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## Case Report

# Periampullary duodenal schwannoma mimicking ampullary neoplasm

Marly Pierina Rubio Sierra, MD<sup>a</sup>, Aydamir Alrakawi, MD<sup>b</sup>, Ahmad Alduaij, MD<sup>c</sup>,  
Dana AlNuaimi, MD<sup>a</sup>, Numan Cem Balci, MD<sup>a,\*</sup>

<sup>a</sup>Imaging Institute, Cleveland Clinic Abu Dhabi, Maryah Island Abu Dhabi United Arab Emirates

<sup>b</sup>Digestive Disease Institute, Cleveland Clinic Abu Dhabi, Maryah Island Abu Dhabi United Arab Emirates

<sup>c</sup>National Reference Laboratories, Cleveland Clinic Abu Dhabi, Maryah Island Abu Dhabi United Arab Emirates

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## ABSTRACT

Schwannomas are neurogenic tumors that arise from Schwann cells in the neural sheath. Gastrointestinal schwannomas occur most often in the stomach, followed by the colon and the rectum. Duodenal schwannomas are rare amongst mesenchymal tumors of the gastrointestinal tract and only a few cases have been reported up to the current date with an incidence of approximately 2%–6%. Duodenal Schwannomas do not have characteristic imaging features thereby cannot be easily differentiated from other submucosal and adjacent extraluminal neoplasms. We present a case of a 76-year old male patient that presented to our hospital with abdominal pain and was diagnosed after an upper gastrointestinal endoscopy with an ampullary duodenal neoplasm that proved to be a periampullary duodenal Schwannoma on histopathology. Duodenal Schwannomas although rare should be considered in the differential diagnosis of ampullary neoplasms.

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## Introduction

The evaluation of submucosal lesions in the alimentary tract is difficult with endoscopy. Cross-sectional imaging can help identify the location of the pathology and characterize its nature [1].

The ampulla and periampullary regions are difficult to evaluate radiologically, and the imaging characteristics of benign and malignant conditions in these areas overlap [2,3].

Furthermore, while some entities can be well evaluated with the high spatial resolution computed tomography imaging other lesions are better visualized with the superior contrast resolution of magnetic resonance imaging [4]. The second portion of the duodenum is aligned with the pancreas and a submucosal lesion at the papillary level can mimic cystic pancreatic neoplasms [5].

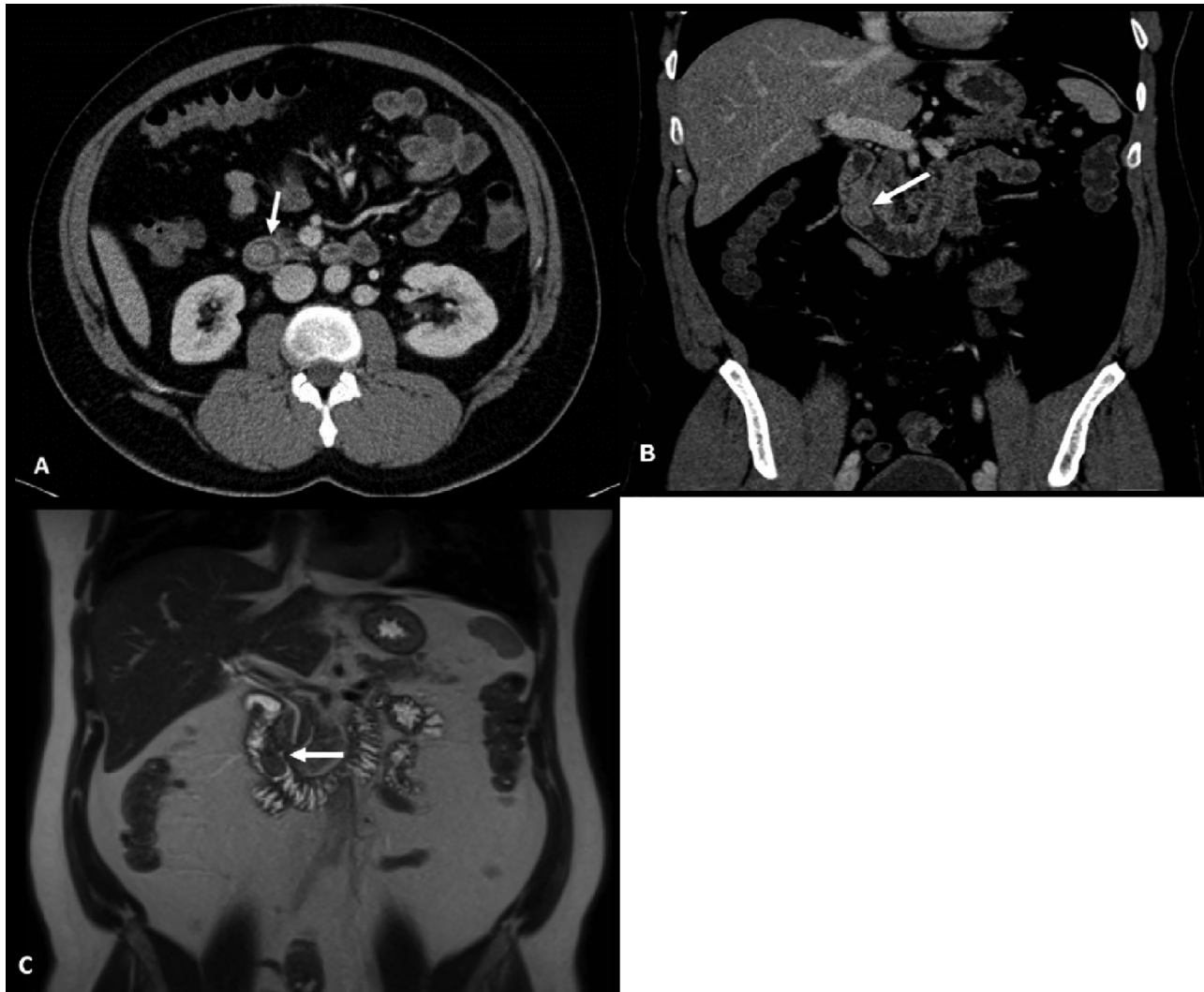
We report a case of a duodenal schwannoma that mimicked an ampullary mass on a cross-sectional imaging and was finally diagnosed on a histopathological specimen obtained during an endoscopic ultrasound.

