

Introduction

- Primary intrahepatic cholelithiasis is associated with brown or pigmented stones and recurrent pyogenic cholangitis.
- Resultant from biliary stasis and infection
- Other important potential chronic complications exist beyond cholangitis of which gastroenterologists should be aware.

Case

- A 57 year-old man with history of diabetes returned from a 13-month stay in Japan to be admitted to an outside facility with DKA and cholangitis.
- Found to have a liver abscess and left hepatic duct stricture treated with sphincterotomy, biliary stenting via ERCP, abscess drainage, and cholecystectomy.
- Followed up externally for stent removal with normalization of liver chemistries
- CT imaging 1 year later showed persistent L hepatic duct stricture with intrahepatic segmental dilation.
- No early satiety, weight loss, or jaundice.
- Referred to our service for further evaluation of chronic indeterminate biliary stricture.
- EUS demonstrated extensive small and large left-sided hepatolithiasis (A).
- Cholangiography during ERCP demonstrated complete filling defect of the L intrahepatic biliary ductal system (B) preventing selective guidewire passage.
- Cholangioscopy (C I-III) resulted in successful electrohydraulic lithotripsy (EHL) of stone burden in the L main hepatic system for selective wire passage and stenting of the L intrahepatic system with a plastic stent. Biliary epithelium appeared smooth and benign.
- The patient was given antibiotics with no complications.

