

Low Phospholipid Associated Cholelithiasis (LPAC) syndrome

Published on 18.06.2017

DOI: 10.1594/EURORAD/CASE.14750

ISSN: 1563-4086

Section: Abdominal imaging

Area of Interest: Abdomen

Procedure: Imaging sequences

Procedure: Localisation

Procedure: Education

Procedure: eLearning

Imaging Technique: MR

Imaging Technique: CT

Imaging Technique: Ultrasound

Special Focus: Dilatation Obstruction / Occlusion

Metabolic disorders Case Type: Clinical Cases

Authors: Dr Filippigh, Dre Varnay

Patient: 26 years, male

Clinical History:

26-year-old non obese male patient consulted the emergency unit with abdominal right upper quadrant abdominal pain.

Hepatic tests: acute hepatic cytolysis (AST 477 UI ; ALT 823 UI) and icteric cholestasis (ALP 175 UI ; GGT 411 UI ; bilirubin 55 $\mu\text{mol/L}$).

Imaging Findings:

The first US examination was made outside our institution, and we did not have access to the images. However, the radiologist reported a dilatation of the intra and extra-hepatic bile ducts without visualization of an obstacle.

The MRI was performed to find the cause of the obstruction. (Figs. 1 and 2)

It showed intra and extra-hepatic biliary dilatation (Fig. 1) and an isosignal content of the distal common biliary duct, cause of the obstruction (Figs. 1 and 2).

On the referral sheet of the second US examination, it was clearly stated that the clinician was looking for signs of LPAC syndrome.

The images produced were pathognomonic: multiple millimetric hyperechogenic images, sometimes associated with comet tail artefacts (Fig. 3).

Discussion:

In our case, the patient first consulted for upper quadrant abdominal pain and icteric cholestasis.

After clinical and biologic examinations, he first underwent a US examination. Then an MRI was performed to find the cause of the biliary obstruction.

The patient then underwent an ERCP (Endoscopic Retrograde Cholangio-Pancreatography) with a sphincterotomy that found sludge and micro-gallstones in the distal common bile duct.

He then had a second US examination that confirmed the final diagnosis.

In our case, the diagnosis was suspected because of symptomatic cholestasis before the age of 30 in a non-obese patient and confirmed by the finding of intra-hepatic millimetric hyperechogenicities with comet-tail artefacts.

The principal components of bile are: 1) phospholipids, 2) bile acids and 3) cholesterol. Most of gallstones appear in

the gallbladder when the balance between these three components is lost.

Usually, gallstones appear because of cholesterol excess.

In this case, there is a lack of phospholipids because of the mutation of the gene (ABCB4) coding for the membrane transporter MDR3 of phosphatidylcholine. The balance between the bile components is then broken and cholesterol precipitates in the bile ducts, provoking intra-hepatic micro-gallstones [2].

These intra-hepatic micro-gallstones, when they are obstructive, provoke upper quadrant abdominal pain and icteric cholestasis.

The pathognomonic US sign in the liver is the presence of small hyperechogenicities with comet tail artefacts [1].

These artefacts are a type of reverberation artefact (multiple echoes), with the two reflective interfaces and thus sequential echoes that are closely spaced. As a result, the artefact is triangular, tapered-shaped [3].

The clinical arguments that should evocate this diagnosis are 1) beginning of the symptoms before the age of 30, 2) relapse of the symptoms after cholecystectomy, 3) clinical history of cholestasis during pregnancy, 4) family history of a first degree relative with symptomatic cholelithiasis before the age of 30. This syndrome is also known for female predominance (role of oestrogens) [4].

The knowledge of this disease by radiologists can be very useful because the patient could benefit of a simple efficient medical treatment (ursodeoxycholic acid) [2].

