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Peripancreatic schwannoma: A case report

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ARTICLE INFO	A B S T R A C T
<i>Keywords:</i> Peripancreatic Schwannoma Pancreatic mass Enucleation	 Introduction: Schwannomas are a benign tumor of peripheral nerve sheath origin. Although most commonly arising in the extremities, head, and neck there have been rare cases of schwannomas presenting within or adjacent to the pancreas reported in the literature. Presentation of case: We present an asymptomatic and otherwise healthy 57-year-old male with an incidental peripancreatic mass measuring 3.5 × 3.7 cm found on abdominal computed tomography (CT). The patient underwent complete enucleation of the mass at our hospital. Pathological analysis of the excised specimen showed spindle shaped cells. Immunohistochemical staining was positive for S100 protein expression, confirming the definitive diagnosis of peripancreatic schwannoma. The patient's postoperative course was uneventful. Discussion: Schwannoma is a commonly benign, encapsulated, and slowly growing tumor arising from myelin producing cells of peripheral nerves. A schwannoma presenting within or adjacent to solid abdominal viscera, such as the pancreas, may be challenging to diagnose preoperatively as it may mimic other lesions. Radiological analysis. The use of endoscopic ultrasound (EUS) with fine needle aspiration (FNA) may assist the clinician in preoperative diagnosis, however these interventions are not widely available. Accurate preoperative diagnosis of a peripancreatic schwannoma is of high importance as it may preclude unnecessary pancreatectomy or radical resection. Conclusion: Although rare, schwannoma should be part of the differential diagnosis of a cystic or solid appearing mass within or surrounding the pancreas. Total resection carries an excellent prognosis with little to no documented recurrence.

1. Introduction

Schwannomas, first described by Verocay in 1910, are benign tumors originating from Schwann cells of the peripheral nerve sheath [1]. The schwannoma commonly arises as a slowly growing and solitary tumor attached to a peripheral nerve [2]. In decreasing frequency, schwannomas occur in the lower extremity, upper extremity, head and neck, retroperitoneum, and the pelvis [3]. Specifically, there have been less than 70 case reports of pancreatic schwannoma reported in the English literature [4]. Schwannomas that abut the pancreas are less common [5].

The pancreas is richly innervated by sympathetic and parasympathetic nerve fibers via the celiac and superior mesenteric plexus [5]. Prior to entering the pancreas, the peripheral autonomic nerves converge at several extra-pancreatic plexuses [5]. By definition, a schwannoma may arise at any point along a peripheral nerve's course.

Clinical features of a peripancreatic schwannoma are dependent on its anatomical location and growth of the tumor. We present a case of complete enucleation of a peripancreatic mass in an asymptomatic patient that was performed at an academic center. Included is a discussion on the pathological and radiological characteristics of schwannoma. The aim of this case report is to propose the inclusion of schwannoma in the differential diagnosis of a cystic or solid mass found within or adjacent to the pancreas. The work has been reported in line with the SCARE criteria [6].

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Case report

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2. Case report

A physically active 57-year-old male, with no significant past medical history, presented for outpatient evaluation of an incidental peripancreatic mass identified on CT. He denied any gastrointestinal symptoms and had no complaints at the time of presentation. He is a non-smoker and drinks alcohol 1–2 times per year. Family history was noncontributory. On physical examination, the patient was anicteric, the abdomen was soft and non-tender, with no signs of organomegaly. No palpable mass was appreciated.

The patient was sent for routine CT of the abdomen by his urologist, who has followed the patient for several years for management mild benign prostatic hyperplasia. The original scan (Fig. 1) showed a 3.5×3.7 cm, mildly heterogeneously enhancing, solid mass at the mid mesentery. Preliminary radiological differential diagnosis included desmoid tumor, malignant lymph node, and exophytic solid mass of the uncinate process of the pancreas. A CT scan from 2 years prior was unremarkable in comparison, with no evidence of this mass. Endoscopic ultrasound (EUS) with fine needle aspiration (FNA) was subsequently performed, and pathological analysis of the obtained specimen identified a spindle cell neoplasm. Immunohistochemical staining was positive for S-100. Staining for CD117, CD34, desmin, and smooth muscle actin (SMA) were negative. The tumor makers CA 19–9 and CEA (16.4 U/mL and 3.0 ng/mL respectively) and all other chemistries were within normal reference limits.

