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A Singular Case of Adenomyoma Ampulla with Parathyroid Adenoma – A Rarest of Rare Association

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Abstract

Adenomyoma is an uncommon benign lesion, rare in the extrahepatic biliary tree and ampulla of Vater, but relatively more common in the gallbladder. Preoperative diagnosis of ampullary adenomyoma is highly challenging, as it often mimics malignancy.

We present a case report of a 58-year-old gentleman who presented with abdominal pain and obstructive jaundice to our institution and underwent standard pancreaticoduodenectomy for suspected periampullary malignancy. Surprisingly, the biopsy revealed adenomyoma of the ampulla. Postoperatively, he developed symptoms of hypercalcemia, which, upon further evaluation, was due to a hyperfunctioning left parathyroid adenoma that was subsequently surgically removed.

The coexistence of adenomyoma of the ampulla and parathyroid adenoma is exceedingly rare and has not been reported in the literature. We present this unique case report to highlight this singular association, hoping that further research in the future may shed light on this phenomenon. Serum calcium levels should be assessed in all periampullary lesions, as this simple test could have identified the parathyroid adenoma preoperatively.

Keywords: Adenomyoma Ampulla, Periampullary Malignancy, Parathyroid Adenoma, Hypercalcemia, Case Report.

Main Manuscript Text

Introduction

Adenomyoma (adenomyomatous hyperplasia, adenomyomatosis or adenomyosis) is an uncommon hamartomatous benign lesion found throughout the gastrointestinal tract, more commonly in gallbladder and notably rare in common bile duct (CBD) and in ampulla of vater (AOV) [1]. Adenomyoma of the AOV is clinically significant as it causes obstructive jaundice, mimicking adenocarcinoma of the AOV or distal CBD. Despite being a benign lesion, it is mostly treated with major surgical resection with high morbidity, as preoperative diagnosis is extremely challenging.

We report a case of a 58-year-old gentleman with adenomyoma ampulla who was treated with standard pancreaticoduodenectomy (PD) without any major post operative surgical complications. Surprisingly, the patient developed postoperative complications from hypercalcemia, caused by a newly detected parathyroid adenoma. This case report highlights both the rarity and diagnostic challenge of adenomyoma of the AOV, as well as the fact that its co-existence with parathyroid adenoma has not yet been reported in the literature.

Case Presentation

A 58-year-old male patient with no comorbidities presented to our institution with complaints of upper abdominal pain for 2

months and yellowish discoloration of eyes for 2 weeks. He had no history of colicky flank pain, bony pain, polyuria, psychosis or any other neuro psychiatric symptoms. There was no family history of any malignancies. Laboratory workup showed a deranged obstructive pattern liver function test with total bilirubin of 3.4 mg/dl, direct bilirubin of 2.8 mg/dl, alkaline phosphatase (ALP) of 330 U/L and normal alanine transaminase (ALT) and aspartate transaminase (AST). Complete blood count was normal, with no elevation of acute phase reactants and a normal amylase level. Cancer antigen 19-9 (CA 19-9) and carcinoembryonic antigen (CEA) were normal. Abdominal contrast enhanced computed tomography (CECT) scan showed a heterogeneously enhancing soft tissue lesion of size 1.5 x 1.4 x 1.3 cm, located in the ampullary region with upstream dilation of CBD of 30 mm and intrahepatic biliary radicles dilation (IHBRD) and dilated main pancreatic duct of 4 mm (Figure 1)..

There was also an incidentally detected right kidney upper pole microlith. Upper gastrointestinal (GI) scopy revealed bulging of ampulla with normal mucosa (Figure 2). Biopsy was not performed since a negative or inconclusive biopsy would not change the treatment plan of the patient as suspicion of malignancy was high.

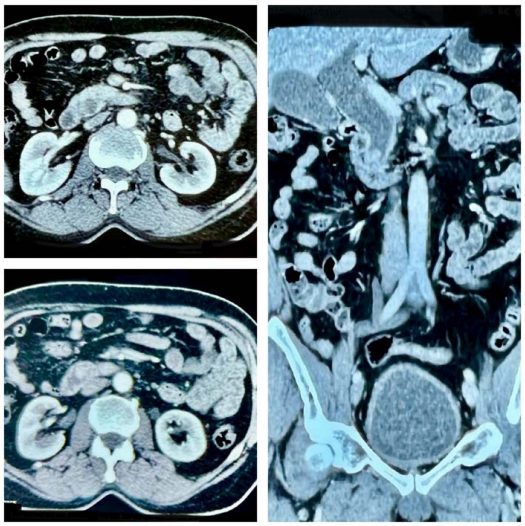

Figure 1: Axial And Coronal View of Computed Tomography Shows Enhancing Lesion in Ampulla with Dilated Common Bile Duct

