Biliary Adenomyomatosis Mimicking Periampular Neoplasia: A Rare Case Report

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Abstract

Adenomyomatosis is a rare benign condition, most commonly found in the gallbladder, although it has also been reported, even more rarely, in different sites throughout the gastrointestinal tract. When occurring in the ampulla and common bile duct, these lesions can lead to biliary obstruction and mimic the behavior of malignant tumors, making their diagnosis and management particularly challenging. We present the case of a 52-year-old female patient with cholestatic symptoms and a 6 kilograms weight loss over 2 months. A Computed Tomography (CT) scan of the abdomen and pelvis revealed a solid, expansive lesion in the pancreatic head, involving the distal common bile duct and causing upstream bile duct dilation. The patient underwent a pancreaticoduodenectomy, and histopathological analysis confirmed distal bile duct adenomyomatosis. Adenomyomatosis, although benign, is extremely rare in the bile ducts and ampulla of Vater. Its ability to simulate malignant characteristics poses a significant diagnostic challenge, often leading to extensive surgical procedures that can increase the risk of complications. Recognizing this condition is crucial to avoid unnecessary interventions and reduce associated risks, since the majority of these lesions present with biliary tract obstruction and mimic malignant behavior.

Keywords: Bile Duct Neoplasms; Adenomyoma; Bile Ducts.

Introduction

Adenomyomatosis is an extremely rare clinical condition that can occur in various locations within the gastrointestinal tract; however, its most common location is the gallbladder. Few cases have been described in the literature involving other sites within the digestive system, such as the bile ducts, the ampulla of Vater, stomach and small intestine.[1-5] Although considered histopathologically benign, adenomyomatosis can mimic the malignant behavior of a carcinoma when it affects the Vater system (ampulla and main bile duct), as it obstructs the bile duct. The clinical presentation can vary depending on the location of the lesion, with the patient potentially being asymptomatic or presenting with jaundice and abdominal pain when the tumor is located in the ampullary region.[1,2]

Diagnostic imaging methods that can be used include computed tomography, magnetic resonance imaging (MRI), endoscopic retrograde cholangiopancreatography (ERCP), and endoscopy.[2,3]

ERCP can be an essential diagnostic tool, allowing for direct visualization of the periampullary area and the bile and pancreatic ducts, and enabling biopsies for histological analysis. 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 required for confirmation, but obtaining a sample 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]

Another method available is EUS-FNA (Endoscopic Ultrasound - Fine Needle Aspiration), which can be performed on ampullary and distal common bile duct masses, offering an overall accuracy of 100%, along with sensitivity, specificity, and positive and negative predictive values all at 100%. However, since most pathologists have limited experience with frozen section adenomyomas, a definitive diagnosis may not be achievable.[2,3]

However, the lesion often manifests radiologically as a cholangiocarcinoma, necessitating histological examination of the surgical specimen for diagnostic confirmation. This can lead to extensive surgical operations, such as pancreaticoduodenectomy, which increases the patient's morbidity and mortality risk.[1-3]

Case Report

A 52-year-old female patient with hypertension, who had undergone a kidney transplant 11 years ago, developed progressive jaundice accompanied by pruritus, choluria, and an estimated weight loss of 6 kilograms over 2 months.

An abdominal examination revealed a soft abdomen, somewhat tender in the right upper quadrant, a negative Murphy's sign, and no visceromegaly. Laboratory tests showed hyperbilirubinemia, predominantly due to direct bilirubin, along with a significant increase in canalicular enzymes, a slight elevation in hepatic transaminases and elevated tumor marker [Table 1].

Table 1: Full blood workup.

Parameter	Value	Reference
Hemoglobin, g/dL	8,4	12,0 - 16,0
White blood cells, µ/L	8.000	4.500 - 11.000
Platelets, µ/L	494.000	140.000 - 500.000
Creatinine, mg/dL	1,05	0,50 - 1,20
Urea, mg/dL	74	20 - 43
Total bilirubin, mg/dL	7,19	0,20 - 1,20
Conjugated bilirubin, mg/dL	5,09	0,00 - 0,50
Alkaline phosphatase, U/L	364	40 - 150
Gamma-glutamyl transferase (GGT), U/L	433	9 -36
CA 19.9, U/mL	354	0 - 37
AST (Aspartate Aminotransferase), U/L	97	5 - 34
ALT (Alanine Aminotransferase), U/L	78	0 - 55