Based on its location and the ambiguity of diagnosis, the decision to surgically excise the mass was made. The patient was admitted to our hospital for surgery several months after the mass was identified on CT scan and the patient remained asymptomatic during this interval. A preoperative CT of the abdomen and pelvis was performed (Figs. 2, 3), noting an ovoid, soft tissue mass of similar size to the previous scan, just anterior to the 3rd portion of the duodenum and adjacent to the tip of the uncinate process of the pancreas. There was no ductal dilatation or calcification of the pancreas. No dilation of the biliary tree was present.

The patient subsequently underwent exploratory laparotomy. A well-circumscribed mass was identified posterior to the uncinate process of the pancreas and superior mesenteric vein, and anterior to the aorta.

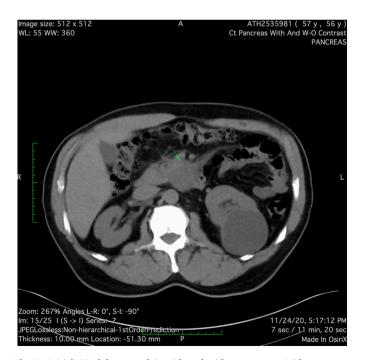


Fig. 1. Initial CT abdomen/pelvis with and without contrast. Mid mesentery mass (green arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)



Fig. 2. Preoperative CT abdomen and pelvis with contrast, sagittal view, showing a circumferential mass adjacent to the pancreas and duodenum.

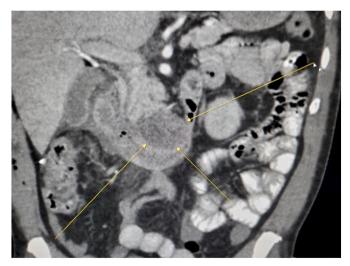


Fig. 3. Preoperative CT abdomen and pelvis with contrast, coronal view, showing an ovoid mass (arrows) posterior to the third portion of the duodenum and abutting the pancreas.

The mass was completely enucleated. Pathological examination of the gross specimen showed a tan, pink to hemorrhagic capsulated mass with a smooth outer surface, measuring $4.5 \times 4.0 \times 2.4$ cm. On cut section, the specimen appeared tan-brown, glistening, and focally hemorrhagic. The specimen was negative for pancreatic parenchymal tissue, affirming our peripancreatic description.

Histopathological analysis showed a fairly well-circumscribed spindle cell neoplasm predominantly arranged in fascicles (Figs. 4 and 5).

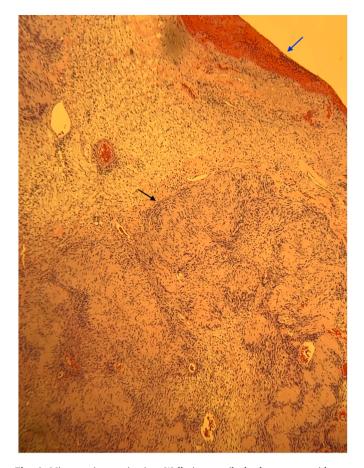


Fig. 4. Microscopic examination. Well circumscribed schwannoma with surrounding capsule (blue arrow). Spindle-shaped cells predominantly arranged in fascicles (black arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

Numerous characteristic Antoni areas with zonally variable cellularity were present. Cellular areas consisted of tumor cells with wavy nuclei palisading around fibrillary processes. On immunohistochemical analysis, the tumor cells stained strongly for S-100 protein. These features strongly supported the diagnosis of schwannoma. Degenerative changes were also noted, including myxoid change, hemosiderin deposition, and focal ossification. Scattered bizarre nuclei were appreciated with no evidence of increased mitotic activity, consistent with benign degenerative change rather than malignant transformation.

The patient tolerated the procedure well without complication. He was discharged home after a 5-day postoperative recovery in the hospital.

3. Discussion

Schwannomas are a benign mesenchymal neoplasm deriving from peripheral nerve sheath Schwann cells. They are the most common tumor of peripheral nerves and very rarely degenerate into malignant forms. Schwannomas may either be sporadic or related to genetic syndromes such as von Recklinghausen disease and neurofibromatosis type 2 [7]. Regardless of etiology, they commonly arise as encapsulated singular tumors from an individual peripheral nerve in the face, neck, major nerve trunks, and extremities [2,5]. Schwannomas rarely arise in the abdomen, including the gastrointestinal tract and abdominal viscera, with 3% of all schwannomas presenting in the retroperitoneal region [2,5]. Schwannomas arising within the pancreas proper are less prevalent, with less than 70 documented case reports describing pancreatic schwannomas over the last 40 years in the English literature [4].

