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BRIEF COMMUNICATION

An Incidental Discovery of Caroli's Disease in an 86-Year-Old Presenting with Cholangitis

Robert Sean O'Neill[1,2]

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Abstract

An 86-year-old woman developed cholangitis post-femoral nail for a left pertrochanteric fracture and was found to have multilobulated intrahepatic bile ducts and hepatic cysts communicating with intrahepatic ducts, along with polycystic kidneys on abdominal imaging consistent with Caroli's disease. Caroli's disease is extremely rare and presentations in the elderly population are exceedingly uncommon. Treatment is usually supportive, with biliary drainage utilised in the first instance for management of cholangitis along with antibiotic therapy. Surgical resection and liver transplantation are utilised in those with end-stage liver disease.

Presentation

An 86-year-old woman developed fevers, right upper quadrant pain and nausea 4 days post-femoral nail for a left pertrochanteric fracture. Her medical history was notable for osteoarthritis, hypercholesterolaemia and hypertension. Medications on admission included amlodipine 5 mg OD, aspirin 100 mg OD, atorvastatin 10 mg nocte, metoprolol 100 mg BD and valsartan 100 mg nocte.

On clinical examination, jaundice was present. Her abdomen was tender to palpation in the right upper quadrant and bilateral kidneys were ballotable. The remainder of her clinical examination was unremarkable.

Her bilirubin was elevated to 88 µmol/L with cholangitic derangement of her liver function tests, while serum lipase was normal. Abdominal ultrasound demonstrated cholelithiasis without features of cholecystitis and a common bile duct diameter of 6 mm. The intrahepatic bile ducts were not dilated. Computed tomography (CT) of the abdomen showed polycystic kidney and liver disease (Fig. 1A). Magnetic resonance cholangiopancreatography (MRCP) demonstrated multilobulated intrahepatic ducts and hepatic cysts communicating with intrahepatic ducts consistent with Caroli's disease (Fig. 1B, arrow). No features suggestive of cholecystitis or choledocholithiasis were present.

She was managed with intravenous antibiotic therapy and regular analgesia. Her bilirubin normalised; however, her admission was complicated by a hospital-acquired pneumonia requiring further intravenous antibiotic therapy.

Discussion

Caroli's disease is a rare autosomal-recessive disorder characterised by abnormal segmental cystic non-obstructive dilation of the intrahepatic bile ducts with concomitant renal cysts [1]. The disease is associated with episodes of recurrent cholangitis along with the potential for life-threatening complications including sepsis, hepatic abscess formation or the development of cirrhosis.

The diagnosis of Caroli's disease is based on typical imaging features on CT such as the 'central dot' sign, along with MRCP demonstrating connections between secular ectasias and the normal biliary tract. Although endoscopic retrograde cholangiopancreatography (ERCP) has the highest level of sensitivity in diagnosing Caroli's disease, it is not routinely used in practice due to its invasive nature and the potential for infection [1].

Treatment is usually supportive, with biliary drainage in the first instance in the management of cholangitis along with antibiotic therapy. Surgical resection and liver

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✉ Robert Sean O'Neill
robert.oneill@health.nsw.gov.au

[1] St Vincent's Hospital, Sydney, NSW 2010, Australia

[2] St Vincent's Clinical School, The University of New South Wales, 290 Victoria Street, Darlinghurst, NSW 2010, Australia

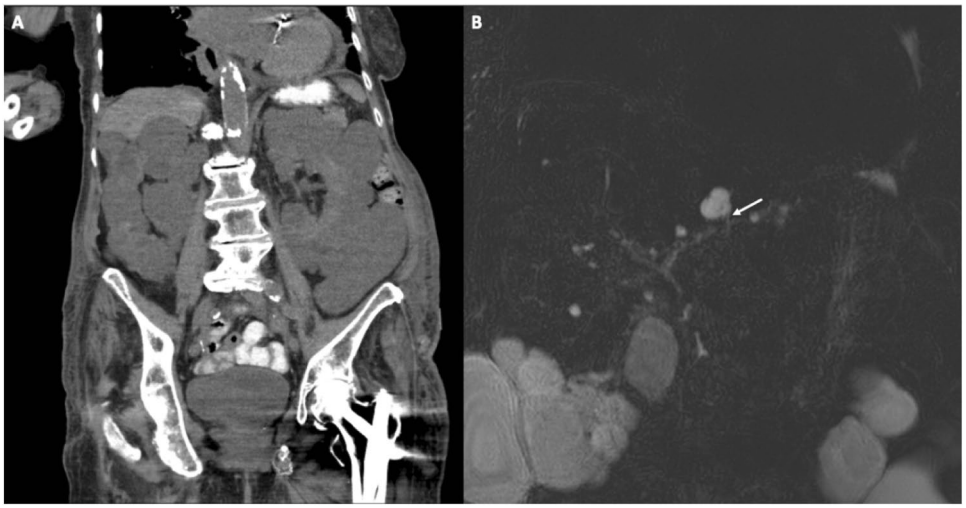

Fig. 1 **A** CT of the abdomen (coronal view) showing polycystic kidney and liver disease. **B** Magnetic resonance cholangiopancreatography (coronal view) showing multilobulated intrahepatic ducts and hepatic cysts communicating with intrahepatic ducts (designated by arrow). Also shown are renal cysts in the coronal plane

transplantation have been utilised previously in patients with end-stage liver disease [2].

The diagnosis of Caroli's disease is usually made during childhood or adolescence; however, it can also occur in adulthood as demonstrated in the presented case. Late presentations of the disease are not uncommon and have previously been published in the literature [3].

Author Contribution RSO consented the patient, cared for the patient, was involved in the conceptualisation of the manuscript, drafting and editing, as well as the submission of the presented manuscript.

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Declarations

Ethics Approval The manuscript was not deemed to require ethics approval by the institutional ethics review committee.

Consent to Participate The patient provided verbal and written consent for the inclusion of their personal information in the presented manuscript for publication.

Consent for Publication The patient provided verbal and written consent for publication.

Conflict of Interest The author declares no competing interests.

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