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

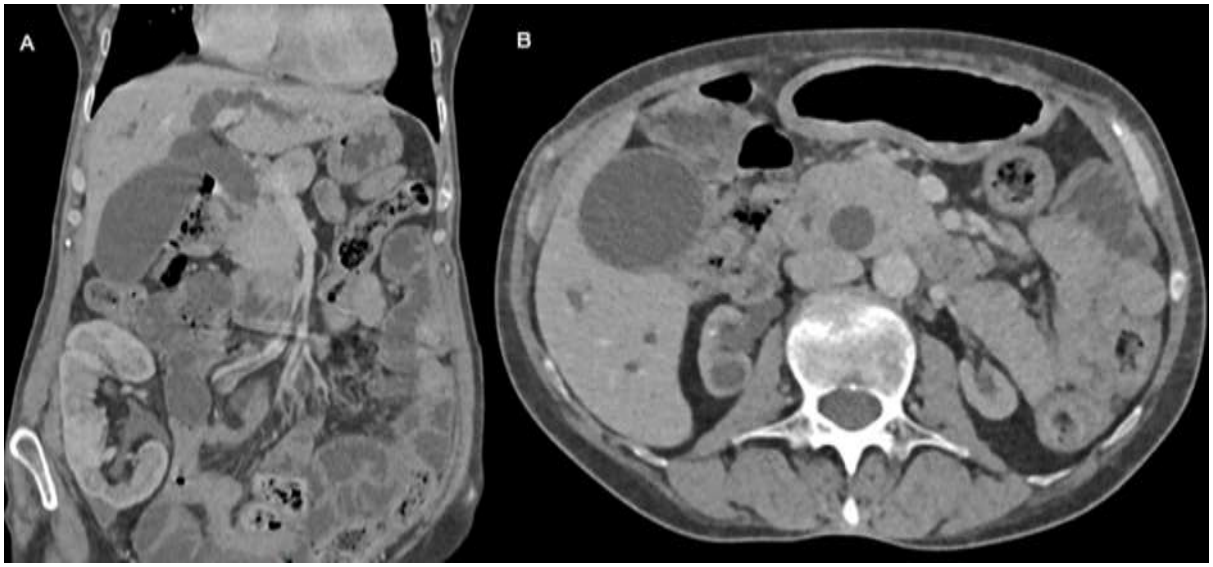

Figure 1: CT image of the abdomen showing a solid, expansive lesion with contrast enhancement in the pancreatic head, measuring approximately 4.5 x 2.5 x 3.0 cm, involving the distal common bile duct, with abrupt narrowing and upstream bile duct dilation. Image A: Coronal view. Image B: Transverse view.

An upper abdominal MRI with contrast and MR cholangiography was performed, revealing findings of diffusion restriction measuring 4.3 cm, in close contact with the distal portion of the common bile duct, causing abrupt narrowing of the duct, with intra- and extrahepatic bile duct dilatation upstream, suggestive of neoplasia. The EUS was not performed due to unavailability of the service.

The case was thoroughly reviewed in a multidisciplinary meeting, where the possibility of neoplasia, indicated by elevated CA 19.9 levels and imaging findings, was carefully considered. Given the patient's history of transplantation, which increases the risk of malignancy, alongside their favorable surgical status, the decision was made to proceed with surgery.

The patient underwent a pancreatoduodenectomy and developed hemodynamic instability, requiring the use of vasopressor medications and volume expansion, with a good response to the measures implemented. The surgical findings showed a hardened tumor about 3 cm 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) with clear bile. The pancreas had a softened consistency, and the pancreatic duct measured approximately 5-6 mm. The liver showed signs of cholestasis. No ascites, peritoneal or hepatic implants, or other signs of involvement were found. The histopathological analysis identified a hardened 3 cm tumor in the pancreatic head, without invasion or contact with the superior mesenteric vein. [Figures 2 and 3].

