CASE STUDY

CONTENTS

"Billiary-cast" syndrome in a patient with acute biliary pancreatitis and pulmonary embolism

Olexii Dronov, Dmytro Vlasenko, Vladyslav Makarov
0.0. BOGOMOLETS NATIONAL MEDICAL UNIVERSITY, KYIV, UKRAINE

ABSTRACT

"Biliary-cast syndrome" ("BCS") is most often encountered in clinical practice as a complication after liver transplantation, there are also described cases of biliary-cast syndrome in patients who did not undergo liver transplantation, isolated cases of "BCS" developing in patients with acute pancreatitis, choledocholithiasis are described in literature. Ischemic damage to bile duct epithelium with development of cholestasis and retrograde biliary tract infection are considered as the main etiological factors. This work presents a clinical case of "Biliary-cast syndrome" in a patient with acute biliary pancreatitis and pulmonary embolism.

KEY WORDS: biliary cast syndrome, BCS, acute pancreatitis

Wiad Lek. 2024;77(12):2559-2562. doi: 10.36740/WLek/194268 DOI

INTRODUCTION

«Biliary-cast» syndrome is a pathological condition in which there is an obstruction of the biliary tract by formations, which are repeating the shape of the bile ducts, unlike gallstones, in the histological structure of the "casts" there is an luminal epithelium of the biliary tracts. Pathophysiologically, gallstone disease develops as a consequence of impaired metabolism of bile pigments and, or cholesterol. «BCS» – develops as a consequence of a violation of microcirculation in the arterial blood supply of the biliary system, at different levels, which leads to desquamatization of the luminal duct epithelium, cholestasis, the formation of a cast of the lumen of the duct with subsequent obstruction of the latter, the development of cholangitis and cholangiogenic abscesses. «Biliary-cast Syndrome" mostly described as a complication after liver transplantation (2.5 – 4%) [1-3]. The main mechanism of development is ischemic damage to the biliary tract, desquamatization of the integumentary epithelium and cholestasis lead to the formation of «casts» of the bile ducts [6]. The clinical picture corresponds to mechanical jaundice, cholangitis. The most sensitive diagnostic method is MR – cholangiopancreatography. Ultrasound can be useful for confirmation of biliary hypertension, but it is not always possible to visualize the «cast» on sonography. Endoscopic retrograde cholangiopancreatography combines both diagnostic and therapeutic techniques, with the help of endoscopic papillosphincterotomy, it is possible to clear lumens of bile ducts. Treatment, in most cases, is surgical, removal of the "cast" in order to restore the passage of the biliary tract.

In our case, there was a total necrosis of the epithelium of the internal bile ducts. With necrosis of the extrahepatic bile ducts.

CASE REPORT

Patient D., born in 1973, was delivered by ambulance. On admission, the patient complains of epigastric pain which is spreading to the right hypochondrium, dry mouth. He has a history of right hip fracture complicated by pulmonary embolism in 2021. Total bilirubin on admission 102 mmol/l. blood amylase 605 U. On abdominal ultrasound: gallbladder is not enlarged, gallblader`s wall is 3 mm. Contains calculi up to 5.0 mm. in diameter; The common bile duct s is dilated to 9 mm.

Patient was diagnosed with Acute biliary pancreatitis. Endoscopic retrograde cholangiopancreatography was performed: biliary sludge and stones up to 2 mm. were identified in the lumen of the CBD. Papillosphincterotomy was performed, stones and debree were removed.

On the second day of the postoperative period, patient's condition deteriorated sharply, complaints of weakness, shortness of breath developed. On examination, cyanosis of the nasolabial triangle is noted, respiration rate up to 22 per minute. Laboratory: D-dimer – 1.08 mg/l. Pulmonary embolism was diagnosed. The patient was transferred to

