Journal of
Medical Case Reports

CASE REPORT

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Splenic artery embolization in a patient with advanced Caroli's syndrome complicated by portal hypertension: a case report

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Abstract

Background Caroli's disease is an inherited disorder characterized by dilatation of intrahepatic bile ducts and congenital hepatic fibrosis. Caroli's disease is complicated by choledocholithiasis, cholangitis, and portal hypertension.

Case presentation A 28-year-old Palestinian female with 7-year history of Caroli's disease complicated by liver cirrhosis, portal hypertension, and esophageal varices presented with hypersplenism and thrombocytopenia. She was managed with splenic artery embolization, which improved her platelet count. The splenic artery embolization was complicated by postembolization syndrome, and she was treated with antibiotics and supportive care. Two months later, she died owing to liver encephalopathy and spontaneous bacterial peritonitis.

Conclusion This case highlights the role of splenic artery embolization in managing hypersplenism associated with Caroli's disease. Despite improvement in platelet count, post-embolization syndrome remains a significant risk. Early liver transplantation should be considered in patients with advanced Caroli's syndrome to improve long-term outcomes.

Keywords Caroli's syndrome, Caroli's disease, Hypersplenism, Splenic artery embolization, Portal hypertension, Case report

Background

Caroli's disease (CD) is an uncommon congenital condition of the intrahepatic bile ducts. Segmental dilation of the intrahepatic bile ducts and liver fibrosis are characteristics of CD [1]. It can present in two forms: the simple form, involving isolated bile duct dilation, and Caroli syndrome (CS), a more complex form associated with congenital hepatic fibrosis [2]. CD is extremely rare, with an estimated incidence of 1 in 1,000,000 [3]. Abnormal differentiation of the ductal plate into mature bile ducts results in bile duct dilatation, intrahepatic cysts, and increased risk for cholangitis and stone formation [4, 5]. *PKHD1* gene mutation is implicated in CS; it encodes the protein fibrocystin, and defects in this protein play an important role in forming biliary cysts and the portal fibrosis characteristic of CS [6].

CD presents with a wide range of symptoms. It may be diagnosed incidentally in asymptomatic patients. Symptomatic patients present with recurrent abdominal pain, jaundice, and cholangitis [7]. In cases of CS, patients have complications due to portal hypertension [8]. Diagnosis is usually initiated by noninvasive imaging modalities such as ultrasonography (USG), computed tomography (CT), and magnetic resonance

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imaging (MRI), Definitive diagnosis is made through histopathological examination [9]. The present case report aims to highlight the role of splenic artery embolization as a therapy option for hypersplenism in the context of portal hypertension.

Case presentation

A Palestinian female in her twenties was diagnosed 7 years ago with Carol's disease, in addition to liver cirrhosis and portal hypertension. She had undergone multiple band-ligation sessions and intensive care unite (ICU) admissions owing to significant upper gastrointestinal bleeding from esophageal varices. Management planned to do a liver transplant, but the patient refused, so she was kept on medical treatment to control the complications.

At the current admission, the patient consulted her gastroenterologist owing to history of left upper abdominal pain and distention for 1 month; an ultrasound was done in the clinic and showed severe splenomegaly. To manage her hypersplenism, the patient was admitted to the hospital. The patient was stable on admission with left upper quadrant tenderness and a left upper quadrant mass crossing the umbilicus. Body temperature was 36.9 °C, heart rate was 90 beats per minute, and blood pressure was 100/60 mmHg. Laboratory tests revealed a platelet count of 29,000/μL, a hemoglobin level of 8.6, and a white blood cell count of 8509/μL. Liver enzymes showed a slight elevation, with an Aspartate Aminotransferase (AST) and Alanine Aminotransferase (ALT) count of 58.8 U/L and 30 U/L, respectively, and a gamma-glutamyl transferase (GGT) of 54 U/L. Bilirubin, prothrombin time (PT), partial thromboplastin time (PTT), and albumin were normal at admission. Owing to her hypersplenism and thrombocytopenia, she was scheduled for splenic artery embolization (SAE) and given a platelet transfusion.