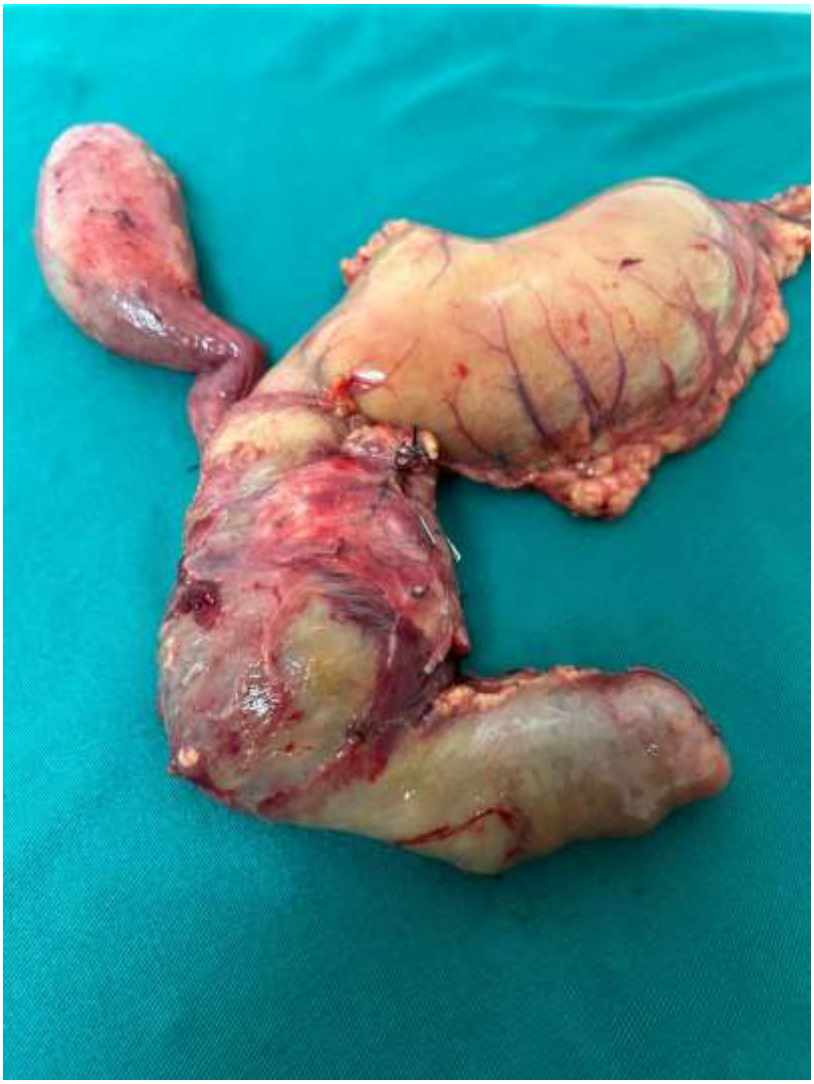

Figure 2: Surgical specimen of pancreatoduodenectomy.

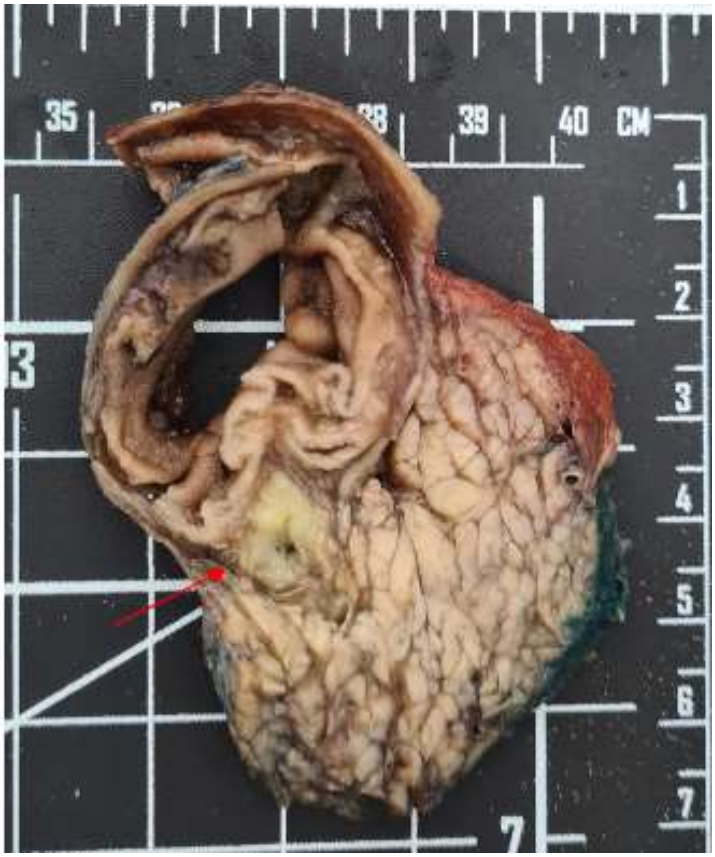

Figure 3: Duodenal segment with preserved folding. Ampulla of Vater with no apparent microscopic lesions. Segment of the pancreas already sectioned, showing intrapancreatic portions of the common bile duct with wall thickening (1.5 x 1.2 x 1.1 cm).