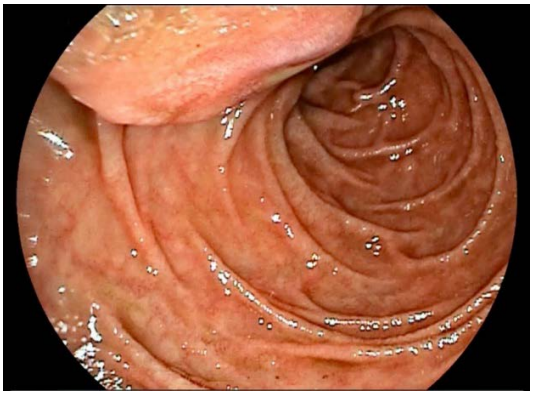

Figure 2: Esophagogastroduodenoscopy Showing Bulging of Ampullary Region with Normal Mucosa Covering It

This case was reviewed in a multidisciplinary gastrointestinal (GI) oncology meeting, and after discussion with the patient, an open standard pancreaticoduodenectomy (PD) was performed (Figure 3). He had a smooth post operative course with no major complications. The resected specimen showed 1.5 x 1.5 x 1 cm nodule at the ampulla that on microscope showed a well circumscribed nodular proliferation of ducts, glands and smooth muscle cells in a disorganized pattern diagnostic of adenomyoma of ampulla (Figure 4).

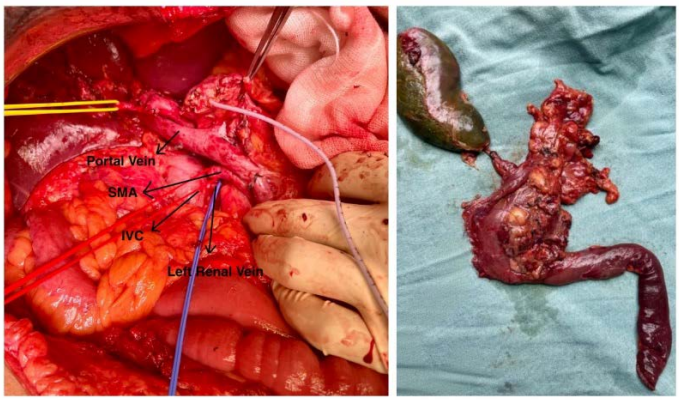

Figure 3: Intraoperative Image Showing Vascular Structures After Pancreaticoduodenectomy (Left) And Standard Pancreaticoduodenectomy Specimen (right). Sma – Superior Mesenteric Artery, ivc – Inferior Vena Cava.

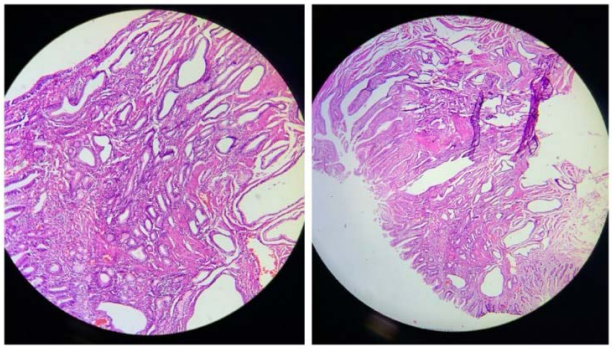

Figure 4: Histopathological Imaging - Hematoxylin and Eosin Staining Shows Nodular Proliferation of Ducts, Glands and Smooth Muscle Cells in A Disorganized Pattern Without Any Atypia or Mitosis, Suggestive of Adenomyoma of Ampulla.

On post operative day (POD) 7 patient was irritable, talking irrelevantly and developed mild slurring of speech. On evaluation he was found to have severe hypercalcemia (total serum calcium of 15.8 mg/dl) and on further investigation, was found to have primary hyperparathyroidism (increased serum parathyroid hormone levels of 663 pg/ml; normal range 10-65 pg/ml). Ultrasonography of the neck revealed a well-defined lesion of 3 x 3 x 1.6 cm in the left parathyroid region. Endocrinology consultation was done, and he was evaluated for associated multiple endocrine neoplasia (MEN) syndrome. Magnetic resonance imaging (MRI) brain was normal with normal prolactin, growth hormone levels and normal thyroid function test. He was found to have none of the associated features of MEN syndrome. Technetium–99m Sestamibi (MIBI – methoxy isobutyl isonitrile) parathyroid scan and MIBI- pertechnetate subtraction study confirmed the diagnosis of left parathyroid adenoma (Figure 5

a). After optimizing the patient, surgery to remove the left parathyroid adenoma was done, and biopsy confirmed the diagnosis of left parathyroid adenoma (Figure 5 b,c,d). He was discharged in stable condition, and he is symptom free and on regular follow up for the last 8 months.

