Adenomyomatous Hyperplasia of Ampulla of Vater and a Concomitant Renal Tumor: A Case Report

Luisa Frutuoso 1, Ana Marta Pereira 1, Lucia Carvalho 1, Gil Gonçalves 2, Mário Nora 3

1. General Surgery, Centro Hospitalar de Entre Douro e Vouga, Santa Maria da Feira, PRT
2. General Surgery, Champalimaud Foundation, Lisbon, PRT
3. General Surgery, Centro Hospitalar Entre o Douro e Vouga, Santa Maria da Feira, PRT

Corresponding author: Luisa Frutuoso, luisafrutuoso04@gmail.com

Abstract

Adenomyomatous hyperplasia is an extremely rare lesion encountered in the ampulla of Vater. Less than 50 cases have been described, most of them with clinical consequences of biliary obstruction, misdiagnosing it as a malignancy. The authors present a concomitant case with a renal tumor, its diagnosis, management, and clinical relevance, as well as a brief revision of the literature. Ampullar and renal tumors were found in a 74-year-old female, in the imagiologic study of a low back pain, in the emergency department. Both were considered malign after further study, and pancreatoduodenectomy with partial nephrectomy was proposed. There is no accurate diagnostic tool to differentiate the benign nature of adenomyomatous hyperplasia and extensive operations are often performed. As an incidental finding in the study of another tumor, this case raises the concern about which and how to treat both tumors, taking into account the morbidity of the respective interventions.

Categories: Pathology, Urology, General Surgery

Keywords: pancreas, pancreatoduodenectomy, renal neoplasms, ampulla of vater, adenomyomatous hyperplasia

Introduction

Benign tumors constitute 6% of all extrahepatic biliary tree neoplasms and are responsible for 0.1% of all biliary tract operations [1]. Adenomyomatous hyperplasia (AH) or adenomyomiosis can occur anywhere in the gastrointestinal tract. It is an extremely rare benign lesion of the extrahepatic biliary tree, more often encountered in the gallbladder, but only a few cases have been described in the ampulla of Vater [2-4].

The histopathogenesis and natural history of these lesions in the common bile duct (CBD), including the ampulla of Vater, is not clear and malignant transformation in other organs as well as recurrence after local resection has been described [1,5]. Diagnostic tools, including endoscopy with ultrasonography and biopsy, are frequently not accurate enough to rule out a malignancy, and these diagnostic difficulties have led to extensive operations. The appropriate management of these tumors is not established.

Unlike the other counterparts in the gastrointestinal tract, the ampulla of Vater’s tumors is responsible for the biliary obstruction, which leads to significant clinical consequences. Jaundice and abdominal pain are the main complaints of these patients although an incidental finding in the study of another neoplasm has been described as well [2]. The latter adds a dilemma about how and when to treat each tumor taking into account both are suspicious of malignancies.

We report a case of a patient AH of the ampulla of Vater and a concomitant renal tumor, its diagnosis, management, and clinical relevance, as well as a brief revision of literature.

Case Presentation

A 74-year-old female, with a past history of renal lithiasis and lumbar hernia, recurred to the emergency department (ED) complaining of acute low back and abdominal pain. An abdominopelvic CT scan in the ED revealed not only the main cause of the pain, calculous hydronephrosis but also two incidentally distinct lesions: a left renal tumor with malignant features and a 15-mm nodular lesion conditioning CBD and wirsung ectasia. No hepatic changes or cholestasis were observed in the laboratory study. Medical treatment for ureterolithiasis was done with complete symptomatic resolution.

Further study was done for both tumors. Laboratory tests showed normal hepatic and renal function, with no cholestasis. Serum carcinoembryonic carcinogen and carbohydrate antigen levels were not increased.

A magnetic resonance cholangiopancreatography (MRCP) confirmed bicanalicular dilation (CBD and wirsung) with abrupt stenosis at the ending of both channels, although no evident nodular lesion, as well the renal
tumor (Figure 1). Fluorodeoxyglucose positron emission tomography-computed tomography scan (PET-FDG/CT) was inconclusive.

FIGURE 1: MRCP showed bicanalar dilation with abrupt stenosis

MRCP - magnetic resonance cholangiopancreatography

Endoscopic ultrasound was attempted and no ampullar changes were demonstrated, but a cystic lesion was apparent at the terminus of the CBD (choledococele), so biopsies were not taken.

Three months later, an MRCP was repeated which revealed at this time an ampullar mass causing bicanalar obstruction, and so, a CBD malignancy could not be ruled out (Figure 2).
A multidisciplinary team discussed the case and the patient was proposed to pylorus-preserving pancreatoduodenectomy in combination with partial nephrectomy, in the same surgical time, to which the patient consented.

Macroscopic examination of the surgical specimen revealed a polypoid lesion in the ampullar region with 1.2 cm.

Pathologic examination demonstrated chronic polypoid fibrous ampullitis with glandular AH. No malignant cells were identified. The partial nephrectomy specimen revealed a clear cell renal carcinoma (pT1aNxVx).

The patient’s condition was complicated with postoperative intra-abdominal abscess followed by surgical drainage. Full recovery and discharge were possible one month after surgery.

Eight months later, the patient is doing well, with no complaints or recurrence of both lesions.

**Discussion**

AH is an extremely rare lesion encountered in the ampulla of Vater and can be misdiagnosed as an ampullar adenoma or carcinoma. To our knowledge, less than 50 cases have been described in the indexed English literature [6]. This is the second case in our institution [4].