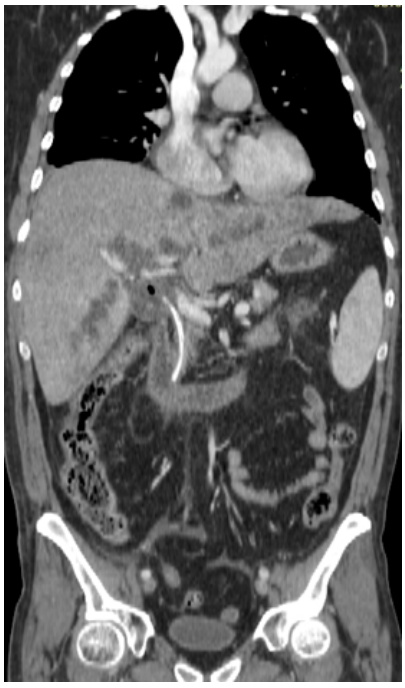

Fig. 1. liver parenchyma, along the course of the biliary tract, multiple confluent hypodense foci surrounded by a zone of perifocal edema are determined. Aerobilia. Internal and extrahepatic bile ducts are diffusely dilated with signs of aerobilia.

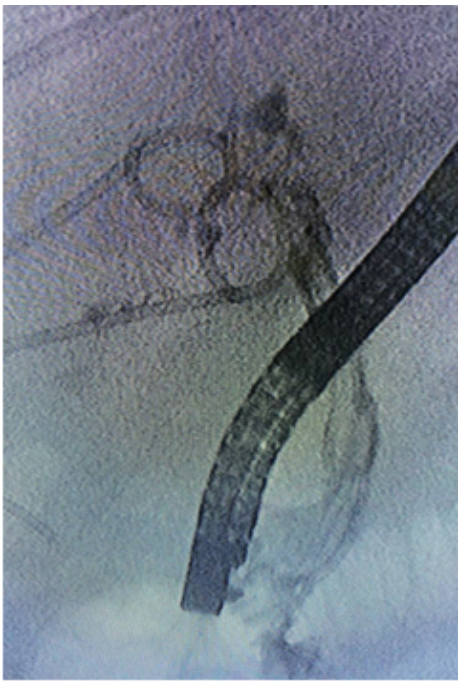

Fig. 2. ERCP: diffuse ductal dilation, proximal choledochal stenosis.

Fig. 3. "Casts" removed from segmental, lobar bile ducts. Necrotic common hepatic and common bile ducts.

the intensive care unit, where cardiac arrest was recorded within 5 minutes, resuscitative measures were carried out, after the restoration of cardiac activity, artificial lung ventilation was started, on which, patient remained for 12 hours. After extubation, patients hemodynamics remained unstable, inotropic support continued up to 3 days. The patient's condition stabilized. Blood pressure 110/70 mmHg, pulse 74/min, Sp02-97%. Leukocytes – 17.5 x 10^9. Hemoglobin – 111 g/l., Bilirubin – 68.1 mmol/l., urea – 9.3 mmol/l., creatinine – 163 mmol/l. On abdominal ultrasound: the liver is enlarged, the echogenicity of the parenchyma is increased. The echostructure is heterogeneous due to multiple anechoic formations localized in the right lobe, ranging in size from 16 to 23 mm. Intrahepatic bile ducts are dilated to 2-3 mm. Common bile duct is dilated up to 10 mm. Contains sludge.

Despite the biliary decompression, the patient retains jaundice and fever. A control ultrasound of the abdominal organs revealed numerous anechoic formations in the right lobe of the liver. Dilated intrahepatic bile ducts. Expanded to 1 cm. common bile duct, filled with echo-positive content. Total bilirubin – 91.2 mmol/L. A decision was made to repeat endoscopic retrograde intervention with stenting of the common bile duct: an endobiliary stent diameter of 8.5 Fr was installed, the outflow of bile into the lumen of the duodenum was observed. On the 26th day from the onset of the disease, computed tomography of the abdominal cavity was performed (Fig. 1).

Under ultrasound guidance, drainage of intrahepatic abscesses was performed. Obtained pus was sent for microbiological evaluation. Result: E.coli 5 x 10^7 CFU, Kl. Pneumoniae 5 x10^7 CFU, St. epidermiditis 5 x 10^5 CFU, sensitive to cefopyrazone. Despite the conservative treatment, the patient's condition remains severe, the patient has hyperthermia up to 38.5^{0}C, white blood cell count up to 15 x 10^9, severe anemia – hemoglobin 70 g/l. On the 37th day of the disease, CT control of the abdominal cavity was performed, on which a positive dynamic was determined – a moderate decrease the size and number of liver abscesses, much less pronounced dilatation of the biliary tract.