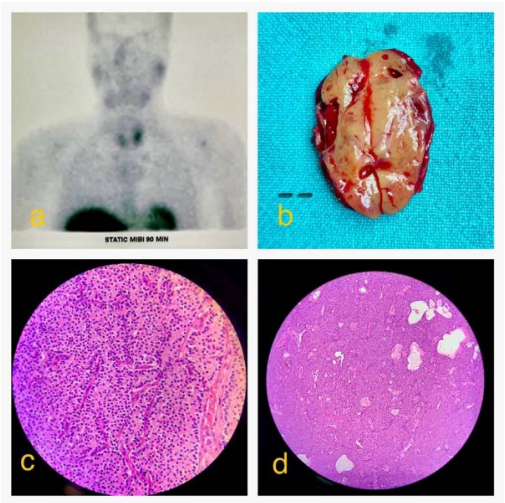

Figure 5: 5a - Sestamibi Scan Showing Increased Tracer Uptake in Left Parathyroid Region with Persistent Hold Up till Delayed Images. 5b – Cut Open Specimen of Left Parathyroid Adenoma. 5c (40x magnification) and 5d (10x magnification) – Hematoxylin and Eosin-Stained Microscopy Images Shows a Benign Neoplasm Composed of Chief Cells with Granular Cytoplasm and Round Vesicular Nucleus Admixed with Areas of Hemorrhage and Cystic Change, Consistent with Parathyroid Adenoma.

Day	Events
0	Patient presented to the hospital with abdominal pain and yellowish discoloration of eyes
5	Multidisciplinary GI oncology meeting – suspected periampullary malignancy and final decision for surgery made
7	Open standard pancreaticoduodenectomy surgery was done with no major complications
14	POD 7 – he was irritable and talking irrelevantly
17	Sestamibi scan confirmed the diagnosis of left parathyroid adenoma
19	Left parathyroidectomy was done
23	Patient discharged in stable condition

Discussion

Adenomyomas are defined as duct-like structures with hyperplasia of smooth muscle cells, combining both epithelial and mes- enchymal elements [2]. Adenomyoma is relatively more com- mon in gallbladder and extremely rare in AOV with less than 60 cases being described in indexed literature so far, reported most- ly as single case reports [3,4]. The exact histogenesis of adeno- myoma in AOV is still unknown with two common hypotheses about their origin. One states that chronic inflammation of papilla causes both muscular and adenomyomatous hyperpla- sia while the other one states that these lesions might be incomplete heterotopic pancreatic tissue [5].

Preoperative diagnosis of adenomyoma of AOV is highly challenging as its presentation mimics that of periampullary malignancy. Radiological imaging (CECT, MRI, MRCP) doesn't reliably differentiate them, as both appear as tumor like mass in ampullary region with upstream biliary dilatation [3].

Diagnosis is confirmed only by histological examination, but it is not always possible on endoscopic biopsies as overall accuracy is low [6]. in their prospective study of 40 patients with papilla of vater tumors, concluded that in case of enlarged or suspicious papillae, endoscopic biopsies from both deep and superficial layers following sphincterotomy should be obtained to improve diagnostic accuracy. In a 10-year retrospective study by Gamble et al. of AOV biopsy specimens from 252 patients, many benign ampullary biopsies were later found to be ns malignant upon resection, indicating that a negative endoscopic biopsy does not rule out malignancy in ampullary lesions. [7].

In our patient this is the reason we didn't proceed with endoscopic biopsy and directly went ahead with PD surgery. Although a few reports [4] suggest that endoscopic papillectomy or surgical ampullectomy is sufficient for these benign ampullary lesions, most patients undergo Whipple procedure and are diagnosed definitively as adenomyoma only after the final biopsy (Table 1). Choosing the appropriate treatment in these cases is highly challenging, because both radiology and endoscopy often cannot accurately distinguish between benign and malignant ampullary lesions as shown in the below table (Table 1)

Serum calcium levels are not routinely checked in the preop- erative evaluation of these patients if kidney function tests are nor-

mal, particularly in resource-limited settings like the Indian subcontinent. Our patient is unique in that he had no symptoms of hypercalcemia preoperatively but developed them post-sur- gery. An incidental CECT finding revealed a right kidney mi- crolith, which was asymptomatic and not further evaluated. In retrospect, assessing this asymptomatic microlith with serum calcium levels could have led to a preoperative diagnosis of the parathyroid adenoma, potentially reducing overall morbidity.

