Biliary Adenomyomatosis Mimicking Periampular Neoplasia: A Case Report

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Abstract

Adenomyomatosis is a rare benign condition, typically found in the gallbladder. When occurring in the ampulla of Vater or common bile duct, it may mimic malignant tumors, making diagnosis and management challenging. We report the case of a 52-year-old woman presenting with cholestatic symptoms and significant weight loss over two months. Imaging revealed a solid, expansive lesion in the pancreatic head, involving the distal common bile duct and causing upstream bile duct dilation, prompting pancreaticoduodenectomy. Histopathology confirmed distal bile duct adenomyomatosis. Although benign, adenomyomatosis can simulate malignant characteristics and poses a significant diagnostic challenge, often leading to unnecessary surgery. Awareness of this condition is crucial to avoid unnecessary interventions and reduce associated risks.

Keywords: Bile Duct Neoplasms; Adenomyoma; Ampulla of Vater; Bile Ducts; Brazil

Introduction

Adenomyomatosis is an extremely rare benign condition that can occur throughout the gastrointestinal tract; its most common location is the gallbladder. Isolated cases have been reported involving other parts of the digestive system, including the bile ducts, ampulla of Vater, stomach, and small intestine.[1,5] Although histologically benign, adenomyomatosis can mimic malignant behavior when it affects the Vater system (ampulla and main bile duct), as it obstructs the bile duct.

Clinical presentation depends on the location of the lesion. While many patients remain asymptomatic, those with ampullary involvement may present with cholestasis, jaundice, or abdominal pain.[1,2] Diagnostic evaluation generally includes imaging methods such as computed tomography (CT), magnetic resonance imaging (MRI), endoscopic retrograde cholangiopancreatography (ERCP), and upper gastrointestinal endoscopy.[2,3]

ERCP is a key diagnostic tool, as it provides direct visualization of the periampullary area and the bile and pancreatic ducts, and enables access for biopsies. Although adenomyoma may present as an ampullary mass or bulging during endoscopy, distinguishing it from other ampullary tumors using ERCP remains difficult. Biopsy is often necessary, but obtaining one can be challenging when the adenomyoma does not protrude into the duodenum or when it is small and located in the submucosal

or muscular layers of the Vater system without ulceration. Consequently, the diagnostic yield of biopsies is typically low.[3,5]

Endoscopic ultrasound with fine-needle aspiration (EUS-FNA) offers improved diagnostic accuracy for ampullary and distal common bile duct masses, with reported sensitivity, specificity, and predictive values all approaching 100%. However, since most pathologists have limited experience with frozen section adenomyomas, achieving a definitive diagnosis is difficult in practice.[2,3]

Radiologically, these lesions may closely resemble cholangiocarcinoma, necessitating histological examination of the surgical specimen for diagnostic confirmation. This diagnostic uncertainty may lead to extensive surgical interventions, such as pancreaticoduodenectomy, increasing morbidity and mortality risks.[1,3]

Case Report

A 52-year-old woman with a history of hypertension and kidney transplant 11 years prior, presented with progressive jaundice, pruritus, choluria, and weight loss of about 6 kg over two months.

On examination, the abdomen was soft with mild tenderness in the right upper quadrant, a negative Murphy's sign, and no visceromegaly. Laboratory tests showed hyperbilirubinemia (mostly due to direct bilirubin), significantly elevated canalicular enzymes, a slight elevation in hepatic transaminases, and markedly elevated tumor marker [Table 1].

Table 1: Complete blood count and biochemical profile.

Parameter	Value	Reference
Hemoglobin, g/dL	8.4	12.0–16.0
White blood cells, µ/L	8000	4500–11,000
Platelets, µ/L	494,000	140,000–500,000
Creatinine, mg/dL	1.05	0.50-1.2
Urea, mg/dL	74	20–43
Total bilirubin, mg/dL	7.2	0.2–1.2
Conjugated bilirubin, mg/dL	5.1	0.0–0.5
Alkaline phosphatase, U/L	364	40–150
Gamma-glutamyl transferase (GGT), U/L	433	9–36
CA 19.9, U/mL	354	0–37
Aspartate aminotransferase (AST), U/L	97	5–34
Alanine aminotransferase (ALT), U/L	78	0–55

Note. *CA 19.9: Carbohydrate Antigen 19-9 tumor marker*.

A CT scan of the abdomen and pelvis showed a solid, expansive contrast-enhancing lesion in the pancreatic head, measuring approximately 4.5 × 2.5 × 3.0 cm, encompassing the distal common bile duct with abrupt narrowing and upstream bile duct dilation [Figure 1].