Histopathological analysis identified the presence of 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 associated with lymphoplasmacytic infiltrate and fibrosis [Figure 4].

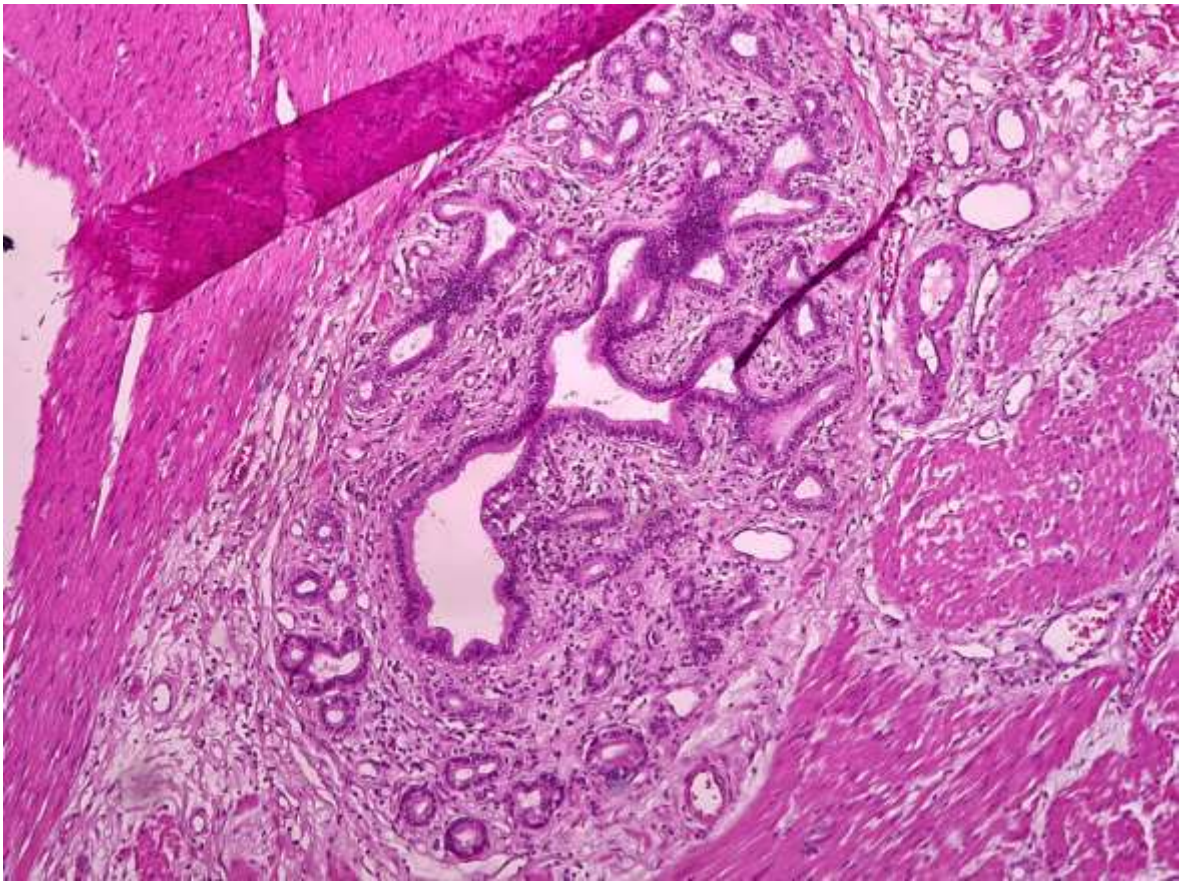

Figure 4: Histopathology slide of gastroduodenopancreatectomy specimen.

The patient remained stable in the immediate postoperative period in the Intensive Care Unit (ICU). On the sixth postoperative day, the patient experienced worsening of the respiratory pattern and a decrease in the level of consciousness, requiring orotracheal intubation. Infectious screening revealed pulmonary consolidation suggestive of pneumonia. Consequently, the patient developed septic shock of pulmonary origin in the postoperative period, along with renal failure, coagulopathy, and eventually, therapeutic failure was determined, leading to death.