The splenic artery embolization (SAE) procedure was performed under local anesthesia through a right femoral artery puncture. Using ultrasound guidance, a 5F femoral introducer sheath was inserted, and a 4F C2 catheter, along with a 0.035×150 cm Terumo guidewire, was advanced into the splenic artery. A baseline splenic angiogram revealed normal vascularization with increased blood flow to the spleen (Fig. 1). Intraarterial administration of 1 g ampicillin, 80 mg gentamicin, and 8 mg dexamethasone was performed. Embolization was then performed distal to the pancreatic branches. The embolization materials included polyvinyl alcohol (PVA) particles and gel foam. Post-embolization angiogram showed near-total blockage of blood flow with only delayed and slow passage of contrast through the main branches (Fig. 2). The patient was moved to the ICU for close monitoring without any immediate complications. Post-embolization computed tomography showed an enlarged spleen measuring about 30 cm with post-embolization changes, multifocal variable-sized cystic dilation of segmental intrahepatic bile ducts with a central dot sign suggesting Caroli's disease. The liver measured about 15 cm in CC dimension at the

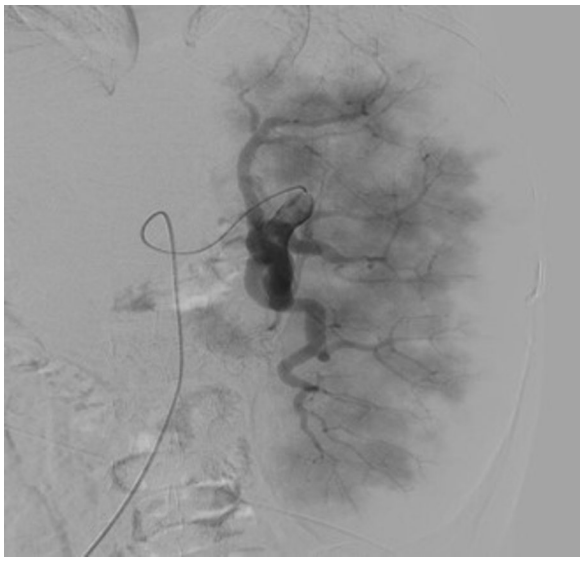

Fig. 1 Splenic angiogram showing extensive blood flow through the splenic artery and its branches, highlighting the high vascularization of the spleen

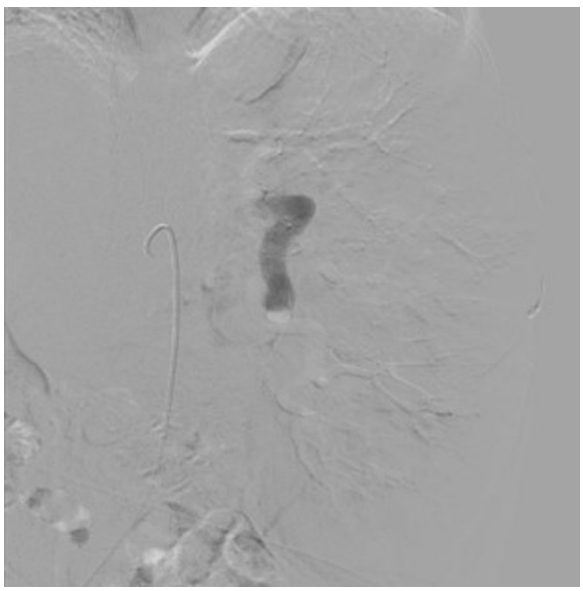

Fig. 2 Angiogram showing near-complete occlusion of the splenic artery. Blood flow is significantly reduced, with only minimal contrast passing through the blocked branches

midclavicular line with a prominent caudate lobe. The gallbladder appeared distended, measuring 6.3×11 cm without stones (Fig. 3). On day 2 after embolization, she experienced fever, tachycardia, and dyspnea. Laboratory testing showed elevated white blood cells (WBCs) of 19,000/μL, C-reactive protein (CRP) level of 157 mg/L, and an improved platelet count of 95,000/μL. Blood and urine cultures were negative. Chest X-ray revealed atelectasis, so she was kept on incentive spirometry. A computed tomography angiogram ruled out pulmonary embolism. Repeated chest x-ray showed left lower lobe infiltration, suggesting pneumonia, and she was given antibiotics. The patient's white blood cells and inflammatory markers improved over time. The platelet count was 394,000/μL on day 10. She was sent home for outpatient clinic evaluation.

Two months later, the patient had spontaneous bacterial peritonitis and hepatic encephalopathy, and she passed away.

Discussion

Caroli's syndrome is a rare congenital disorder characterized by segmental dilatation of the intrahepatic bile ducts. CS is complicated by recurrent cholangitis, biliary stones, and portal hypertension [10].

