

Biliary Cast - Complication of Cholangitis and Pancreatitis in a Pancreas Divisum Patient

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Rezumat

Tromb biliar - complicație secundară colangitei și pancreatitei în cazul unui pancreas divisum

Prezentăm o cauză rară de "tromb biliar" secundar colangitei și pancreatitei, la o pacienta în vârstă de 60 de ani, cu pancreas divisum care a fost internată în spitalul nostru cu o pancreatită acută (etiologia alcoolică a fost exclusă) complicată cu abces pancreatic și icter obstructiv. Pacienta a suferit o intervenție chirurgicală complexă: colecistectomia, coledocotomie cu extragerea mularului biliar, drenaj biliar extern cu tub Kehr, evacuarea abcesului pancreatic, sequestrectomie, lavaj peritoneal și drenaje multiple. În ciuda tratamentului chirurgical și de terapie intensivă, drenajul biliar prin tubul Kehr s-a redus și icterul obstructiv a reapărut într-un mod chiar mai accentuat. Colangiografia endoscopică retrogradă a evidențiat pancreas divisum complet și stenoze difuze multiple alternând cu dilatarea arborelui biliar intrahepatic (similar colangitei sclerozante). O proteză biliară a fost plasată endoscopic până în canalul hepatic drept. În ciuda utilizării ERCP (colangiopancreatografia endoscopică retrogradă) combinată cu tratamentul cu UDCA (acid ursodeoxicolic) pentru gestionarea sindromului de tromb biliar, evoluția a fost nefavorabilă cu insuficiență hepatică acută, șoc septic și în final deces. Necropsia a relevat o distribuție biliară extinsă a trombului biliar în întreg arborele biliar și colangita piogenă.

Cuvinte cheie: tromb biliar, pancreas divisum, stent biliar, abces pancreatic, colangită purulentă

Abstract

We report a rare cause of "biliary cast" secondary to cholangitis and pancreatitis, in a 60-year-old female patient with pancreas divisum. She was admitted in our hospital with an acute pancreatitis (alcoholic etiology was excluded) complicated with pancreatic abscess and obstructive jaundice. The patient had undergone a complex surgical intervention: cholecystectomy, choledocotomy with extraction of the biliary thrombus, external biliary drainage through a T tube, evacuation of the pancreatic abscess, sequestrectomy, peritoneal lavage and multiple drainages. In spite of the surgical and intensive care support, the biliary drainage through the T tube had ceased and the obstructive jaundice had reappeared in a more accentuated fashion. Endoscopic retrograde cholangiography showed complete pancreas divisum and diffuse multiple stenosis alternating with dilatation of the intrahepatic biliary tree (a pattern of sclerosing cholangitis). An endoscopic prosthesis was placed inside the right hepatic bile duct. Despite the use of the combined endoscopic plus UDCA (ursodeoxycholic acid) treatment for the management of the biliary cast syndrome, the evolution was unfavorable with hepatic coma, septic shock and finally death. The necropsy revealed an extensive biliary cast in the entire biliary tree and pyogenic cholangitis. The patient had a fatal outcome despite all the surgical, endoscopic and conservative efforts, with development of intraductal biliary obstruction and secondary pyogenic cholangitis. Biliary cast syndrome is a rare but very aggressive entity and its management is often difficult despite the advances in surgery and endoscopy treatments.

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Key words: biliary cast, pancreas divisum, biliary stent, pancreatic abscess, suppurative cholangitis

Introduction

Biliary cast syndrome is a very rare and dangerous entity and is most frequently diagnosed in liver transplant patients (1-4). It was first described by Starzl et al. in 1977 (4). The cast formation is caused by unknown conditions most frequently by dehydration, cholangitis, tumors, hemolysis, ischemia, biliary duct lesions and recent surgery (5,6). The main cause in liver transplant patients is biliary duct ischemia.

We describe a female patient with pancreas divisum who developed a biliary cast following a severe angiocholitis and acute pancreatitis. The patient underwent an endoscopic retrograde cholangiography with a biliary stent placement, along with ursodeoxycholic acid administration, but the cholestasis remains unchanged. She developed a septic shock with hepatic coma.