On the 58th day of the disease, endoscopic retrograde cholangiopancreatography was repeated: the left and right lobar ducts were dilated, diffusely, unevenly. The zone of stenosis of the proximal part of the CBD was determined (Fig. 2).

Given the lack of a positive response to conservative treatment, septic condition of the patient. A decision was made on surgical intervention in the scope of debridement and drainage of intrahepatic abscesses. Intraoperatively, the liver is enlarged, the right lobe is

violet – blue in color, jelly-like consistency. In the 4b segment, there is an abscess up to 3 cm. in diameter. The gallbladder is wrapped in omentum, contents: a single calculus up to 1 cm. in diameter. The hepatoduodenal ligament is infiltrated, edematous. After division of hepatoduodenal ligament, a necrotized common bile duct was identified, with a defect along the posterior wall up to 1 cm. Necrotic changes extended up to the confluence, left and right lobe ducts. During the removal of necrotized walls of the common hepatic duct, cast like formations of the segmental bile ducts were removed from the liver parenchyma, mucous pus with bile removed from the wound canals. (Fig. 3). External drainage of the intrahepatic bile ducts was performed.

In the postoperative period, patient remained jaundiced, bilirubin 187.2, direct fraction – 147 mmol/l. White blood cell count – 14.0 x 10^9, ALT – 196, AST – 233. The debit of bile through drains, installed in the intrahepatic ducts, was up to 100 ml of bile with purulent material, per day. Patient stayed in intensive care unit, antibiotic therapy was performed: Tigecycline, Amikacin. For 37 days from the moment of surgical treatment, the patient's condition remained severe, without significant dynamics. On the 38th day of the postoperative period, the patient's condition deteriorated sharply. Complaints of weakness, shortness of breath, tachycardia were noted. Hemoglobin – 65 g/l. Intra-abdominal hemorrhage was diagnosed. The patient was taken to the operating room, a relaparotomy was performed, the source of bleeding was identified – a defect in common hepatic artery up to 1 centimeter in diameter. Hemostasis was accomplished by suturing common hepatic artery. The patient was transferred to the intensive care unit in critical condition, in which he remained for 2 days. On the 105th day from the onset of the disease, patient died of cardiac arrest.

"Biliary-cast" syndrome is a rare phenomenon. In clinical practice, often described as a complication in patients after liver transplantation [1-3], cases of this syndrome have also been described in patients with biliary pancreatitis [4] and choledocholithiasis [5]. The main mechanism of development is ischemia of the bile ducts, desquamatization of intraluminal epithelium and cholestasis lead to the formation of "cast-like" formations inside bile ducts. By their structure and chemical composition are similar to that of gallstones [6]. These "casts" may form intrahepaticaly and extrahepatically, cause mechanical obstruction of the bile ducts, followed by an infection and development of cholangitis, necrosis of the biliary tract, and the formation of cholangiogenic abscesses. Lemmers et al. in their paper, where 14 cases of BCS in patients after liver transplantation were prospectively evaluated, propose the hypothesis that "Biliary-cast" syndrome is not a variant of prolonged ischemic damage to the bile ducts, but is an independent disease. In favor of this hypothesis, the authors note the specific nature of secondary strictures of the biliary tract. Absence of time correlation from the moment of liver transplantation prior to the onset of BCS or other non-anastomotic complications[3]. Voigtländer et al. In their retrospective study in liver transplant patients, ischemic injury is preferred in determining the etiology of BCS. Mainly, microcirculation disorders as a consequence of renal failure in patients after liver transplantation[7]. Currently, there is no concensus on management of biliary cast syndrome. This is due to the occurrence of BCS in liver transplant patients of 2.5%[1]. And even fewer patients with BCS who do not have a history of liver transplantation.

