



Biliary Schwannoma in a Post-Liver Transplant Patient Presenting as Biliary Obstruction

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Introduction

- Schwannomas are benign, mesenchymal cell-derived neoplasms which present as solid, slow growing tumors
- They are most commonly observed in the head, neck, spinal cord, and extremities
- Schwannomas have been reported in the gastrointestinal tract, with rare cases reported in the biliary system
- We present a unique case of a bile duct schwannoma in a post-liver transplant patient

Case Presentation

A 77-year-old man with a history of alcohol-related cirrhosis and hepatocellular carcinoma treated with orthotopic liver transplantation (16 years prior) presented with epigastric tenderness and jaundice.

Labs

AST (U/L)	ALT (U/L)	Alkaline Phosphatase (U/L)	Total Bilirubin (mg/dL)	Direct Bilirubin (mg/dL)
181	189	625	7.9	5.0

Diagnostic work-up

- MRCP: Enhancing mass with stricture in CBD (Figure A)
- EUS: Fine needle biopsy of mass in the mid CBD
- ERCP: Severe biliary stricture seen at the level of biliary anastomosis. Treated with balloon dilation and placement of a covered metal stent

Pathology

- Low-grade spindle cell tumor
- Staining positive for S100 and negative for CD117 and desmin (Figure B and C)

MRCP and Pathology

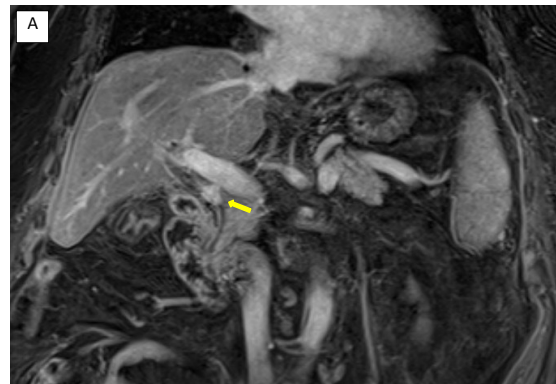


Figure A. MRCP T1-weighted coronal image demonstrating a 16 x 15 x 11 mm enhancing mass with diffusion restriction (yellow arrow) at the mid common bile duct causing an abrupt biliary stricture with upstream dilation

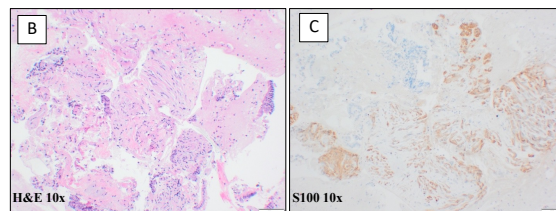


Figure B. Spindled, wavy nuclei in a fibroblastic stroma consistent with a spindle cell neoplasm in a background of unremarkable columnar biliary epithelium

Figure C. Diffuse nuclear and granular cytoplasmic positivity consistent with nerve sheath origin

Hospital Course

- Complicated by post-sphincterotomy bleed and post-ERCP pancreatitis
- Liver function continued to deteriorate despite stent placement, with patient's total bilirubin rising to 23.3 mg/dL
- Surgical intervention for schwannoma was deferred due to patient's tenuous clinical status
- Patient passed away shortly after transition to comfort care

Discussion

- The biliary tract is innervated by dense networks of extrinsic and intrinsic nerves found in the nonepithelial layer. Although schwannomas of the biliary tract are extremely rare, it is thought to arise from these peripheral nerves
- The clinical presentation of a biliary schwannoma is similar to cholangiocarcinoma, hence, tissue diagnosis is required
- Pathology typically shows highly cellular spindle cells
 - Stains positive for:
 - Vimentin – nonspecific marker of mesenchymal origin
 - S100 – marker of nerve sheath and melanocytic origin
 - Stains negative for:
 - CD117 – proto-onco gene activated in gastrointestinal stromal tumors
 - Desmin – marker of myogenic differentiation
- Resection is the treatment of choice

References

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