Case report

A 60-years old woman with diabetes mellitus type II was admitted in our service with acute pancreatitis of unknown etiology (alcoholic etiology was excluded) and a pancreatic pseudocyst suspicion (first admission). Her medical history reveals diabetes mellitus diagnosed two years ago and a surgical intervention for hip prosthesis placement. Physical examination reveals epigastric pain and diminished intestinal transit. Total bilirubin level was 1.06 mg/dl (< 1.2 mg/dl ULN), direct bilirubin was 0.46 mg/dl ($< \text{ULN}$), alkaline phosphatase was 1538 UI/L ($< \text{ULN}$), gamma glutamyl transpeptidase was 893 UI/L ($< \text{ULN}$), serum amylase was 324 U/L ($< \text{ULN}$), glycemia was 311 mg/dl ($< \text{ULN}$), hemoglobin was 8.6 g/dl ($< \text{ULN}$), hematocrite was 26.2% and the rest of the biochemical and hematological values within normal limits. Body temperature varies between 36-37.5 °C. Abdominal ultrasound revealed the presence of a hypoechogenic mass at the level of the tail of the pancreas, having a diameter of 63 mm, fluid collection in the right sub-phrenic space and also in the anterior and posterior sub-hepatic space; transonic para-duodenal fluid collection measuring 3 mm width; a distended gall bladder without lithiasis, without any dilatation of common bile duct and intrahepatic biliary duct. She was discharged after 11 days of conservative treatment.

After about 5 weeks from the date of discharge, she returns with the signs of sclero-tegumentary jaundice, dark urine, pale stool, pruritus and mild hepatomegaly without abdominal sensibility upon palpation (second admission). The patient was diagnosed with obstructive jaundice with a pancreatic fluid collection with necrotic tissue (sequester), revealed by ultrasound examination and confirmed by

CT-scan. She underwent a surgical intervention and the intraoperative findings were calculous cholecystitis and pancreatic abscess with a large necrotic sequester. Because of the obstructive character of the jaundice, the common bile duct was also explored and the presence of some "biliary cast" was found, extending into both right and left hepatic ducts, from where complete extraction and removal were impossible because of its friable nature.

Cholecystectomy, choledochotomy with a T-tube insertion for external biliary drainage, evacuation of the pancreatic abscess and necrosectomy were performed. The initial post-operative evolution of the patient was favourable resuming the intestinal transit along with the reinstitution of per-oral alimentation and also the remission of the sclero-tegumentary jaundice, while the external bile draining T tube remained functional. The condition deteriorated starting with the 8th post-operative day when a reduction of the volume of the bile drained through the T tube was observed along with an increase in the value of the serum bilirubin level which accentuated the jaundice.

The abdominal ultrasound examination and internal medicine reevaluation diagnosed cholangitis and multiple hepatic microabscesses in the 4th segment of the liver in remissive status and the recommended treatment was: Ursofalk (ursodeoxycholic acid) and hepatoprotective medication. The patient continued to have an unfavourable evolution. She underwent repetitive ultrasound examinations at different intervals of time and the presence of a hepato-choledochal intraluminal mass was confirmed, which had an obstructive character and which also might be the cause of the modified ultrasound pattern seen in the hepatic parenchyma. A structural modification of the parenchyma at the level of the hilum of the liver was also observed. Suppuration at the level of the parietal incision appeared, which was thought to be due to a possible suppuration at the level of the pancreatic bed. During the evolution, she achieved at a point a status of partial amelioration and upon request of the family members she was discharged with a pre-programmed medication which she was supposed to continue after her discharge.

Evolution of the patient remained stationary at her domicile for a period of time after which she developed severe jaundice and she complained of severe abdominal pain mainly localized in the abdominal right upper quadrant; for this reason she was readmitted after 1 month and 11 days from her latest discharge date (third admission). On admission, the biochemical analysis revealed a total bilirubin level of 18 mg/dl, direct bilirubin level of 13.5 mg/dl, ASAT level of 36 UI/L and ALAT level of 26 UI/L.

