

Pancreatic intraductal papillary mucinous neoplasm masquerading as ampullary adenoma: a diagnostic puzzle

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Abstract

Intraductal papillary mucinous neoplasm (IPMN) is the most common pancreatic cystic lesion, remaining mostly asymptomatic. An atypical presentation of such a lesion, initially thought to be an ampullary adenoma, is presented herein. A 78-year-old white male with painless jaundice was treated in a tertiary hospital. Imaging and endoscopic investigations pointed towards an ampullary adenoma obstructing and causing dilatation of both bile and pancreatic ducts. Endoscopic papillectomy was carried out and histology revealed tubulovillous adenoma. Follow-up duodenoscopy 3 months later showed a recurrent lesion with mucous leaking from the pancreatic duct. Cytology revealed mucin-rich atypical cells, consistent with main-duct IPMN. Pancreatoduodenectomy was performed, finally revealing main-duct IPMN protruding through Vater's ampulla. Cystic pancreatic lesions are increasingly found and IPMN is the most common of these. On the other hand, the management of ampullary adenomas has been revolutionized by endoscopic treatment and the advent of endoscopic papillectomy, with expanding indications. Meticulous clinical and imaging work up of these patients is essential to avoid suboptimal treatment. IPMN should be included in the differential diagnosis of ampullary adenomas, especially in the presence of a grossly dilated pancreatic duct.

Keywords Cystic pancreatic lesion, intraductal papillary mucinous neoplasm, ampullary adenoma, endoscopic papillectomy

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Introduction

Intraductal papillary mucinous neoplasm (IPMN) of the pancreas is the most common cystic lesion of this gland, accounting for 20% of all cystic pancreatic tumors [1]. IPMNs derive from mucin-secreting cells in the main pancreatic duct or its branches, and are categorized accordingly as main-duct, branch-duct or mixed-type IPMN. These lesions possess malignant potential and are associated with pancreatic duct dilatation [2]. Clinical manifestations of jaundice or abdominal pain may trigger investigations leading to this diagnosis; however, IPMNs remain mostly asymptomatic. A rare case of an IPMN protruding through Vater's ampulla, endoscopically resembling an ampullary adenoma and initially treated as such,

is presented herein, along with the diagnostic dilemmas and puzzlement of this bizarre clinical scenario. To the best of our knowledge, there is no other case of an IPMN presenting as a large Vater's ampulla mass lesion, causing pancreatic duct dilatation across its length as well as jaundice with bile duct dilatation.

Case report

A 78-year-old white male was investigated for painless jaundice, with a total bilirubin level of 6 mg/dL on initial presentation. His personal medical history was clear. Abdominal ultrasound scan showed a dilated common bile duct (CBD) up to 20 mm, with concomitant dilatation of the intrahepatic bile ducts and a normal gallbladder without gallstones. Further imaging with magnetic resonance cholangiopancreatography (MRCP) revealed a dilated CBD up to 20 mm, while the pancreatic duct measured 10 mm (Fig. 1). Endoscopy followed and an ampullary lesion was found, giving the endoscopic impression of an ampullary adenoma measuring about 3 cm in size (Fig. 2). Endoscopic retrograde cholangiopancreatography (ERCP) was performed, with findings similar to those of MRCP and without evidence

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Conflict of Interest: None

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of intraductal extension. Endoscopic papillectomy was carried out, along with endoscopic sphincterotomy (Fig. 3). Histopathological examination of the specimen showed an ampullary tubulovillous adenoma.

Three months later, on scheduled follow-up duodenoscopy, a recurrent lesion was revealed of similar texture, initially considered as a recurrent ampullary adenoma. Interestingly, at the time mucinous fluid was noticed leaking from the pancreatic duct and was sampled for cytology, which showed mucin-rich atypical cells, consistent with main-duct IPMN (Fig. 4).

The patient underwent a pylorus-preserving pancreaticoduodenectomy. His postoperative course was uneventful and the patient was discharged on the 10th postoperative day. Final histopathology revealed main-duct IPMN with high-grade dysplasia, protruding into Vater's ampulla, mimicking a recurrent ampullary adenoma. The patient remains disease free one year later on follow up.

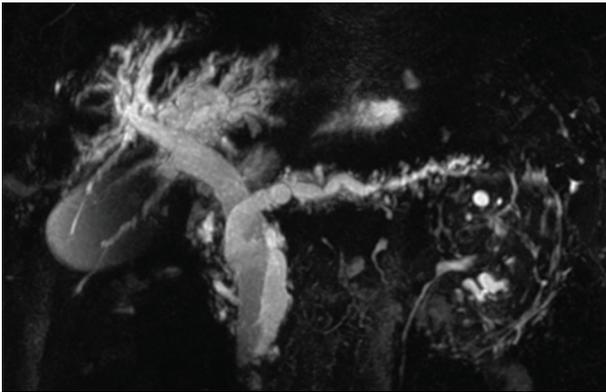


Figure 1 Magnetic resonance cholangiopancreatography showing dilatation of both pancreatic and common bile ducts

