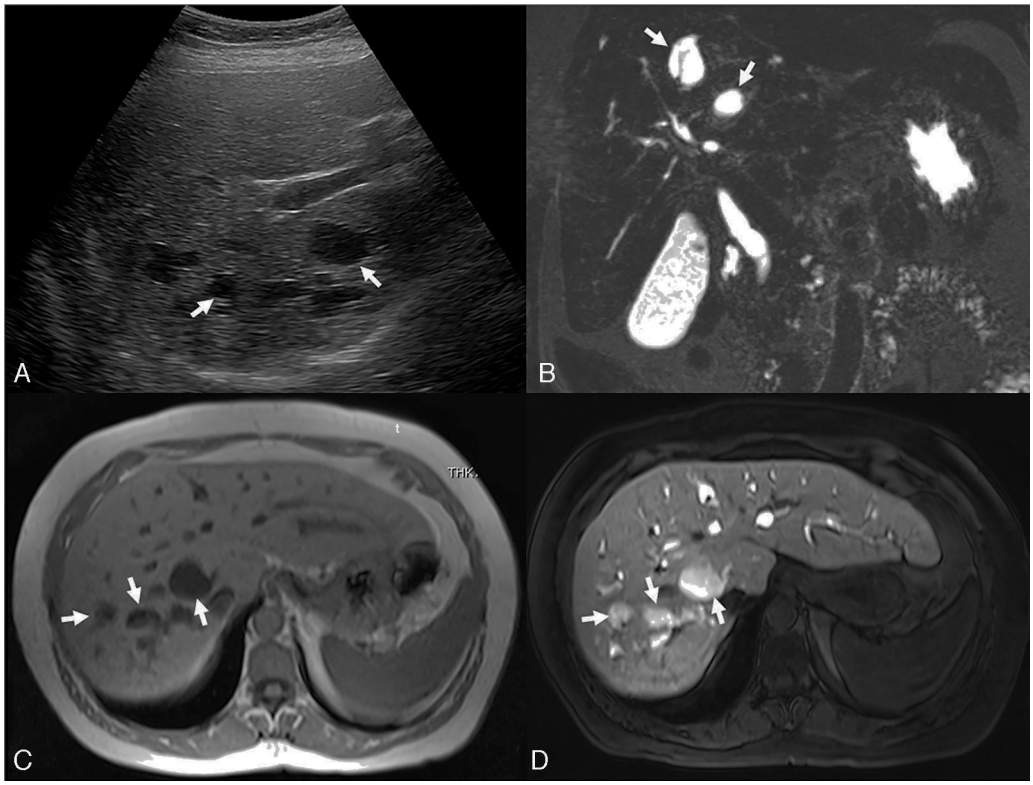

Figure 1 Ultrasound image (A) demonstrates multiple intrahepatic cysts (arrows). Coronal conventional T2-weighted MR cholangiography (B) and axial pre-contrast T1-weighted MR image (C) confirm the finding of multiple liver cysts in close relation to the biliary tree (arrows), some of them with the central 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.

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 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]

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Conflicts of interest

The authors declare no conflict of interest.

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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

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

Mujer de 44 años, sin alergias medicamentosas ni hábitos tóxicos, con artritis seronegativa de inicio a los 18 años, en forma de oligoartritis asimétrica, de predominio en grandes articulaciones. A los 22 años inició coxitis derecha rápidamente destructiva, que precisó prótesis de cadera derecha (PTC) a los 42 años. Los estudios de factor reumatoide, anticuerpos antipéptido citrulinados, anticuerpos antinucleares y HLA-B27 fueron negativos. Continuó con tratamiento inductor de remisión con hidroxicloroquina, sales de oro (supendidos por ineficacia) y metotrexate (suspendido por intolerancia). En 2006 se inició tratamiento con adalimumab (suspendido a los 3 meses por *rash* cutáneo) y, posteriormente, con etanercept (2008-2009), con respuesta parcial y requerimiento de glucocorticoides en los brotes de artritis. Posteriormente recibió rituximab cada 6 meses (2009-2011, interrumpido para intervención de PTC). Por nuevo brote de artritis precisó infiltración del carpo derecho, y se reinició rituximab en junio del 2014 con repuesta parcial.

En julio de 2014 inicia pérdida ponderal, empeoramiento del estado general y de la artritis, anemización y elevación de reactantes de fase aguda VSG 64 mm/h y PCR 56 mg/l (valor normal: < 5 mg/l), por lo que se decidió cambio de tratamiento biológico. Por el marcado componente sistémico y la necesidad de utilizarlo en monoterapia (intolerancia a MTX), se decidió iniciar tratamiento con tocilizumab ev mensual, en octubre de 2014.

En enero de 2015, ingresa en el servicio de gastroenterología por cuadro de deposiciones diarreicas (5-7 día), sin productos patológicos, con dolor abdominal y pérdida de 10 kg de peso, de un mes de evolución. En la exploración física destacaba dolor abdominal de predominio en flanco izquierdo, sin masas ni visceromegalias palpables. Analíticamente sin leucocitosis ni anemia (hemoglobina 14,2 g/dl), PCR 0,24 mg/l, albumina 44 g/l. Para completar el estudio se realizó una fibrogastroscopia que resultó normal y una colonoscopia que mostró desde 12 cm del margen anal hasta ciego úlceras extensas y profundas con mucosa interlesional sana, íleon terminal, afecto 15 cm con aftas milimétricas (fig. 1). La anatomía patológica resultó compatible con EII. Posteriormente se cursan niveles de calprotectina fecal que resulta elevada 2.714 mg/kg heces (VN < 100 mg/kg). Se orientó como un brote grave de enfermedad de Crohn (EC) íleo-cólica con artropatía periférica asociada. Se inició tratamiento con corticoterapia a dosis plena (40 mg/día), azatioprina 150 mg/dia y nutrición enteral total (NET), presentando correcta evolución clínica, con disminución en el número de deposiciones, así como resolución del dolor abdominal y articular, siendo dada de alta a domicilio.