A T-tube cholangiography with high pressure injection was performed showing strictures and dilatations of the intrahepatic biliary ducts and a leak at the insertion site into the common bile ducts. (Fig. 1)

This time she underwent endoscopic retrograde cholangiopancreatography (ERCP), where the result showed a complete pancreas divisum and a minimal dilatation of the intrahepatic bile ducts, which was opacified only after the injection under pressure of the contrast material that revealed multiple stenoses and dilatations of the biliary tracts. This specific

aspect was diffuse in all the intrahepatic branches of the bile duct.

A prosthesis of 10 Fr of 12 cm length was inserted in the right hepatic duct. The case was interpreted as a sclerosing cholangitis developed secondary to severe cholangitis. The patient continued to have an unfavorable evolution later developing hepatic coma, and eventually died.

At the time of autopsy examination the viscera had the specific aspect of septic shock, along with other findings like acute necrotico-hemoragic pancreatitis, suppurative cholangitis (pus was exteriorized upon compression on the intraparenchymal biliary canaliculi), as well as the presence of intra-luminal biliary cast, formed by the precipitation of bile salts, in the dilated intrahepatic biliary canaliculi. (Figs. 2-4)

The specimen obtained from the bile ducts at the time of autopsy reveals abundant fibro-leucocytic infiltration, steato-necrotic remains of the acute pancreatitis process and bile. No viable cellular or tissue elements were found. From these findings the extensive necrosis of the biliary tract

epithelium was also considered to be one of the ongoing pathological processes. (Figs. 5-6)

Discussion

The case presented here catches our interest from the point of view of a rare complication which might appear secondary to the attack of cholangitis and of acute pancreatitis – the biliary cast.

The data in the literature about biliary cast are rare and it appears mainly as a posttransplant pathology (2,4,7). Also, there are other causes of tumoral thrombus (8) or hemobilia. Regarding the non-transplant etiology there are only few references (5,9-12).

The causes of biliary cast syndrome (“biliary thrombus”) in our case are: severe infection with multiple stenoses and dilatation of the biliary tree – a pattern of secondary sclerosing cholangitis; severe dehydration with a low choleresis and viscous bile. We cannot exclude a minute hemobilia (the

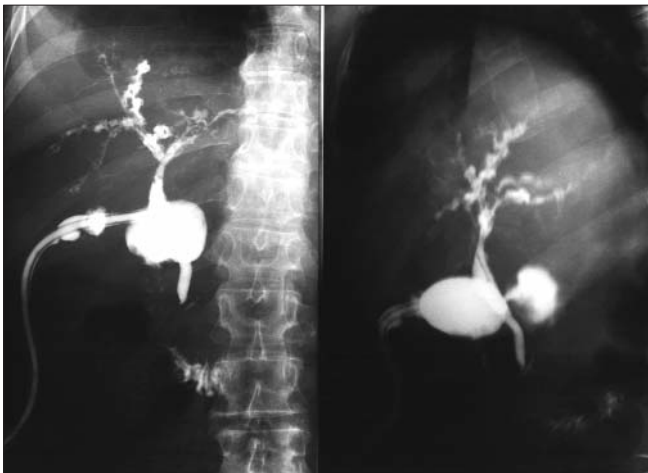


Figure 1. T-tube cholangiography. A small leak appears because of the high pressure used to inject the contrast