\* Corresponding author.

E-mail address: [balcin@clevelandclinicabudhabi.ae](mailto:balcin@clevelandclinicabudhabi.ae) (N.C. Balci).

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**Fig. 1 – Computed Tomography (A, B) and MRI (C) of the patient. A-B. Axial and coronal CT images reveal an enhancing polypoidal lesion at the ampulla mostly intraluminal (arrow). C. Coronal T2 weighted images showing the same lesion with no evidence of biliary dilatation (arrow).**

## Case Report

A 76-year-old male patient presented to our healthcare facility with a long history of upper abdominal pain. The pain has been intermittent for the past 10-15 years with mild to moderate intensity and was not related to meals or exertion.

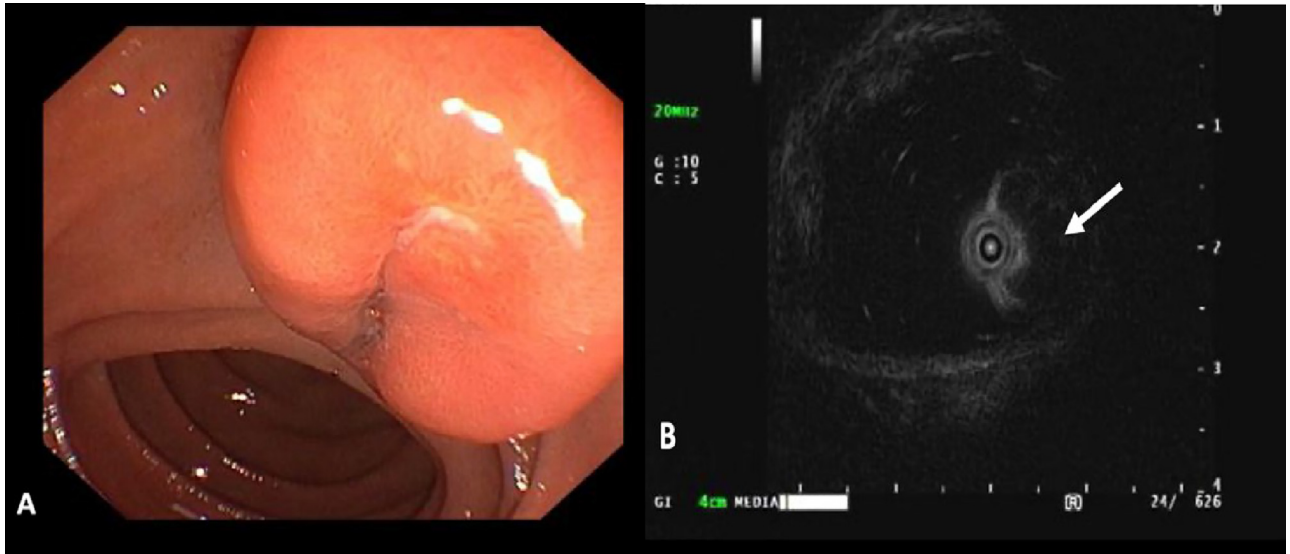
The blood work-up was unremarkable. CA19-9, lipase, ALT, AST, ALP, creatinine, electrolytes, and blood glucose levels were all within the normal limits. The patient had an upper gastroduodenal endoscopy a few months back in another facility which showed a submucosal duodenal mass at the periampullary location.