It is difficult to determine the real incidence, but a frequency of 0.13% has been reported in a series of consecutive 3,131 endoscopic retrograde cholangiograms (four cases) [7]. Higher frequency was noted in a post-mortem specimens study, in which 54% of 100 unselected patients had this lesion, with no clinical significance [8]. Another interesting observation of this study was the presence of other concomitants, more important findings, apparently not correlated with the AH, such as thyroid adenomas, gastric, colonic and uterine polyps, and even adenocarcinomas of the stomach, colon, and rectum, although no constant association was observed. This could bring up the hypothesis of the AH as a sentinel lesion for other tumors, giving this case as an example, once the renal tumor was the main concern of this patient’s condition.

Histologically, AH is defined as a nodular lesion or a mucosal thickening with a proliferation of smooth muscle and epithelial components, as well as glandular hyperplasia without cellular atypia. Its histogenesis is not clear and many theories have been proposed. Incomplete heterotopic pancreas (type III according to Heinrich classification) seems to be the most widely accepted theory, although some authors defend that this lesion may have an inflammatory origin [9,10]. Baggenstoss also highlights the fact that the papillar region as a transitional epithelial zone, is susceptible to local irritation inducing polypoid hyperplasia [11]. This case represents a glandular AH associated with a chronic inflammatory polypoid ampullar lesion, which goes with this hypothesis.

The preoperative diagnose is challenging and imaging is not accurate enough to distinguish an AH from adenoma or carcinoma. A definitive diagnose is only possible with histological examination of the completed resected mass [2,12,13]. Preoperative biopsies are often useless once the absence of malignant features cannot exclude malignancy. In a series of 15 patients with AH of the ampulla of Vater, preoperative
Adenomyomatous hyperplasia of the ampulla of Vater is an extremely rare tumor of the biliary tree. Unlike in other organs, this location has important clinical consequences, being misdiagnosed as an adenoma or carcinoma in most of the cases. There is no accurate method to distinguish these lesions from an adenoma or carcinoma, and so, extensive surgery is frequently adopted.

Efforts must be done to improve the diagnostic tools. Cases with concomitant neoplasms are not negligible in the literature, with decision-making implications and increased surgical risk.

Conclusions
Adenomyomatous hyperplasia of the ampulla of Vater is an extremely rare tumor of the biliary tree. Unlike in other organs, this location has important clinical consequences, being misdiagnosed as an adenoma or carcinoma in most of the cases. There is no accurate method to distinguish these lesions from an adenoma or carcinoma, and so, extensive surgery is frequently adopted.

Efforts must be done to improve the diagnostic tools. Cases with concomitant neoplasms are not negligible in the literature, with decision-making implications and increased surgical risk.

References
1. Burhans R, Myers RT: Benign neoplasms of the extrahepatic biliary ducts. Am Surg. 1971, 37:161-6.
2. Handrz-Luca A, Terris B, Couvelard A, Bonte H, Flejou JF: Adenomyoma and adenomyomatous hyperplasia of the Vaterian system: clinical, pathological, and new immunohistochemical features of 13 cases. Mod Pathol. 2003, 16:530-6. 10.1097/01.MP.0000075752.71096.85
3. Choi YH, Kim MJ, Han JH, et al.: Clinical, pathological, and immunohistochemical features of adenomyoma in the ampulla of vater. Korean J Gastroenterol. 2013, 62:352-8. 10.4166/kgj.2013.62.6.352
4. Guimarães M, Rodrigues P, Gonçalves G, Oliveira V, Nora M, Monteiro MP: Vater papilla adenomyosis [Article in Portu- guese]. Revista Portuguesa de Cirurgia. 2014, 45-7.
5. Kneafsey PD, Demetrick DJ: Malignant transformation in a pyloric adenomyoma: a case report. Histopathology. 1992, 20:433-5. 10.1111/j.1365-2559.1992.tb01015.x
6. Gouveia C, Fidalgo C, Loureiro R, Oliveira H, Maio R, Cravo M: Adenomyomatosis of the common bile duct and ampulla of vater. GE Port J Gastroenterol. 2021, 28:121-33. 10.1119/000507788
7. Hammarström LE, Holmin T, Stenram U: Adenomyoma of the ampulla of Vater: A rare cause of bile duct obstruction. Surg Laparosc Endosc. 1997, 7:388-95.
8. Dardinski VJ: Inflammatory adenomatoid hyperplasia of the major duodenal papilla in man. Am J Pathol. 1931, 5:519-22.
9. Narita T, Yokoyama M: Adenomyomatous hyperplasia of the papilla of Vater: a sequel of chronic papillitis? Ann Diagn Pathol. 1999, 3:174-7. 10.1016/s1092-9154(99)00045-8
10. von Heinrich H: Ein Beitrag zur Histologie des sogen akzessorischen Pankreas. Virchows Archiv für pathologische Anatomie und Physiologie und für klinische Medizin. 1909, 198:392-401. 10.1007/BF02085527
11. Baggergård AH: Major duodenal papilla: variations of pathologic interest and lesions of mucosa. Arch Pathol. 1938, 26:855-68.
12. Kayahara M, Ohta T, Kitagawa H, Miwa K, Urabe T, Murata T: Adenomyomatosis of the papilla of Vater: a case illustrating diagnostic difficulties. Dig Surg. 2001, 18:139-42. 10.1159/000050115

13. Iwaki K, Shihata K, Ohta M, et al.: Adenomyomatous hyperplasia of the common bile duct: report of a case. Surg Today. 2008, 38:85-9. 10.1007/s00595-007-5558-9

14. Kwon TH, Park DH, Shim KY, et al.: Ampullary adenomyoma presenting as acute recurrent pancreatitis. World J Gastroenterol. 2007, 13:2892-4. 10.3748/wjg.v13.i20.2892

15. Ulich TR, Kollin M, Simmons GE, Wilczynski SP, Waxman K: Adenomyoma of the papilla of Vater. Arch Pathol Lab Med. 1987, 111:588-90.