Differential Diagnosis List: LPAC (Low Phospholipid Associated Cholelithiasis) syndrome, Common biliary colic, Cholecystitis, Cholangitis, Sclerosing cholangitis

Final Diagnosis: LPAC (Low Phospholipid Associated Cholelithiasis) syndrome

References:

Benzimra, Derhy, Rosmorduc, Menu, Poupon, Arrivé (2013) Hepatobiliary anomalies associated with ABCB4/MDR3 deficiency in adults : a pictorial essay. Springer (PMID: [23591976](#))

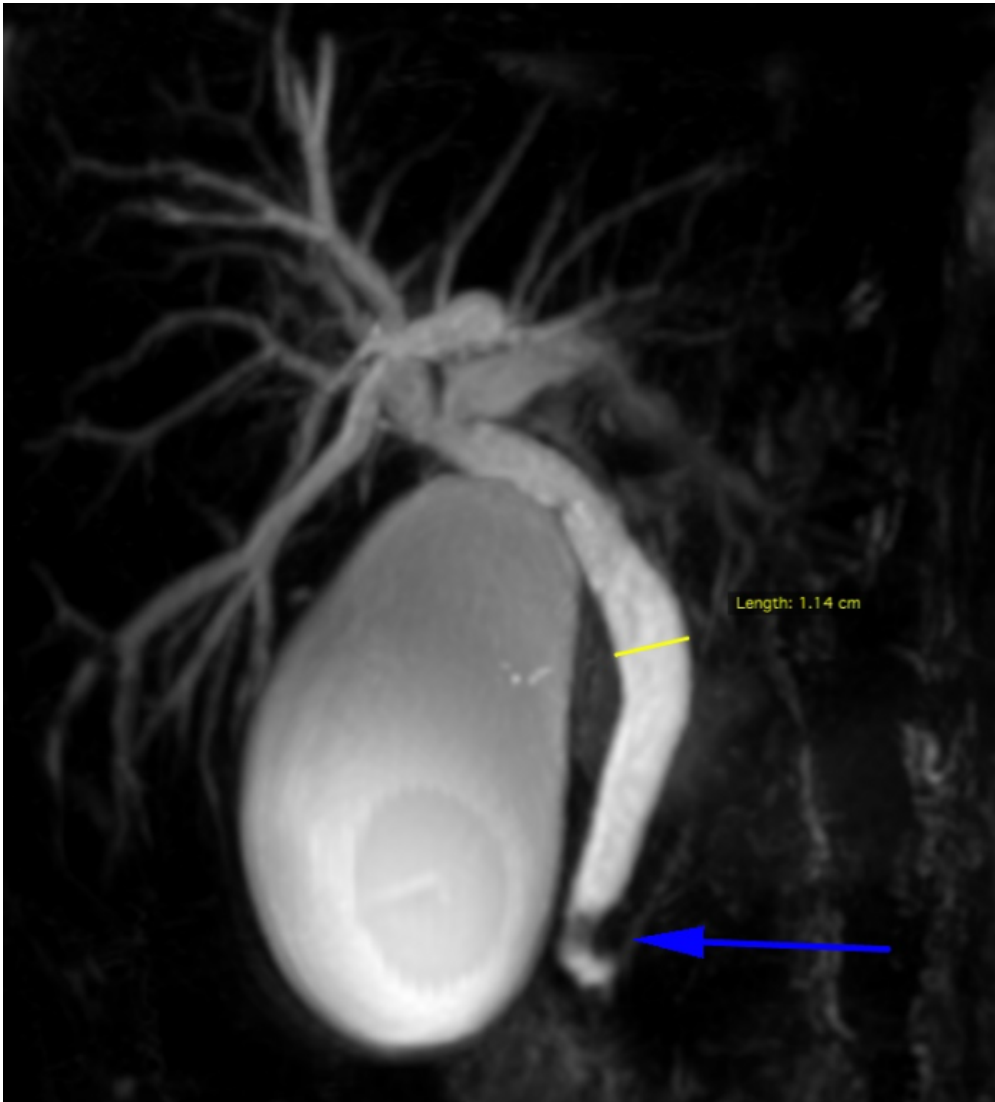
Bertrand Condat (2016) Le syndrome LPAC (Low Phospholipid Associated Cholelithiasis) : mythe ou réalité?. POST'U

Feldman, Katyal, Blackwood (2009) US artifacts. Radiographics (PMID: [19605664](#))