Endoscopy & Radiology

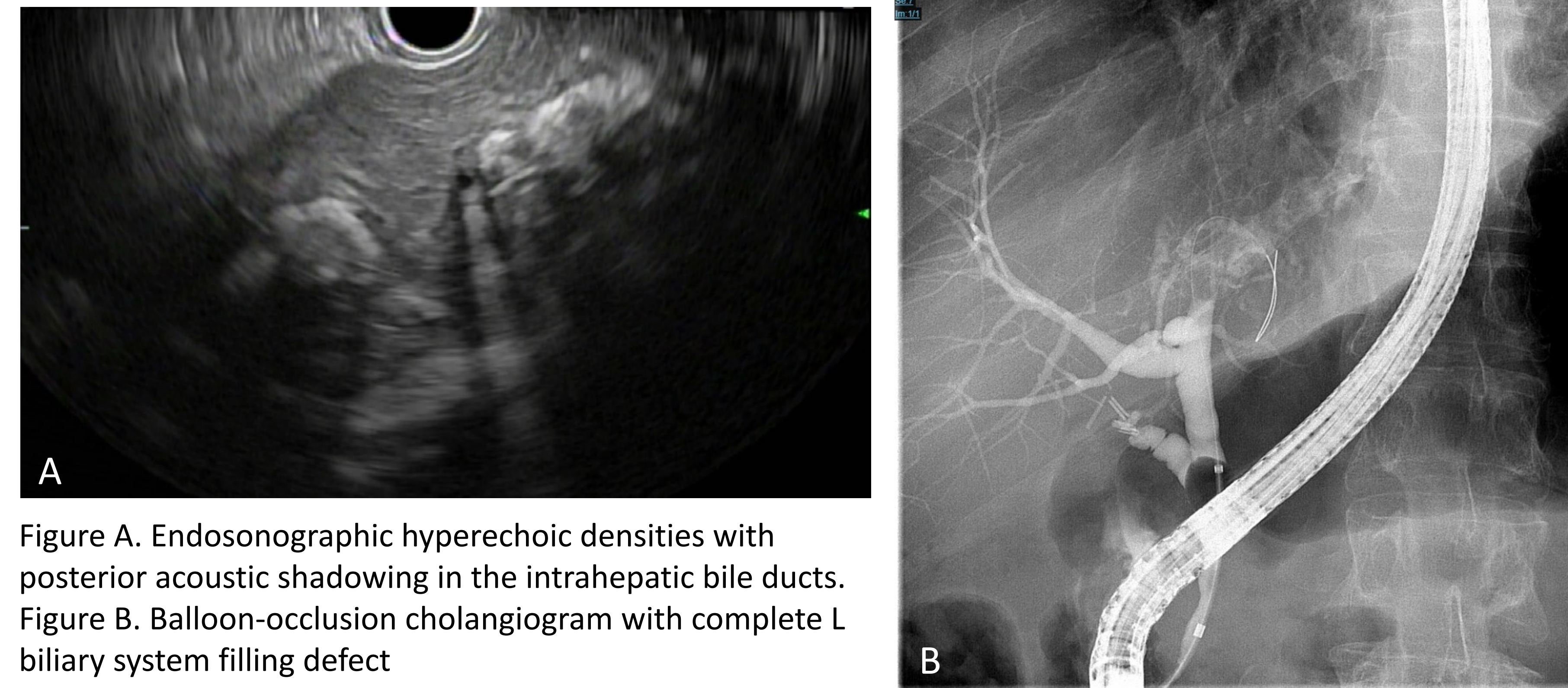


Figure A. Endosonographic hyperechoic densities with posterior acoustic shadowing in the intrahepatic bile ducts.
Figure B. Balloon-occlusion cholangiogram with complete L biliary system filling defect

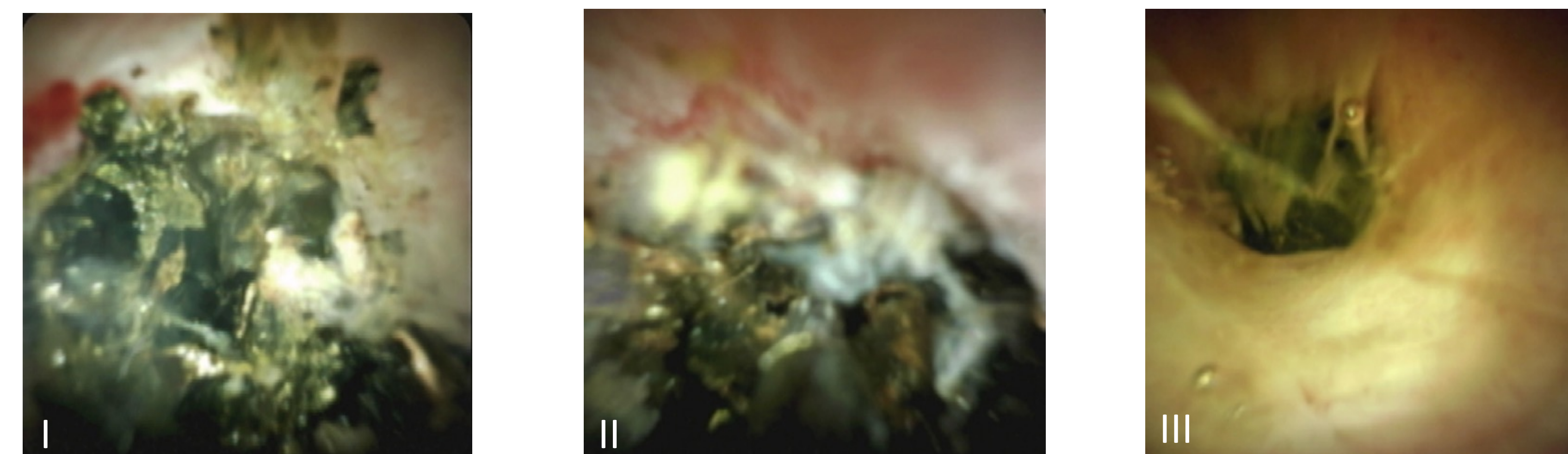


Figure C I-III. Cholangioscopy with stone fragmentation, benign-appearing biliary epithelium