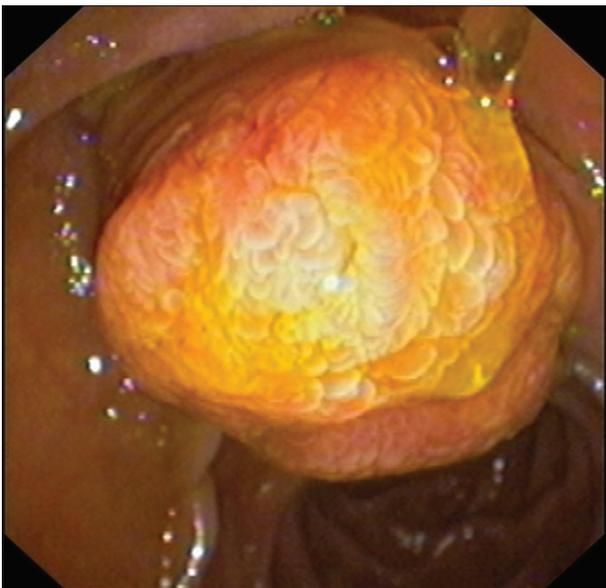


Figure 2 Duodenoscopy revealing an ampullary adenoma with max diameter about 3 cm

Discussion

Cystic lesions of the pancreas are increasingly found on imaging investigations, mostly on magnetic resonance scans [3]. Most of these cysts are asymptomatic; however, symptoms such as pancreatitis episodes, abdominal pain, nausea, or vomiting and jaundice may arise according to the cyst's location, size and nature [4]. The fundamental question regarding the clinical management of a pancreatic cystic lesion lies in the malignant potential associated with this cyst. Several reviews and guidelines exist on the appropriate work-up and management of pancreatic cysts [5].

On the other hand, the management of ampullary adenomas has been revolutionized by endoscopic treatment and management, with the advent of endoscopic papillectomy

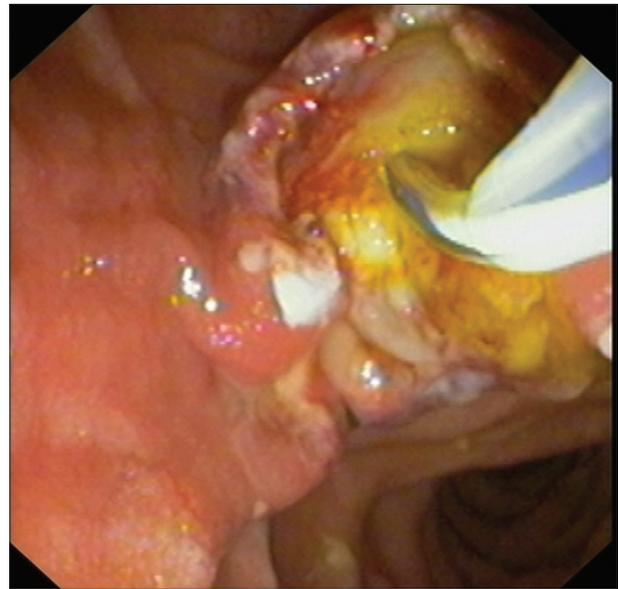


Figure 3 Endoscopic view post endoscopic papillectomy

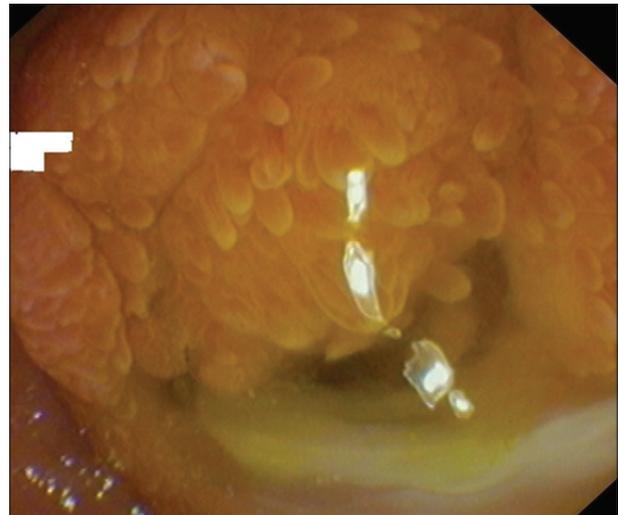


Figure 4 Recurrent ampullary lesion with mucinous fluid leaking from the pancreatic duct

(EP), and advances in endoscopic ultrasound (EUS) and ERCP [6]. Treatment modalities for ampullary tumors have included pancreatoduodenectomy and transduodenal excision, which are surgical procedures, and endoscopic papillectomy [7,8]. Ampullary adenomas seem to follow the well-established adenoma-to-carcinoma sequence; they are thus considered premalignant and warrant endoscopic or surgical resection [8]. The indications for EP are under constant review and expanding, while size larger than 5 cm, ulceration, friability or spontaneous bleeding are contraindications for EP. Intraductal extension is another contraindication for EP and can be assessed by EUS or ERCP. EUS performs similarly to ERCP in evaluating intraductal extension [8,9].

In the case presented above, obstructive jaundice and the endoscopic image of an ampullary mass suggested the diagnosis of ampullary adenoma. The dilatation of both bile and pancreatic ducts was attributed to obstruction from the adenoma. MRCP and ERCP did not show any intraductal extension and despite the large size, initial treatment with *en bloc* EP was deemed reasonable. On recurrence though, this study emphasizes the need for further sampling and reevaluation of the patient. A rare pathology, an IPMN protruding through Vater's ampulla, was actually the underlying lesion, but was not accurately diagnosed in the first instance, even histopathologically. The latter cannot easily differentiate between an ampullary tubulovillous adenoma and a papillary mucinous tumor, without the relevant clinical suspicion. In conclusion, IPMN should be included in the

differential diagnosis for endoscopically detected ampullary adenomas, especially in the presence of a grossly dilated pancreatic duct.

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