Condat B1, Zanditenas D, Barbu V, Hauuy MP, Parfait B, El Naggar A, Collot V, Bonnet J, Ngo Y, Maftouh A, Dugué L, Balian C, Charlier A, Blazquez M, Rosmorduc O. (2013) Prevalence of low phospholipid-associated cholelithiasis in young female patients. Elsevier (PMID: [23684896](#))

Figure 1

a

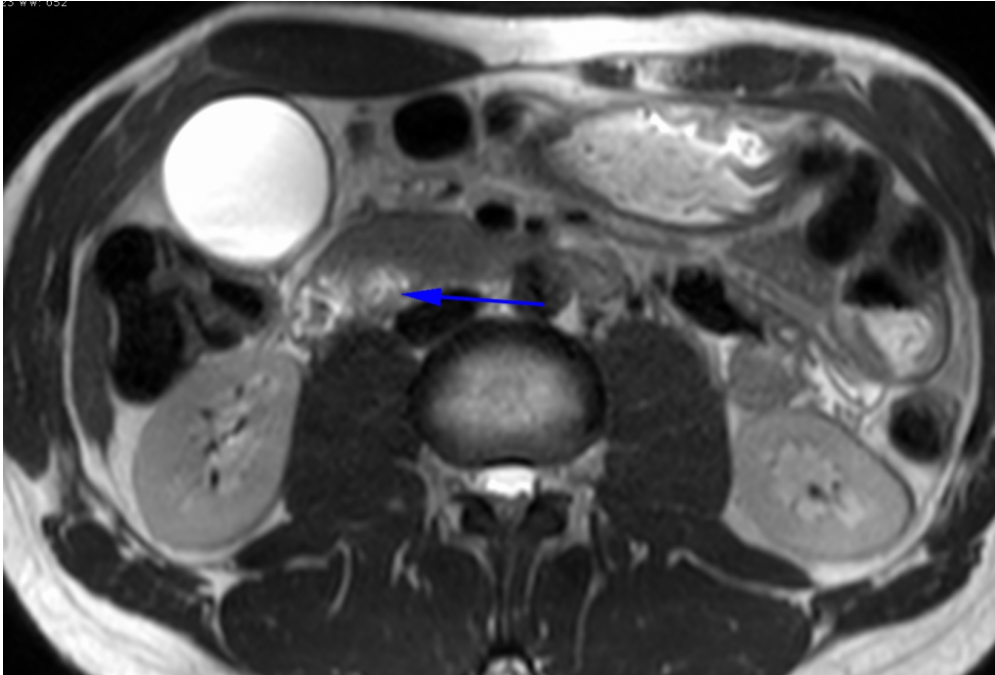


Description: Intra and extra-hepatic biliary dilatation.

The blue arrow shows a low signal content of the distal common bile duct. **Origin:** HUG