Figure 2. Biliary cast with the prosthesis in the intrahepatic biliary ducts

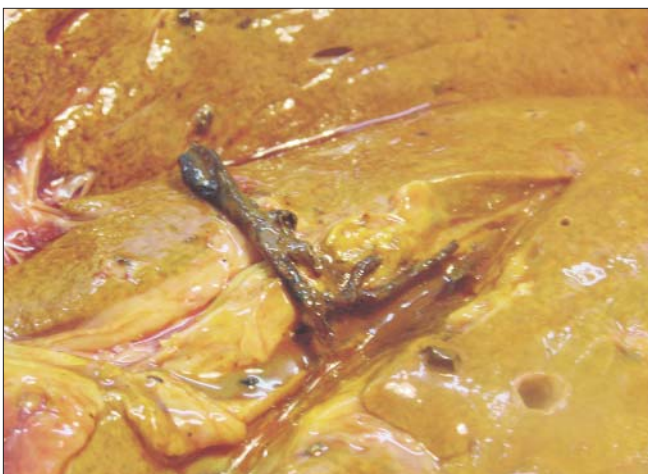


Figure 3. Branched biliary cast with pus in the intrahepatic biliary ducts



Figure 4. Branched biliary casts removed from the liver

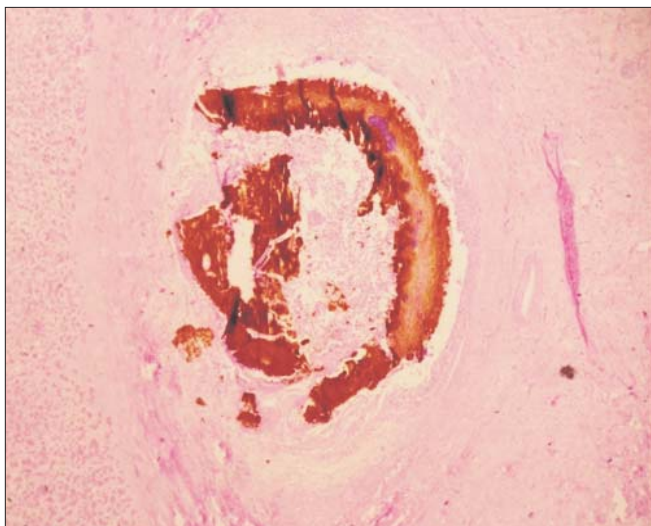


Figure 5. Histopathological aspect: abundant fibro-leucocytic infiltrate, no viable cellular elements (4x)

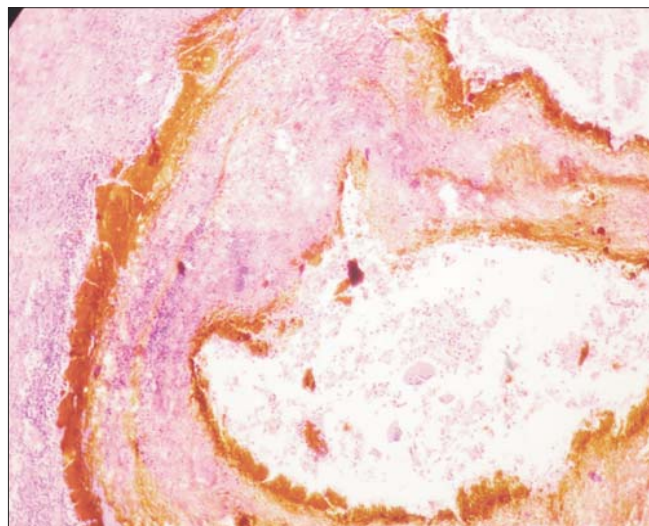


Figure 6. Histopathological aspect: abundant fibro-leucocytic infiltrate, no viable cellular elements (10x)

patient was anemic) or the treatment with cephalosporins as cofactors.

In our case neither lithiasis nor the pancreas divisum can be confirmed as the initiating agent of the chain of pathophysiologic processes. In the literature an abundant number of acute pancreatitis cases posing different grades of severity can be found, all having the same gallstone etiology, but very less often do we come across those cases where we find intrahepatic biliary canaliculi obstruction due to acute pancreatitis, as this pathology is more often seen in chronic pancreatitis (13,14).

The studies had shown the possibilities of different types of hepatic resections for different types of regional intrahepatic lithiasis, but in the case of our patient it was not possible due to the diffuse nature of the intrahepatic obstruction, to which later added the extremely severe systemic inflammatory response as an additional cause due to which such a measure could no longer be taken. Practically the treatment administered was directed towards remission of the acute pancreatitis process, treatment of the obstructive jaundice and to prevent any episode of cholangitis, the pathology which finally led to the appearance of a septic shock phenomenon.

The presented case shows a rare complication of acute pancreatitis, consequence of the pancreas divisum and of cholangitis - the biliary cast - which is formed by the precipitation of bile salts as a consequence of intrahepatic bile duct obstruction.

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