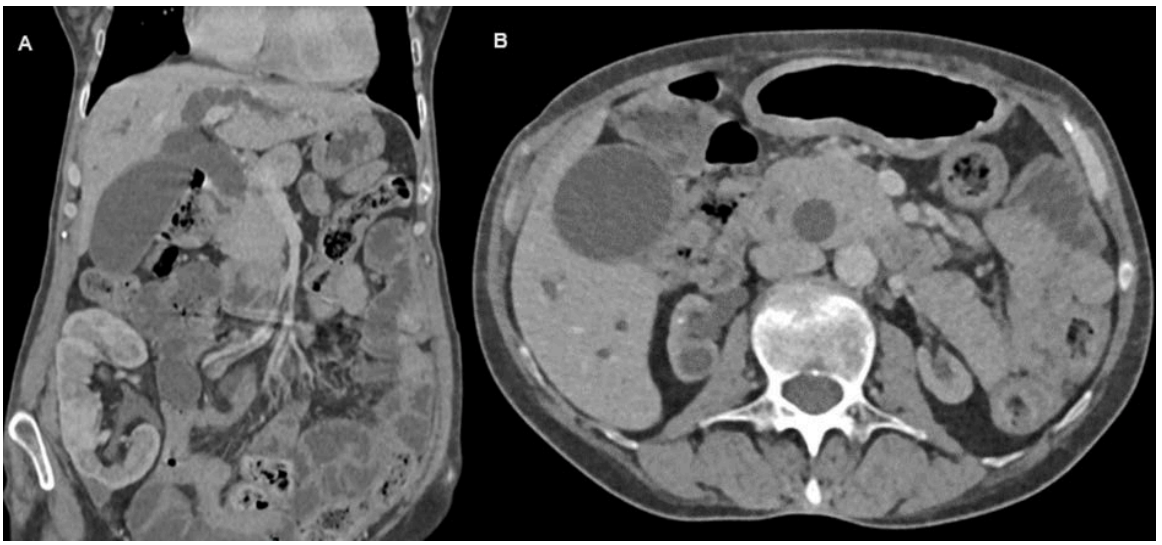

Figure 1: Abdominal computed tomography image of the abdomen showing a solid, contrast-enhancing lesion in the pancreatic head involving the distal common bile duct. A: Coronal view. B: Transverse view.

A subsequent contrast-enhanced magnetic resonance imaging (MRI) with MR cholangiography revealed a 4.3 cm lesion in close contact with the distal portion of the common bile duct, with diffusion restriction and abrupt narrowing of the duct, with intra- and extrahepatic bile duct dilatation upstream, suggestive of neoplasia. An endoscopic ultrasound (EUS) was not performed due to unavailability of the service.

A multidisciplinary team reviewed the case. Given the imaging findings, significantly elevated CA 19.9, and the patient's post-transplant status (which increases the risk of malignancy), the decision was made to proceed with surgical exploration.

The patient underwent a pancreatoduodenectomy. Intraoperatively, a firm 3 cm tumor was noted in the head of the pancreas, without invasion or contact with the superior mesenteric vein. Nonspecific lymphadenopathy was observed in the superior mesenteric vein. The bile duct was significantly dilated (about 3 cm), containing clear bile. The pancreas was soft, with the duct measuring 5–6 mm. No ascites, peritoneal or hepatic implants, or other signs of involvement were identified. The liver showed signs of cholestasis. The excised surgical specimen consisted of a hardened 3 cm tumor in the pancreatic head, without invasion or contact with the superior mesenteric vein [Figures 2 and 3].