Discussion

Adenomyoma is a term generally applied to nodular lesions showing proliferation of both epithelial and smooth muscle components.[6] It is a rare benign lesion of the gastrointestinal tract, most commonly found in the gallbladder. Few cases have been described in the literature involving other sites within the digestive system, such as the bile ducts and the ampulla of Vater.[1-5]

Patients may be asymptomatic or present with nonspecific symptoms, depending on the tumor's location. Clinical manifestations can include epigastric pain and signs of bile duct obstruction, such as cholestatic syndrome, necessitating differential diagnosis with malignant periampullary tumors.[1,2]

The diagnosis of adenomyoma in the common bile duct and ampulla of Vater presents significant challenges, as patients typically show symptoms of biliary obstruction and cholestasis, with imaging studies often revealing bile duct blockage or a mass.[1,3]

Endoscopic procedures, including biopsies, EUS, and brush cytology, commonly detect atypical cells or dysplasia. However, these findings are often a result of the endoscopic manipulation itself, complicating the diagnostic interpretation.[1,3]

Although ERCP may allow direct visualization of the periampullary area, confirmation of a definitive diagnosis before surgery remains difficult due to the limited diagnostic yield of biopsies and the inability to differentiate between benign and malignant lesions.[4,5]

The management of this condition is not consensual in the literature, as obtaining a preoperative histological diagnosis is challenging, often leading to patients undergoing surgical approaches with the risk of complications such as pancreatic fistula and delayed gastric emptying. However, given a satisfactory histological sample obtained through non-surgical methods, a conservative approach and clinical observation are possible.[1-3,6]

Patients with lesions in the biliary tract, depending on the size, location, and preoperative/intraoperative diagnosis, have three treatment options. These include local surgical resection or pancreaticoduodenectomy, which is the traditional surgical approach. An alternative option is endoscopic mucosal resection, followed by awaiting the pathology report.[1-,3,5]

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 accurate diagnosis and awareness among healthcare providers. A conservative approach may be indicated for asymptomatic cases, and less invasive treatment options should be considered. Further investigation is essential to refine diagnostic criteria and treatment strategies for this rare condition.

Disclosure

The authors declared no conflicts of interest. Unfortunately, we couldn't contact the family in 2 years with repeated trial of contact. All communications made did not elicit any response or return from the family.

References

1. Gouveia C, Fidalgoa C, Loureiro R, et al. Adenomyomatosis of the Common Bile Duct and Ampulla of Vater. GE Port J Gastroenterol 2021;28:121-133.

2. Ramos-Muñoz F, Hinojosa-Arco LF, Jorge Francisco Roldán-de la Rúa JF, García-Salguero AI, Suárez-Muñoz MA. Obstrucción biliar por adenomioma de la ampolla de Vater. Gastroenterol Hepatol 2022;45:69-70.

3. Frutuoso L, Pereira AM, Carvalho L, Gonçalves G, Nora M. Adenomyomatous Hyperplasia of Ampulla of Vater and a Concomitant Renal Tumor: A Case Report. Cureus 2021;13(12):e20258.

4. Choi YH, Kim MJ, Han JH, et al. Clinical, pathological, and immunohistochemical features of adenomyoma in the ampulla of vater. Korean J Gastroenterol 2013;62(6):352-358.

5. Kwon HJ, Kim SG, Park J. Adenomyoma of the ampulla of Vater mimicking malignancy: A case report and literature review. Medicine (Baltimore). 2023 16;102(24):e34080.

6. Matsumoto K, Kato H, Nishida K, Okada H. Adenomyomatosis hyperplasia arising in the bile duct. Dig Liver Dis 2019;51(7):1060.

7. Park Y, Lee J, Kim Y, Cho BS, Park KS, Woo CG. Differentiation of Adenomyoma from Localized Adenocarcinoma of the Ampulla of Vater Using Multidetector CT. Taehan Yongsang Uihakhoe Chi. 2021;82(2):393-405.

8. Byung-Uk LEE; Jei-So BANG; Soo-Hyun YANG; et al. A Case of Ampullary Adenomyoma Associated with Dilatations of Pancreatic and Biliary Ducts. Korean Journal of Gastrointestinal Endoscopy. 2010; 391-395.

9. Kwon TH, Park DH, Shim KY, Cho HD, Park JH, Lee SH, et al. Ampullary adenomyoma presenting as acute recurrent pancreatitis. World J Gastroenterol 2007;13(20):2892-2894.