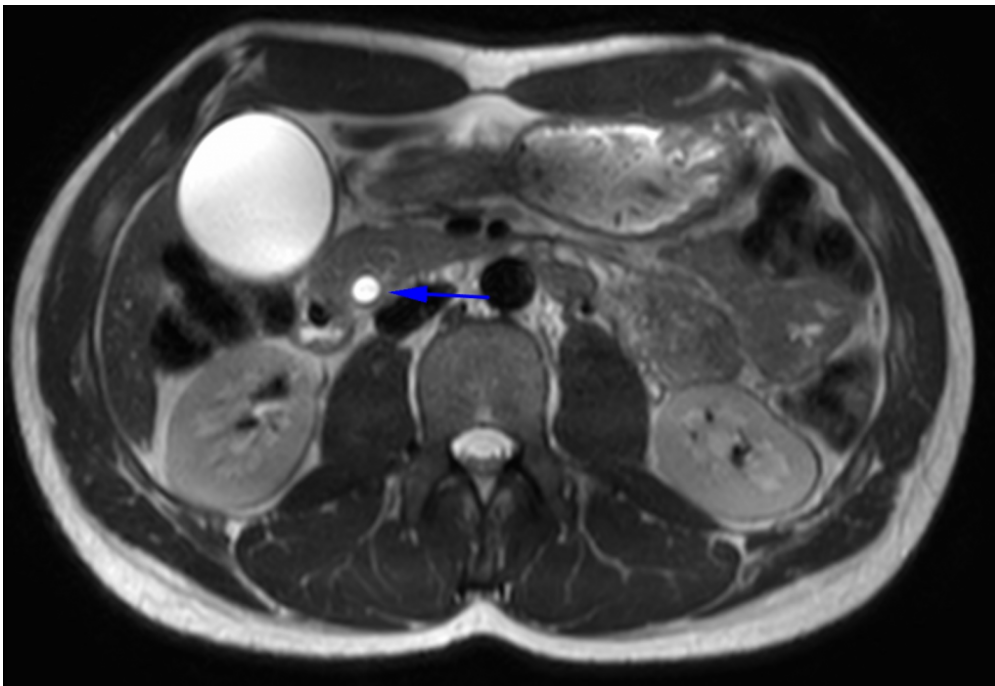
Figure 2

a



Description: The blue arrow shows the distal common bile duct. It is obstructed by material that was revealed to be sludge and micro-gallstones at ERCP (Endoscopic Retrograde Cholangio-Pancreatography). **Origin:** HUG

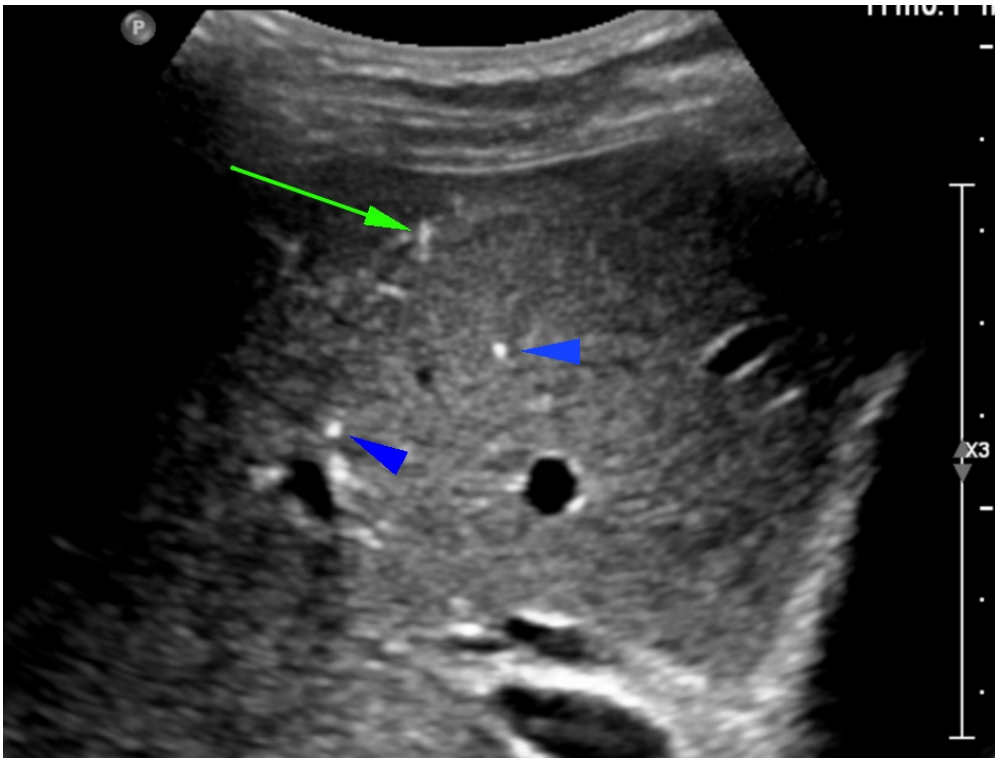
b



Description: Previous slice.
The blue arrow shows the dilated common bile duct. **Origin:** HUG

Figure 3

a



Description: The blue arrowheads show millimetric hyperechogenicities, and the green arrow shows the comet tail artefact.

Both images represent intra-hepatic millimetric gallstones. **Origin:** HUG