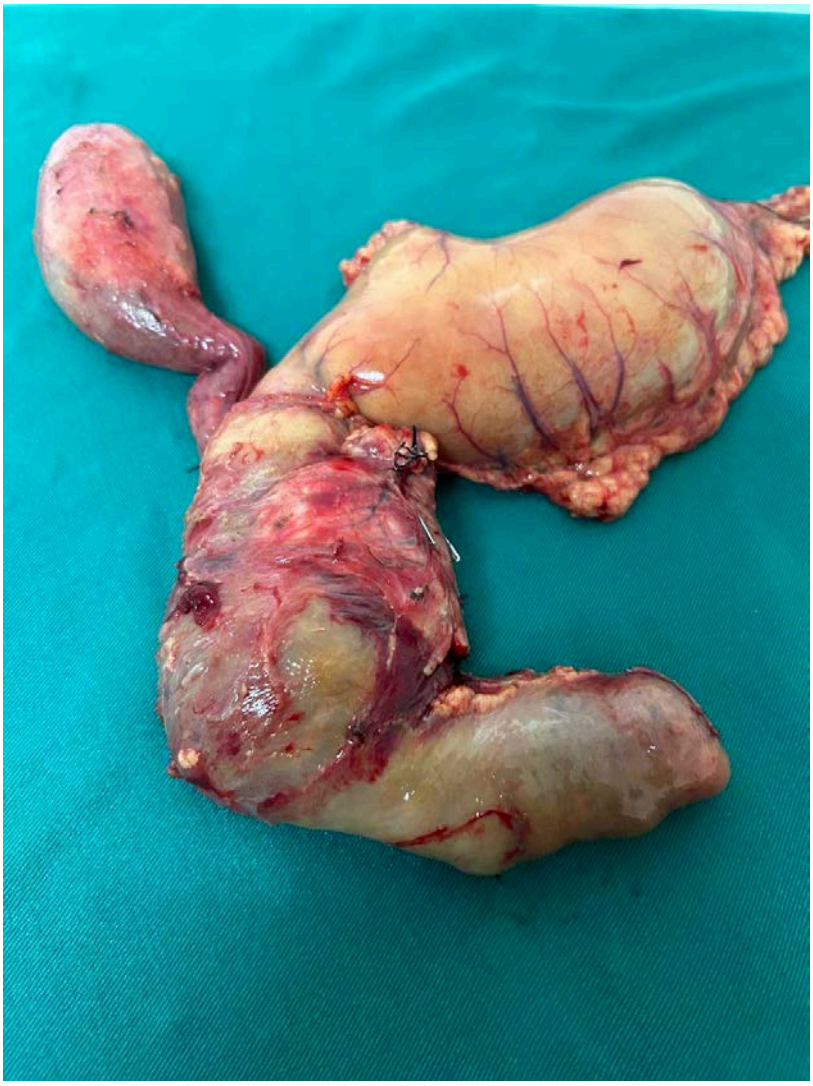

Figure 2: Surgical specimen from pancreatoduodenectomy.

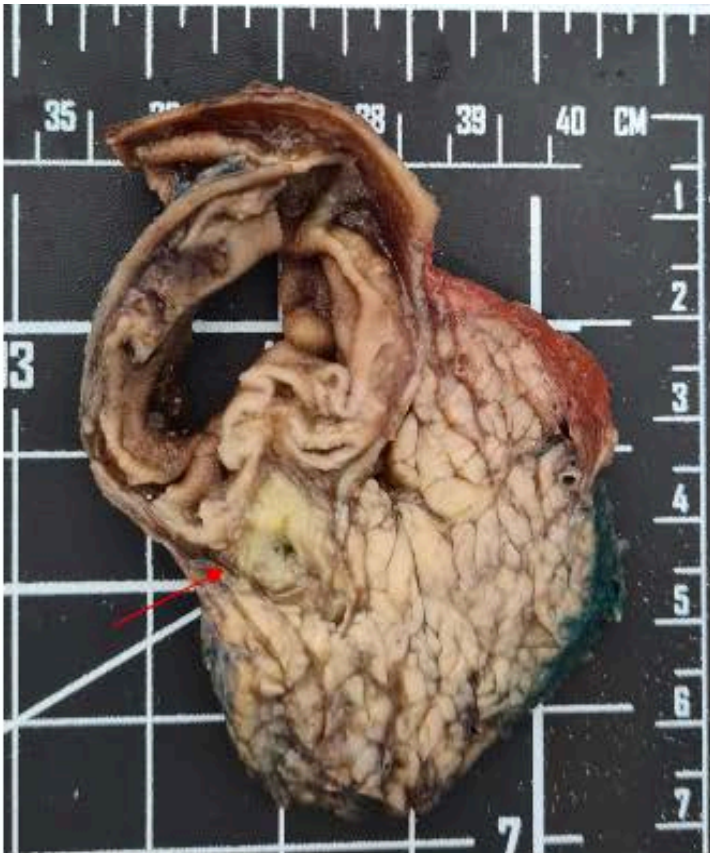

Figure 3: Duodenal segment with preserved folding. Ampulla of Vater with no apparent microscopic lesions. Sectioned pancreas showing intrapancreatic portions of the common bile duct with wall thickening (1.5 × 1.2 × 1.1 cm).

The patient remained stable in the immediate postoperative period in the intensive care unit (ICU). On the sixth postoperative day, her respiratory pattern worsened, with decrease in level of consciousness, requiring orotracheal intubation. Infectious screening revealed pulmonary consolidation suggestive of pneumonia. The patient developed septic shock of pulmonary origin, renal failure, and coagulopathy. Despite intensive support, she died from septic complications.

Histopathological analysis identified low-grade mucinous intraepithelial neoplasia in the pancreas with no invasive neoplasia after extensive sampling, as well as adenomyomatous hyperplasia of the distal bile duct with lymphoplasmacytic infiltrate and fibrosis [Figure 4].

