

A Unique Case of Poorly Differentiated Metastatic Signet Ring Cell Carcinoma of the Ampulla of Vater Presenting With Obstructive Jaundice With Non-Dilated Biliary Tree

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Introduction

- Ampullary cancers are rare and account for 0.2% of all gastrointestinal malignancies (1).
- Signet ring cell carcinoma (SRCC), a histological type of adenocarcinoma, is highly malignant and associated with poor outcomes.
- This case involves Ampullary SRCC which presented with fulminant liver failure; interestingly, there was obstructive cholestasis with no common bile duct (CBD) dilation.

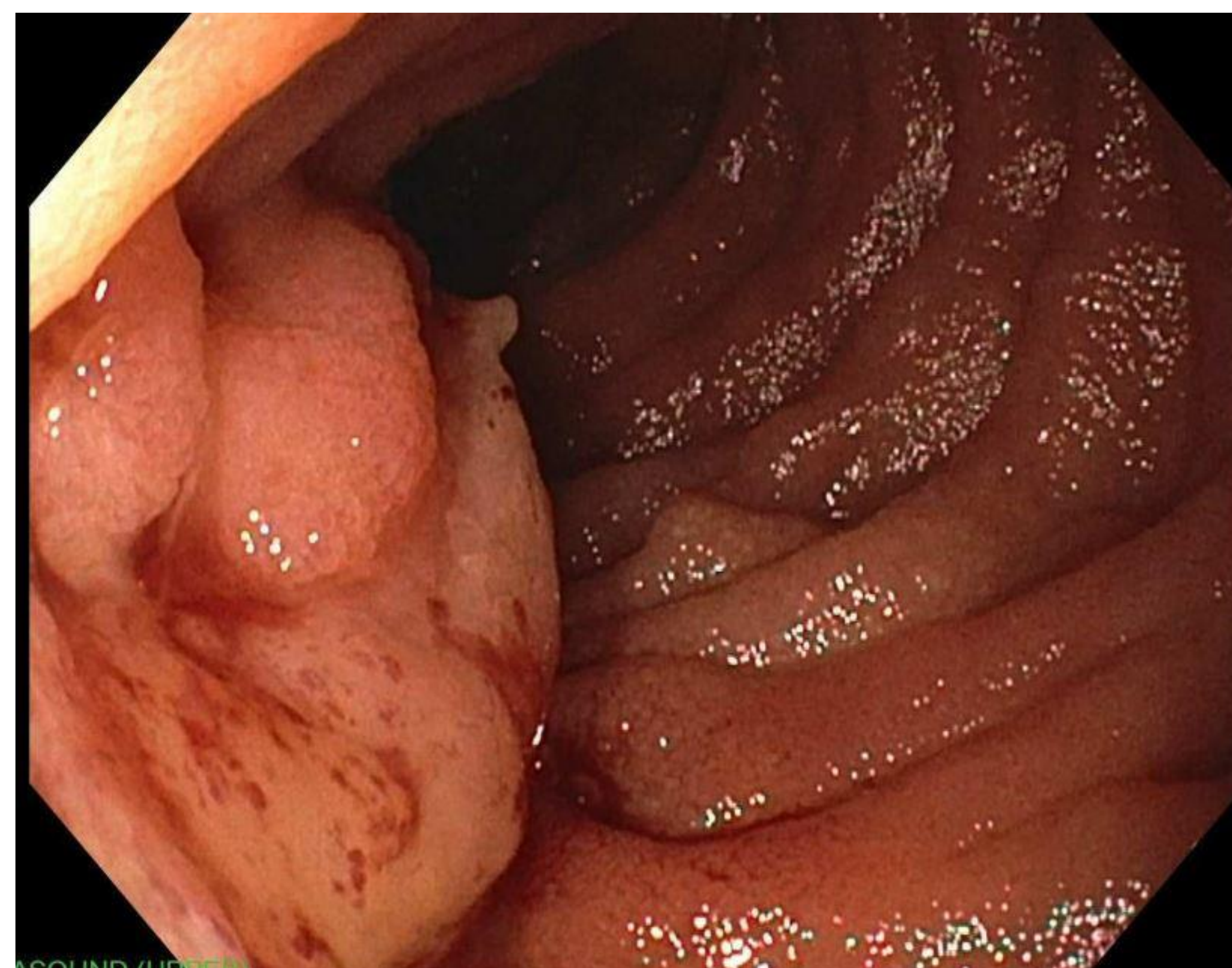
Case Presentation

- 65 year-old male with a past medical history of morbid obesity, diabetes, atrial fibrillation & heart failure.
- Presented with a one-month history of dull abdominal pain, weight loss (30lbs over 6 months) and jaundice.
- Examination: nontender abdomen without palpable mass.
- Lab tests showed a cholestatic pattern (Table 1). Anti-mitochondrial & anti-smooth muscle antibodies negative. Hepatitis B, C, EBV & CMV negative.
- Abdominal ultrasound and MRCP was negative for cholelithiasis & CBD dilation. CT imaging raised suspicion for malignancy due to extensive periportal and retroperitoneal lymphadenopathy. Liver biopsy significant for hepatitis, bile stasis & sinusoidal congestion; no neoplasia or fibrosis.
- Cervical lymph node biopsy revealed poorly differentiated adenocarcinoma with signet ring features (CDX2 and CK20 positive) in keeping with intestinal type malignancy.
- Esophagogastroduodenoscopy (EGD) revealed a 2 cm ampullary mass partially obstructing the major papilla (Image 1). Biopsy positive for SRCC.
- ERCP showed strictures in the proximal CBD with marked ductopenia of intrahepatic ducts with pruning.
- Hospital stay was complicated by fulminant liver failure, renal failure and death within one month of admission.

Table 1: Lab Investigations

Time	AST (U/L)	ALT (U/L)	ALP (U/L)	Total Bili (mg/dL)	Direct Bili (mg/dL)	Lipase (U/L)
1 month pre-admit	12	18	33	0.5		
On admission	249	318	610	8.2	6.6	95
2 weeks into admission	140	219	817	19.7	14.5	

Image 1: EGD displaying 2 cm mass partially obstructing the major papilla in the duodenum.



Discussion

- SRCC of the ampulla is a very aggressive neoplasm and scarcely described in the literature with only 81 cases documented (1).
- Signet ring cells are identified by the presence of vacuoles with high mucin content, staining PAS positive.
- Our case is unique as the patient presented with stage IV metastatic disease which is rare during the initial presentation (11% of cases) (1).
- Additionally, despite the ampullary mass and lab evidence of cholestasis, MRCP showed no signs of biliary dilatation – eventually found to be related to proximal CBD strictures and ductopenia on ERCP.
- Paraneoplastic ductopenia is well established in Hodgkin's Lymphoma (2); However, there are no cases to date with SRCC associated ductopenia. Other potential causes of ductopenia were ruled out and an idiopathic etiology would be a great coincidence in setting of metastatic disease.
- The median survival time for SRCC is 17 months versus other adenocarcinomas which is 25 months (1).
- Definitive treatment is pancreatoduodenectomy with reconstruction. However, this patient was not fit for surgery.
- Key learning point is to consider intrahepatic ductopenia & CBD strictures in advanced SRCC (& other gastrointestinal malignancies) which present with cholestasis with absence of CBD dilation.

References

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