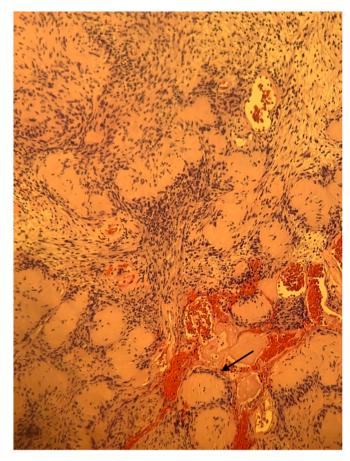


Fig. 5. Microscopic examination. Spindle cells with wavy nuclei palisading around focal acellular areas of myxoid material (black arrow).

Gross pathological examination of schwannoma commonly shows a soft, encapsulated tumor with a smooth nodular outline [7]. Cut section shows yellowish to tan color, often with areas of hemorrhage or cystic change. Microscopic analysis characteristically yields spindle shaped tumor cells. A schwannoma is microscopically composed of two areas: Antoni A and Antoni B. Antoni A areas are hypercellular, with fascicles of Schwann cells that may feature nuclear palisading. This often sharply contrasts with Antoni B areas, which are predominantly acellular, and composed of loosely textured myxoid components [7,8]. Immunohistochemical analysis stains strongly for S-100 protein. Negative staining for desmin, CD34, SMA, and CD117 differentiates schwannoma from other neoplasms of mesenchymal origin. Additionally, degenerative changes in the forms of cyst formation, hemorrhage, and hyalinization of vasculature may be present within some schwannomas [9].

An accurate preoperative diagnosis of an abdominal schwannoma may be difficult due to the nonspecific radiological features of these tumors [5]. This may be particularly challenging when the tumor is bordering or located within visceral organs, as it may mimic other more common lesions. The CT appearance of schwannomas has classically been described as a well-circumscribed homogenous soft tissue mass [10]. However, schwannomas have also been reported to present with mixed attenuation on CT, which may be mistaken for complex cysts or malignant lesions [8,9,11,12]. It has been suggested that the aforementioned pathologic degenerative changes within the tumor may account for the heterogeneous contrast uptake seen on imaging [1,3,6].

The clinical features of peripancreatic schwannomas are also nonspecific. Schwannomas are typically slowly growing tumors, and the size at which they become symptomatic is unknown [5,13]. Kuo et al. [5] described the clinical presentation of 17 patients with peripancreatic schwannoma: 10 were asymptomatic and incidentally found on health screening, 5 had abdominal pain, and 2 had an abdominal mass on physical examination. Additionally, the authors report that 45 out of 63 patients with other retroperitoneal schwannomas were asymptomatic [5]. Regardless of location, surgical excision is the recommended course of treatment, as pathological examination is required for definitive diagnosis. A schwannoma arising within pancreatic parenchyma typically requires more extensive resection, such as pancreatectomy or Whipple procedure [13,14].

Intraoperative frozen sections are valuable to confirm the diagnosis of a benign schwannoma. When malignant transformation cannot be completely ruled out preoperatively, pancreaticoduodenectomy or distal pancreatectomy is generally favored [14]. In proven benign cases, simple enucleation is generally sufficient [15], and unnecessary radical resection may safely be avoided [5,15]. After complete excision of the tumor, long-term prognosis is excellent, and no recurrence has been documented in multiple case reports [4,5,13].

4. Conclusion

Incidental abdominal masses are identified with increasing frequency with the increased use of multi-sectional imaging. Preoperative diagnosis may be challenging due to ambiguity of radiologic features across a wide differential of potential pathological processes. Although rare, schwannoma should be considered in the differential diagnosis of a cystic or solid peripancreatic mass. Depending on their anatomical location, confirmed benign schwannomas can be enucleated, which may preclude unnecessary radical resection.

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Ethical approval

This case report is exempt from ethical approval by our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Sumer Moussa: 1. Wrote the case report, 2. Revised and edited the case report, 3. Approved final version of case report.

Stephanie Cruz: 1. Participated in patient care 2. Revised and edited the case report, 3. Approved final version of case report.

Mark Ingram: 1. Participated in patient care 2. Revised and edited the case report, 3. Approved final version of case report.

Jamshed Zuberi: 1. Revised and edited the case report, 2. Approved final version of case report.

Registration of research studies

This is not a "first in humans" report, so it is not in need of registration.

Guarantor

Sumer Moussa.

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Data sharing policy

The authors declare that data supporting the findings of this study are available within the article (and its supplementary files).

Declaration of competing interest

The authors report no declaration of competing interest.

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