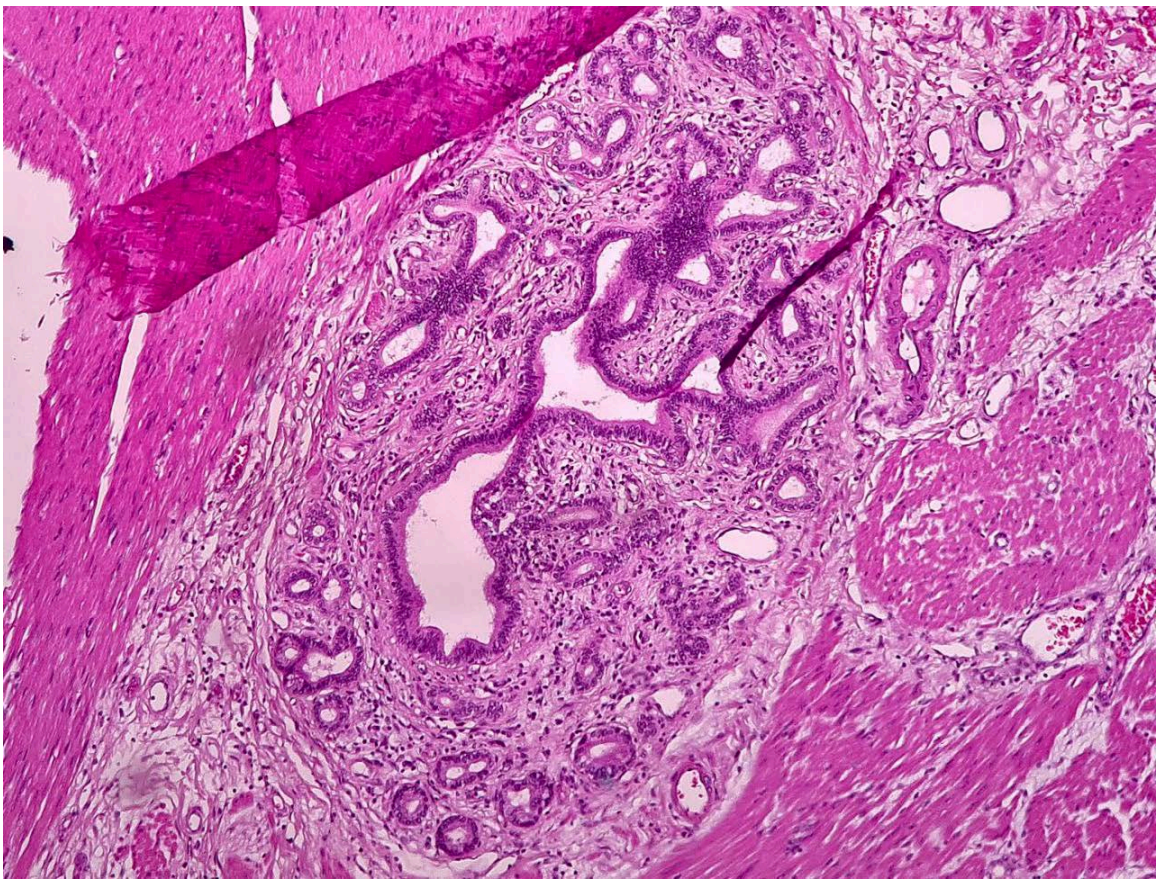

Figure 4: Histological slide of gastroduodenopancreatectomy specimen.

Discussion

The present case illustrates an extremely rare occurrence of adenomyoma in the ampulla of Vater, and underscores the difficulty of distinguishing benign from malignant lesions in the distal bile duct, particularly when imaging and laboratory findings are highly suggestive of cancer.

Despite uncertainty regarding whether the lesion was benign or malignant, factors including biliary obstruction, weight loss, and elevated CA 19-9 prompted surgical exploration and excision. Further, EUS-FNA, a key tool for assessing subepithelial or non-protruding lesions, was not available.

Histopathology ultimately revealed adenomyomatous hyperplasia with no invasive malignancy, showing that surgery was not called for. The patient's fatal postoperative course highlights the need for better preoperative differentiation to avoid unnecessary surgery.

This outcome reflects a broader challenge in clinical practice: rare benign lesions like adenomyomatosis can closely mimic malignancy. In post-transplant patients, such interventions carry added risks due to immunosuppression and comorbidities. However, if a satisfactory histological sample can be obtained through non-surgical methods, a conservative approach including clinical observation are possible.[1–3, 6]

Conclusion

Adenomyomatosis presents significant diagnostic challenges due to its potential to mimic malignant lesions in the bile ducts and ampulla of Vater. The tendency for over-treatment can lead to unnecessary surgical interventions, emphasizing the importance of awareness and accurate

diagnosis. A conservative approach may be indicated for asymptomatic cases. Further investigations are essential to refine the diagnostic criteria and treatment strategies for this rare condition.

Disclosure

The authors declare no conflicts of interest. Unfortunately, we could not contact the family in two years despite repeated attempts.

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