Association of hyperfunctioning parathyroid adenoma with ampullary lesions is very rare, although association with Gastro-Entero-Pancreatic (GEP) neuroendocrine tumor is reported in MEN type 1 syndrome [12]. Our patient had a normal MRI brain and pi- tuitary hormone levels with negative GEP neuroendocrine work- up. Despite extensive literature search, the association of adeno- myoma of ampulla with a hyperfunctioning parathyroid adenoma has not been reported before and further research might be needed to shed some light on this unique association.

Table 1: Literary Review of Similar Case Reports of Adenomyoma of Ampulla

Authors	Age /Sex	Clinical presentation	LFT	Imaging/Endoscopy	Preoperative biopsy	Treatment
Tanimu et al [9], 2014	61/M	Pain upper abdomen	Obstructive pattern (elevated direct bilirubin and ALP)	CT: acute pancreatitis with dilatation of CBD and dilated pancreatic duct. EUS: multilobulated hypoechoic ampullary density (2.4 × 2.1 cm)	EUS-FNA: reactive cells. Endoscopic biopsies: inflammatory polyp versus inflammatory changes	Endoscopic ampullectomy
Choi et al [10], 2016	42/M	Jaundice and upper abdominal pain	Obstructive pattern	CT: abrupt narrowing of the distal CBD and proximal biliary dilatation	Endobiliary biopsies: chronic inflammation with fibrosis and dysplastic change	PD
Gialamas et al [5], 2018	73/F	Jaundice	Obstructive pattern	MRCP: stenosis of the distal CBD at ampullary level, with dilatation above this region. EUS: retro-ampullary mass	Endoscopic biopsies: atypical cells and chronic inflammation without dysplasia	PD
Gouveia et al [3], 2019	58/M	Upper abdominal pain	Mildly elevated transaminases with normal bilirubin and ALP	Endoscopy: bulging papilla. EUS: mass in the ampulla area	EUS-FNA: epithelial cell groups, some with benign characteristics, others in favour of epithelial dysplasia	PD
Frutuoso et al [11], 2021	74/F	Incidentally detected for evaluation of renal colic pain	Normal	CT – nodular lesion in distal CBD with proximal biliary dilation and dilated MPD and left renal tumor. MRCP: ampullary mass causing bicanalar obstruction	Not done	PPPD with partial nephrectomy
Kwon et al [12], 2023	47/F	Abdominal pain	Obstructive pattern	CT: mild extrahepatic biliary dilation with diffuse wall thickening and smooth distal CBD tapering	Endoscopic biopsy: chronic inflammation with no atypical cells	PPPD

LFT – liver function test; CT – computed tomography; CBD – common bile duct; EUS- endoscopic ultrasound; FNA- fine needle aspiration; PD- pancreaticoduodenectomy; MRCP- magnetic resonance cholangiopancreatography; ALP – alkaline phosphatase; MPD – main pancreatic duct; PPPD – pylorus preserving pancreaticoduodenectomy.

Conclusion

Adenomyomas of the AOV and distal CBD are rare benign lesions, typically diagnosed through postoperative biopsies, as they often mimic malignant neoplasms and rarely receive a definitive preoperative diagnosis from radiology or endoscopy. There are currently no established guidelines or protocols for evaluating and managing this condition. Urgent research is needed to implement newer techniques, such as EUS, immunohistochemistry (IHC), and advancements in frozen sections and pathology, to improve diagnostics and enhance patient outcomes.

Serum calcium levels should be assessed in all periampullary lesions, as this simple test could have identified the parathyroid adenoma preoperatively, particularly given the patient's kidney microliths. This case highlights the importance of thoroughly evaluating even minor incidental findings before major resections, even if the patient is asymptomatic.

Authors' Contributions: Karthikayan V R was the Senior Res- ident surgeon in-charge of the overall care of the patient and was involved in the day-to-day patient care and follow up. Karthikayan V. R. wrote the first draft of the manuscript and collected valuable data on the case compilation. Vinay B.N was the Professor and Head of department, who did the primary surgery and was involved in supervising the entire management of the patient and did the final proof reading of the manuscript. Arun H N was the Surgical oncologist who operated the parathyroid adenoma. Gautham M V was the first assistant in both the surgeries for the patient and was also involved in proof reading of the manuscript.

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Nil

Declaration of Conflicting Interests

The authors declare that there is no conflict of interests regarding the publication of this case report.

Informed Consent for Publishing

Obtained from the patient concerned.

Ethical Approval

Not applicable /not necessary at our institution to publish an anonymous case report.

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