CASE REPORT

Secondary sclerosing cholangitis rapidly leading to liver cirrhosis: a possible post-ICU treatment sequel

M.C. Reichert¹, C. Jüngst¹, F. Grünhage¹, F. Lammert¹ and M. Krawczyk¹,²

From the ¹Department of Medicine II, Saarland University Medical Center, Homburg, Germany and ²Laboratory of Metabolic Liver Diseases, Department of General, Transplant and Liver Surgery, Medical University of Warsaw, Warsaw, Poland

Address correspondence to Dr M. Krawczyk, Department of Medicine II, Saarland University Medical Center, Saarland University, Kirrberger St. 100, 66421 Homburg/Saar, Germany. email: marcin.krawczyk@uks.eu

Learning points for clinicians
Secondary sclerosing cholangitis in critically ill patients (SSC-CIP) should be suspected in patients who present with cholestasis after treatment on intensive care unit, regardless of the duration of the intensive care therapy. Increased awareness of this condition is necessary, because it may rapidly lead to liver cirrhosis. ERCP, ursodeoxycholic acid and transplantation represent currently available therapeutic options.

Case presentation
A 64-year-old man was admitted to our department with increasing jaundice (bilirubin 9.1 mg/dl, gamma-GT 1492 U/l, AP 1745 U/l). He had a negative history of viral and non-viral liver diseases (including primary sclerosing cholangitis) and displayed no signs of inflammatory bowel disease in colonoscopy. Twenty-five days prior to admission, he underwent aortic valve replacement (biological aortic valve) due to high-grade aortic valve stenosis and right hemicolectomy for localized colonic adenocarcinoma (T4bN1bM0). The post-operative course was complicated by a 9-day stay on the intensive care unit (ICU) due to acute acalculous cholecystitis and subhepatic abscess with consecutive peritonitis. The patient underwent open cholecystectomy with percutaneous drainage of the abscess. Mechanical ventilation (average PEEP 5.25 mmHg, maximal PEEP 6 mmHg, FiO₂ < 0.5) and low-dose catecholamine therapy (maximal dose of norepinephrine 0.3 μg/kg/min) were necessary for a total of 4 days.

Given the increasing cholestatic jaundice after admission to our unit, we decided to perform endoscopic retrograde cholangiography (ERC) with sphincterotomy and biliary stent placement. The cholangiogram and magnetic resonance cholangiography showed typical findings for secondary sclerosing cholangitis in critically ill patients (SSC-CIP), namely diffuse rarification of the intrahepatic bile ducts (Figure 1A) and presence of biliary casts (Figure 1B). Despite optimal biliary drainage and treatment with 500 mg ursodeoxycholic acid twice daily, the patient developed repeated episodes of acute cholangitis, which had to be treated with antibiotics and biliary stent replacements. Biliary casts had to be removed repeatedly. The patient developed liver cirrhosis (currently Child-Pugh stage B), as indicated by transient elastography (liver stiffness 25.1 kPa), within a timeframe of 5 months. Of note, a CT scan performed 2 months before the ICU admission did not show any signs for liver cirrhosis (Figure 1C).

Discussion
SSC-CIP is a newly described liver condition,¹ which should be suspected in all patients presenting with cholestasis after treatment on ICU.² The formation of biliary casts containing lithogenic material mixed with necrotic biliary epithelium is a typical sign of SSC-CIP.³ Our case appears to be exceptional since the intensive care treatment was short. Most of the described SSC-CIP cases occurred after prolonged and intensive treatment on the ICU,¹ which was not the case in our patient. Despite this fact liver cirrhosis developed rapidly after the onset of SSC-IP. Overall, SSC-CIP often represents a progressive condition with poor prognosis. Although currently no causal treatment or prophylaxis for SSC-CIP are available, clinicians should be aware of this condition and include it in the differential diagnosis of increased cholestatic markers after treatment on ICU, even after short stays.
Authorship

All authors had access to the case information and collaborated in writing this article.

Conflict of interest. None declared.

References


Figure 1 (A) Cholangiogram presenting an irregular bile duct system with multiple diffuse strictures, prestenotic dilatations and rarefactions of intrahepatic bile ducts. (B) Duodenoscopy showing a biliary cast protruding from the major papilla (C) CT scan of the liver 2 months before admission to ICU.