He underwent a contrast-enhanced computed tomography (CT) examination in the portal venous phase upon his admission in our facility and the CT showed a polypoidal vegetative mass at the ampulla of Vater with intraluminal growth into the duodenum (Fig. 1). A hypodense lesion was also detected in the liver. The patient then underwent magnetic resonance

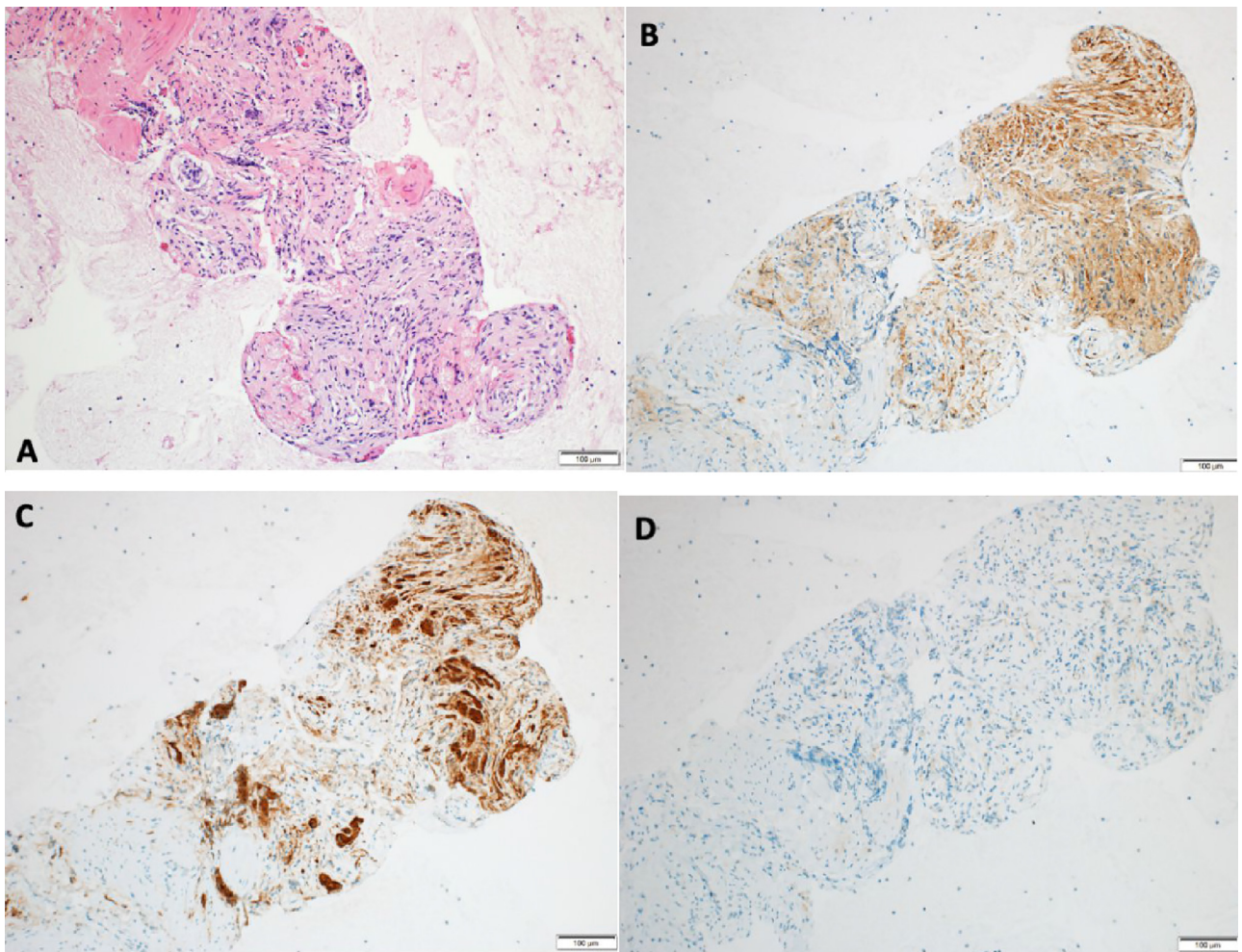
imaging (MRI) to characterize the liver lesion and to visualize the extent of the ampullary mass and its relationship to the common bile duct and pancreatic duct. It showed the same lesion at the ampulla but without evidence of biliary or pancreatic duct dilatation (Fig. 1).