In this case, the patient had portal hypertension complicated by esophageal varices and hypersplenism. Management of portal hypertension often involves medical management, endoscopic interventions, and surgical options [11]. The primary clinical manifestation in this case was hypersplenism. Splenic artery embolization was performed to manage hypersplenism. SAE is a minimally invasive procedure that reduces arterial inflow to the spleen, which subsequently lowers venous outflow pressure. This decrease in splenic venous pressure translates to reduced portal inflow, effectively lowering portal vein pressure, which results in therapeutic benefits for patients with portal hypertension, such as reduced risk of variceal bleeding, improved control of ascites, alleviating hypersplenism, improving peripheral blood counts, and enhancing systemic circulation [12, 13]. However, embolization may weaken splenic immunity. Even without complete functional asplenia, SAE-induced infarcts and necrosis of splenic tissue may lead to localized immunosuppression, increasing the risk of splenic abscess, spontaneous bacterial peritonitis (SBP), or systemic inflammatory response syndrome (SIRS) [14].

In this case, the embolization successfully raised the patient's platelet count. Despite the initial improvement following SAE, the patient developed post-embolization syndrome, including fever, leukocytosis, and localized chest complications (atelectasis and pneumonia). Post-embolization complications highlight the importance of postoperative monitoring, as these patients are at risk for infections and other complications [15].

The long-term management of Caroli's disease remains a topic of debate among clinicians. Surgical resection is an adequate treatment option in cases of localized disease. Liver transplantation is required for patients with advanced disease or those who develop significant hepatic fibrosis. In this patient, the presence of liver cirrhosis and portal hypertension suggests that transplantation may improve her condition [16, 17].

Conclusion

This case highlights the role of splenic artery embolization (SAE) as a treatment for hypersplenism in patients with Caroli's syndrome. The procedure increased platelet counts and stabilized the patient's condition in

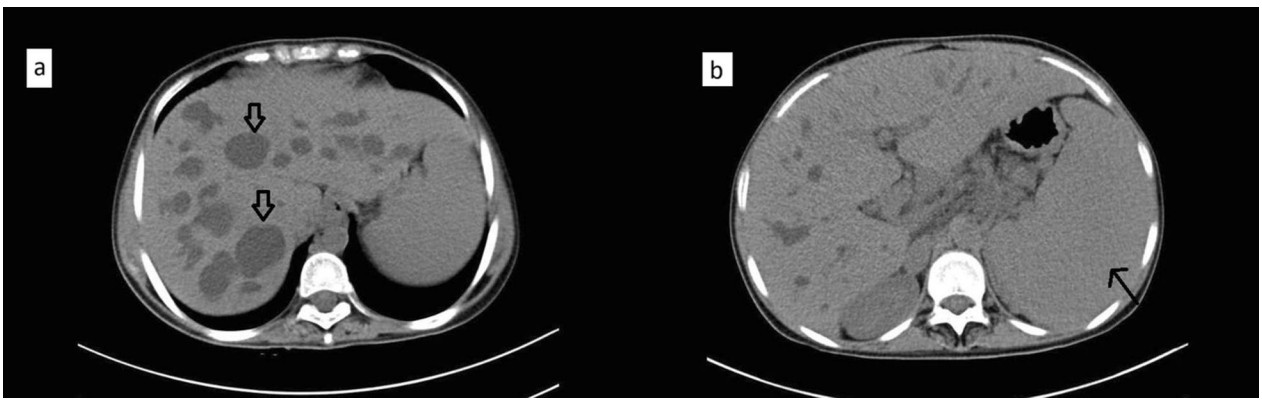

Fig. 3 **a** Axial contrast-enhanced computed tomography scan of the abdomen, demonstrating multiple intrahepatic cystic lesions (black arrows), representing saccular dilatation of the intrahepatic bile ducts, characteristic of Caroli's disease. **b** Axial contrast-enhanced computed tomography scan showing a larger saccular dilatation (black arrow) in the liver, along with hepatomegaly (enlarged liver) and splenomegaly (enlarged spleen)

the short term. However, post-embolization syndrome emphasizes the need for post-procedural monitoring. Early consideration of liver transplantation remains crucial for improving outcomes in patients with advanced Caroli's syndrome.

Acknowledgements
We thank all staff of the An-Najah National University Hospital who contributed to this report.

Author contributions
A.Z., M.A., and A.K. contributed to the conception and design of the study, acquired and analyzed case data, drafted the manuscript, and reviewed relevant literature. Y.K. interpreted imaging findings and contributed to manuscript revisions. Q.A. supervised patient care, ensured clinical content accuracy, and provided critical revisions. All authors critically reviewed the manuscript, approved the final version for publication, and agreed to be accountable for the accuracy and integrity of the work.

Funding
No external funding was provided for this study.

Data availability
All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Written informed consent was obtained from the patient's next of kin for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare no competing interests related to this case report.

Received: 31 January 2025 Accepted: 22 July 2025
Published online: 23 October 2025

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