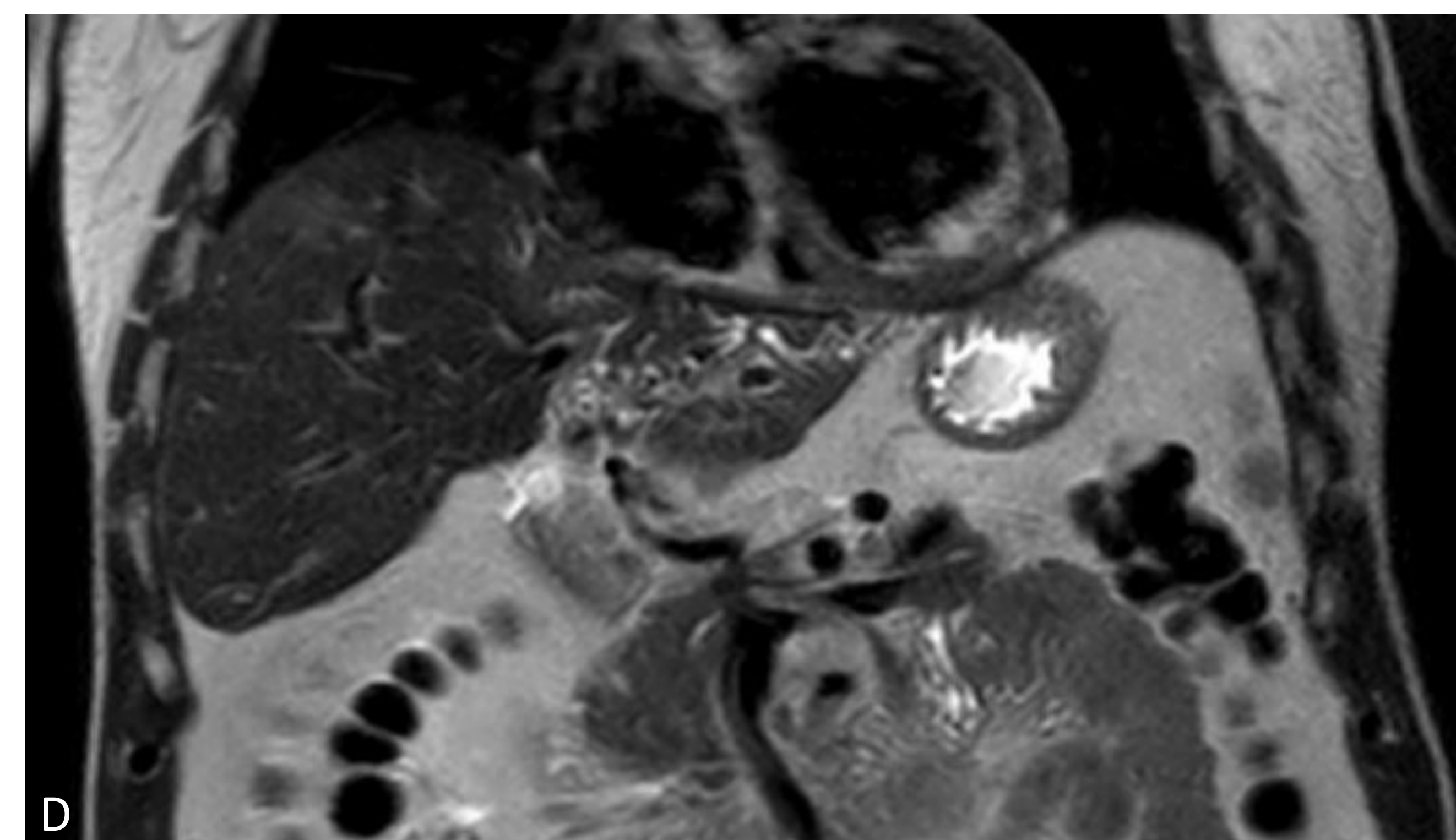


Figure D. Magnetic resonance imaging with atrophic left liver lobe in diseased segments

Case (Continued)

- ERCP 6 weeks later w/ cholangioscopy + EHL and stone removal in L intrahepatic ductal branches but ductal disease remained throughout smaller distal branches of the L lobe on cholangiogram.
- The main duct was protected with a fully-covered metal stent and antibiotics were given.
- Follow-up MRI/MRCP demonstrated atrophic left liver lobe (D) in addition to our findings.
- Left partial hepatectomy was thus performed for definitive therapy of the diseased lobe.
- The patient did well post-operatively with benign pathology.
- Ultimately followed up with stent removal and resolution of stone burden.

Discussion

- Treatment of other chronic sequelae of severe primary hepatolithiasis is important to reduce risk of progression to cirrhosis.
- Antibiotics are a cornerstone of therapy but may not be sufficient in cases of severe stone disease.
- Multiple interventional endoscopic methods are available and should be utilized to establish the diagnosis and optimize therapy.
- Surgery may be indicated in severe cases.
- While ursodeoxycholic acid (UDCA) would be insufficient as primary therapy, consideration can be given to a prophylaxis course after treatment of severe intrahepatic stone disease.

References

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