



Rarely Diagnosed IgG4-Related Sclerosing Cholangitis Identified with SpyGlass™ DS II Direct Visualization System

Pushpak Taunk, M.D.,
Associate Professor of Medicine
Division of Digestive Diseases and Nutrition
University of South Florida Morsani College of Medicine
Tampa General Hospital
Tampa, Florida



Patient History & Assessment

A 64-year-old male presented with obstructive jaundice in 2020. He was found to have a 4cm irregular pancreatic head mass and a dilated common bile duct (CBD) of 1.2cm on a CT scan. The endoscopic ultrasound (EUS) revealed a pancreatic head mass and Fine-needle aspiration (FNA) of the mass yielded atypical lymphoid infiltrate, suspicious for B-cell lymphoma, but further testing with flow cytometry was negative. Endoscopic retrograde cholangiopancreatography (ERCP) was notable for a mid CBD stricture, for which cytology and Fluorescence in Situ Hybridization (FISH) were negative. Because of inconclusive results, repeat EUS-FNB was performed and showed evidence of chronic pancreatitis, but no mass. Fine-needle biopsy (FNB) of the pancreas showed fibrosis and chronic inflammation. He then underwent an open, surgical, transduodenal biopsy of the pancreas which, again, showed chronic inflammation. Over the course of the following year, an MRI/MRCP was obtained which revealed irregular contour of the common hepatic and common bile ducts, raising concern for sclerosing cholangitis. Given this concern, repeat ERCP—now utilizing SpyGlass DS II Direct Visualization System—was performed.

Procedure Plan

The plan was to perform ERCP with the SpyGlass DS II Direct Visualization System and use SpyBite™ Max Biopsy Forceps to obtain biopsies of the biliary stricture and irregularities identified during direct visualization.

Procedure/Techniques Used

Cannulation was first achieved using a 9mm Extractor™ Pro retrieval Balloon Catheter and 0.035in x 260cm Hydra Jagwire™ Guidewire. Occlusion cholangiography revealed diffuse intrahepatic strictures. There was also a 3mm x 5mm mid-CBD stricture with irregular contour. Dilation of the mid CBD stricture was performed with 4mm x 4cm and 6mm x 4cm Hurricane™ RX Biliary Balloon Dilation Catheters. The SpyScope™ DS II Catheter was then advanced to the level of the mid-CBD stricture. The mucosa appeared nodular, erythematous and edematous. There were no irregular vessels. SpyBite Max was inserted through the SpyScope DS II Catheter and a total of 6 biopsies were taken from the irregular biliary mucosa.

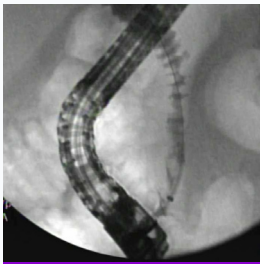


Figure 1
There are multifocal extrahepatic strictures present within the common bile duct in a continuous fashion.

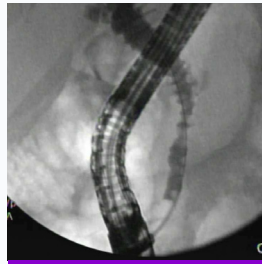


Figure 2
The strictures involve the mid to distal common bile ducts. The extrahepatic bile duct has an "accordion-like" appearance.

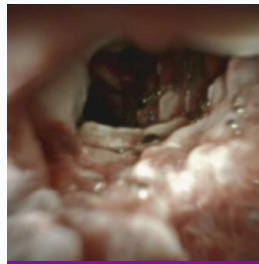


Figure 3
The biliary mucosa of the extrahepatic biliary strictures is erythematous, nodular and smooth in appearance.

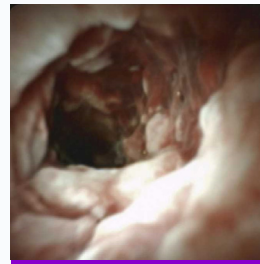


Figure 4
There are fibrinous white exudates, but without evidence of irregular vessels or mass lesions.



Figure 5
Direct visualization with SpyGlass™ DS II Direct Visualization System enabled a consistent view of these features throughout the bile duct.

Case Outcome

Upon pathological review, biopsy results showed extensive stromal lymphoplasmacytic inflammation with increased IgG4 plasma cells and eosinophils. These results were compatible with a diagnosis of IgG4 sclerosing cholangitis, a rare autoimmune disorder. The patient was started on steroids for treatment and responded well with a decrease in liver enzymes.

Case Discussion

The SpyGlass DS II Direct Visualization System enabled direct visualization and biopsy which supported the diagnosis of autoimmune IgG4 sclerosing cholangitis. Over the course of 18 months, the patient underwent two EUS procedures, one ERCP examination and a surgical biopsy – all without a definitive diagnosis. After using the SpyGlass DS II Direct Visualization System, in conjunction with SpyBite™ Max, a tissue sample was obtained which led to a diagnosis. The SpyScope DS II Catheter enabled direct visualization of a concerning inflammatory condition rather than a malignant disease. The device was also able to help the endoscopist visualize the biliary lesion for direct, targeted sampling which clinched an otherwise elusive diagnosis. The patient was able to undergo the right treatment due to the visualization and access provided by the SpyGlass DS II Direct Visualization System with SpyBite Max.

IMPORTANT INFORMATION: These materials are intended to describe common clinical considerations and procedural steps for the use of referenced technologies but may not be appropriate for every patient or case. Decisions surrounding patient care depend on the physician's professional judgment in consideration of all available information for the individual case. Boston Scientific (BSC) does not promote or encourage the use of its devices outside their approved labeling. Case studies are not necessarily representative of clinical outcomes in all cases as individual results may vary.

This case study was produced in cooperation with Dr. Pushpak Taunk. Results from case studies are not predictive of results in other cases. Results in other cases may vary.

CAUTION: U.S. Federal law restricts this device to sale by or on the order of a physician.

Images provided courtesy of Dr. Pushpak Taunk.

All trademarks are the property of their respective owners.

2022 Copyright © Boston Scientific Corporation or its affiliates. All rights reserved.

ENDO-1474810-AA