The liver lesion had a characteristic imaging feature of a hemangioma. Thereafter, the patient underwent endoscopic ultrasound (EUS) and biopsy of the ampullary mass. On EUS, an intramural (subepithelial) hypoechoic lesion was found in the second portion of the duodenum. The lesion appeared to originate from the submucosal third echo layer (Fig. 2).

Fine needle biopsy was performed. There was no sign of significant pathology in the common bile duct. On histopathology, the sample showed bland spindle cells with S100 immunostain and focal synaptophysin expression, most suggestive for a neural origin such as a schwannoma (Fig. 3). Follow-up was recommended due to the patient's unwillingness for surgical excision.



**Fig. 2 – (A)** The endoscopic image showing a polypoid submucosal lesion at the ampulla of Vater. **(B)** Endoscopic ultrasound reveals a hypochoic submucosal lesion (arrow).



**Fig. 3 – (A-D)** Histopathology of the fine needle biopsy revealing S100 positive spindle cells in the periampullary lesion **A.** The hematoxylin and eosin staining shows bland appearing spindle cells. **(B-C)** S100 and Synaptophysin immunostains are expressed in the spindled cells. **D.** DOG-1 is negative.



## Discussion

Schwannomas are rare peripheral nerve sheath tumors and may arise in any nerve where Schwann cells are present. Gastrointestinal schwannomas are rare mesenchymal tumors and were first described by Daimaru et al. in 1988 [6]. They arise from Schwann cells of the nerve sheath of Auerbach's plexus, in contrast to conventional schwannomas which arise from peripheral nerves of the skin, connective tissue, and internal organs [7].

Gastrointestinal Schwannomas are frequently found in synchronous locations in the same patient and are often associated with Von Recklinghausen disease. The stomach is the most frequent location of gastrointestinal Schwannomas [8]. Duodenal Schwannomas are very rare and only a few cases are reported in the literature.

Three forms of duodenal schwannomas have been described: submucosal intraluminal, subserous extrinsic or intramural schwannomas and each presents with different symptoms with the submucosal intraluminal type being the most common type [9].

Duodenal Schwannomas exhibit slow growth thereby delaying symptoms. Most often they remain unrecognized clinically. Intraluminal Schwannomas may cause mucosal ulceration and intestinal bleeding which may be their initial presentation [10,11]. They may remain indolent and vague with nonspecific symptoms related to intraluminal compression and patients present with abdominal pain, indigestion, and vomiting. In our case, the schwannoma was submucosal intraluminal type and the patient had vague upper abdominal symptoms possibly due to mild intraluminal obstruction.

Duodenal Schwannomas are mostly located in the second or third portion of the duodenum [12]. In our case, the location of the tumor mimicked an ampullary neoplasm on cross-sectional imaging because of its location [13]. Ampullary neoplasms are associated with upstream biliary dilatation, which was not pronounced in our case. Other differential diagnoses of submucosal duodenal lesions include leiomyomas and gastrointestinal stromal tumors (GIST) [14].

Only a few case reports described CT and MRI findings of duodenal schwannomas that appear similar on imaging to other gastrointestinal tract schwannomas showing arterial contrast enhancement on CT and MRI. Schwannomas tend to have mixed signal intensity on T1 and T2 weighted MRI images, cystic degeneration can also occur [15]. If the Schwannoma is hypervascular, it cannot be differentiated from neuroendocrine tumors [16,17].

Malignant transformation of Schwannomas may occur and are difficult to distinguish from malignant neurogenic tumors by cross-sectional imaging alone [18,19]. Biopsy and histopathologic evaluation is needed for diagnoses and to establish the malignant transformation of the schwannoma. It was done by endoscopic ultrasound in our case. The Schwannoma was located in the subepithelial third echo layer which is not a usual location. Duodenal Schwannomas are hypoechogenic and their expected location is in the fourth echo layer which represents the muscularis [20].

On histological examination, schwannomas are encased by the intact mucosa and usually involve the

submucosa and muscularis propria. They are composed of spindle-shaped nuclei with high and low cellularity regions. Since conventional 6 hematoxylin and eosin staining cannot distinguish neurogenic and myogenic tumors, immunohistochemistry is essential for the differentiation between schwannomas and GISTs or leiomyomas. Cells of schwannomas are 100% immunoreactive with S-100 protein [21,22]. GISTs and leiomyomas are negative for S-100 protein [23].

The treatment of choice is complete surgical resection, with an approach that depends on tumor size, localization, and histological features [24].

In conclusion, we described a rare case of duodenal schwannoma which resembled an ampullary neoplasm on cross-sectional imaging. Endoscopic ultrasound aided in differentiating the submucosal mass from an ampullary neoplasm and histopathologic evaluation confirmed the final diagnosis.

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