The clinical picture is similar to cholangitis, patients complain of jaundice, pain in the right hypochondrium, chills. Ultrasound diagnostics makes it possible to confirm the mechanical etiology of jaundice, the presence of dilated bile ducts and to detect hyperechoic "casts" in the lumen of the ducts. Magnetic resonance cholangiopancreatography has diagnostic sensitivity and specificity of 0.95 and 0.92, respectively. On MRCP, the "casts" appear as hypointense filling defects inside the bile ducts, surrounded by a ring of hyperintense bile. On computed tomography with contrast, it is possible not to identify "casts", especially in the early stages of the disease, due to the fact that the latter can be of the same density as the liver parenchyma. Severe cases of "Biliary-cast" syndrome have manifestations of severe biliary hypertension, with intraluminal hypodense formations, with increased contrast and thickening of the walls of the bile ducts.

The main direction in treatment of «Biliary-cast» syndrome is the elimination of the mechanical block of the bile ducts, with restoration of bile passage. Endoscopic papillosphincterotomy with endoscopic debridement and extraction is used as a therapeutic method with success in 25% to 60% of cases[9,10]. If endoscopic retrograde papillosphincterotomy is ineffective, percutaneous transhepatic biliary decompression techniques under ultrasound and radiological control are used.

CONCLUSIONS

The problem of "Biliary-cast" syndrome remains insufficiently studied, now this syndrome occurs less and less often as a complication after liver transplantation due to the evolution of surgical technique. The occurrence of "Biliary-cast" syndrome without liver transplantation is represented by isolated cases in the world literature, often in severe, comorbid patients.

REFERENCES

1. Gor NV, Levy RM, Ahn J et al. Biliary cast syndrome following liver transplantation: Predictive factors and clinical outcomes. Liver Transpl. 2008;14(10):1466-72. doi: 10.1002/lt.21492. DOI
2. Zhu XD, Shen ZY, Chen XG, Zang YJ. Pathotyping and clinical manifestations of biliary cast syndrome in patients after an orthotopic liver transplant. Exp Clin Transplant. 2013;11(2):142-9. doi: 10.6002/ect.2012.0035. DOI
3. Lemmers A, Pezzullo M, Hadefi A et al. (2021). Biliary cast syndrome after liver transplantation: A cholangiographic evolution study. J Gastroenterol Hepatol. 2021;36(5):1366-1377. doi: 10.1111/jgh.15318. DOI
4. Kusnierz K, Nowakowska-Dulawa E, Pilch-Kowalczyk J, Lampe P. Biliary cast syndrome in a non-transplant patient with acute pancreatitis. In Digestive and Liver Disease. 2012;44(6):534–535. doi:10.1016/j.dld.2012.01.011. DOI
5. Park JC, Park JG. (2021). A case of biliary cast syndrome after endoscopic and percutaneous management of common bile duct stone. Taehan Yongsang Uihakhoe Chi. 2022;83(1):224-229. doi: 10.3348/jksr.2021.0013. DOI
6. Yang Y-L, Zhang C, Lin M-J et al. Biliary casts after liver transplantation: Morphology and biochemical analysis. World J Gastroenterol. 2013;19(43):7772–7777. doi:10.3748/wjg.v19. DOI
7. Voigtländer T, Negm AA, Strassburg CP et al. Biliary cast syndrome post-liver transplantation: risk factors and outcome. Liver Int. 2013;33(8):1287-92. doi: 10.1111/liv.12181. DOI

CONFLICT OF INTEREST

The Authors declare no conflict of interest

CORRESPONDING AUTHOR

Vladyslav Makarov
O.O. Bogomolets National Medical University
13 T. Shevchenko boulevard, 01601Kyiv, Ukraine
e-mail: vlad111776@gmail.com

ORCID AND CONTRIBUTIONSHIP

Olexii Dronov: 0000-0003-4033-3195 A E F
Dmytro Vlasenko: 0000-0002-8218-2703 B
Vladyslav Makarov: 0009-0009-7778-7079 B D

A – Work concept and design, B – Data collection and analysis, C – Responsibility for statistical analysis, D – Writing the article, E – Critical review, F – Final approval of the article

RECEIVED: 27.02.2024
ACCEPTED: 08.10.2024