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## Pancreatic schwannoma: A rare case and a brief literature review



Metin Ercan<sup>a</sup>, Mehmet Aziret<sup>a,\*</sup>, Ali Bal<sup>a</sup>, Adem Şentürk<sup>a</sup>, Kerem Karaman<sup>a</sup>,  
Zeynep Kahyaoğlu<sup>b</sup>, Havva Belma Koçer<sup>a</sup>, Birol Bostancı<sup>a</sup>, Musa Akoğlu<sup>a</sup>

<sup>a</sup> Sakarya University of Faculty of Medicine, Department of General Surgery, Sakarya, Turkey

<sup>b</sup> Sakarya University of Faculty of Medicine, Department of Pathology, Sakarya, Turkey

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## ABSTRACT

**INTRODUCTION:** Pancreatic schwannoma (PS) is an extremely rare benign tumor. Less than 50 cases of pancreatic schwannoma have been described in the English literature over the past thirty years.

**PRESENTATION OF CASE REPORT:** A 63-year-old female underwent left modified radical mastectomy 2 years ago due to breast cancer. During her routine check-up, a 65 × 63 × 55 mm measured calcified, well-demarcated, cystic-mass having septations and calcifications that localized to the pancreatic head was detected by abdominal computerized tomography. She was asymptomatic and her tumor markers were in normal ranges. A standard Whipple procedure was performed, and the histo-pathological diagnosis of the resected specimen was reported as ancient schwannoma with clear surgical margins. Patient's post-operative course was eventful. She had a biliary leakage after surgery which was managed conservatively. She is under follow-up.

**DISCUSSION:** Pancreatic schwannoma also known as neurilemoma or neuroma is a slowly growing, encapsulated, mostly benign tumor with smooth well-delineated margins that originates from myelin producing schwann cells located on the nerve sheath of the peripheral epineurium of either the sympathetic or parasympathetic autonomic fibers. PS's are extremely rare. The head of pancreas being involved in the vast majority of cases (40%), followed by its body (20%). Management of pancreatic schwannomas remains largely controversial. Both enucleation and radical surgical resections have revealed great therapeutic efficiency, with a well prognosis without recurrences.

**CONCLUSION:** Although rare, PS's should be considered in the differential diagnosis of the other solid or cystic masses of the pancreas.

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## 1. Introduction

Schwannoma or neurilemoma is usually a benign tumor which arises from schwann cells that present in the sheath of cranial, spinal nerve roots and peripheral nerves [1,2]. Schwannom is seen in extremities, head, neck, retroperitoneum, mediastinum, pelvis and rectum [2,4]. PS is an extremely rare and equally hold both genders in adults [5,6]. Less than 50 cases of pancreatic schwannoma have been described in the English literature over the past thirty years. The behaviour of this tumor is not clear which can show sometimes malignant degeneration. Two thirds of PS's have degenerative changes such as cystic formation, calcification, and hemorrhage mimicking pancreatic cystic tumors [5,6,8]. Patients with PS have usually abdominal pain, nausea and vomiting. Weight loss and jaundice may sometimes accompany [5–7]. Diagnosis of preoperative PS is very difficult and contrast enhanced computed tomography (CT), ultrasonography (USG) or

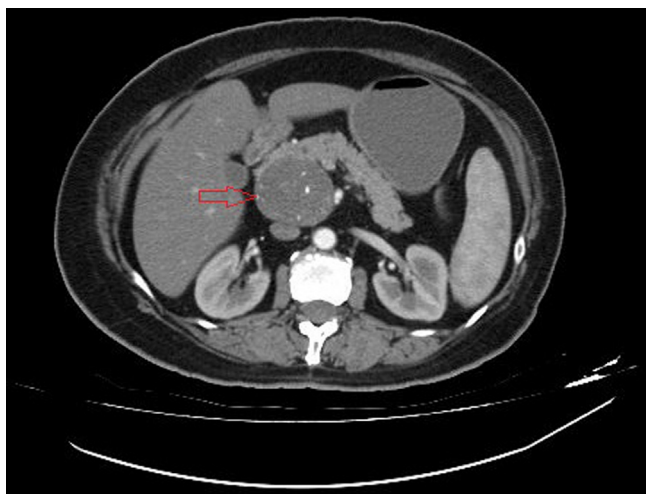
ultrasonography-guided fine-needle aspiration (USG-FNA) may helpful [8–10]. Surgery is the only curative treatment modality consisting of a variety options changing from the simple enucleation to pancreaticoduodenectomy [3,6,11,12]. The following report presents a case with PS who underwent Whipple procedure for a preoperative assumed diagnosis of a pancreatic cystic tumor, with review of literatures.

## 2. Case report

A 63-year-old female patient underwent modified radical mastectomy and axillary dissection two years ago due to left breast cancer (BC). During her follow-up, a cystic mass of the pancreatic head was dedected on abdominal CT. Her tumor markers were in normal range. A complex cystic mass measuring 5.5 × 4.5 mm in diameter with rough calcifications focusing on the internal structure having high density mural nodulation and irregularities in the location of uncinate process was detected by abdominal USG. Contrast enhanced CT detected a cystic lesion 65 × 63 × 55 mm in size with smooth contoured wall including calcifications and septa that hold contrast and also contained millimetric nodules and soft tissue

\* Corresponding author.

E-mail address: [mhmtaziret@gmail.com](mailto:mhmtaziret@gmail.com) (M. Aziret).

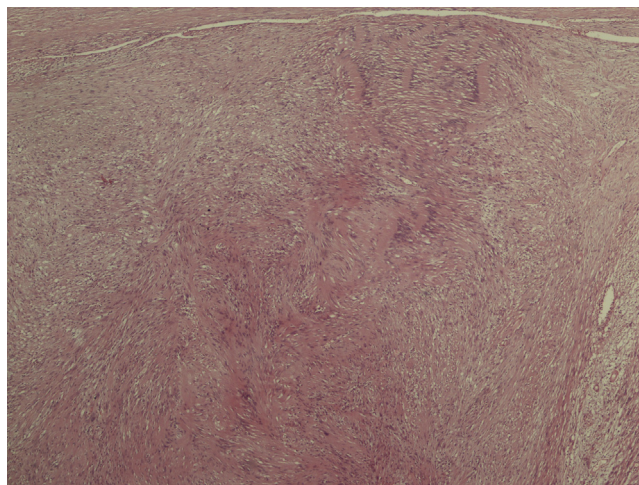


**Fig. 1.** CT image of pancreatic schwannoma in relation with duodenum, pancreas and vascular structures (red arrow).

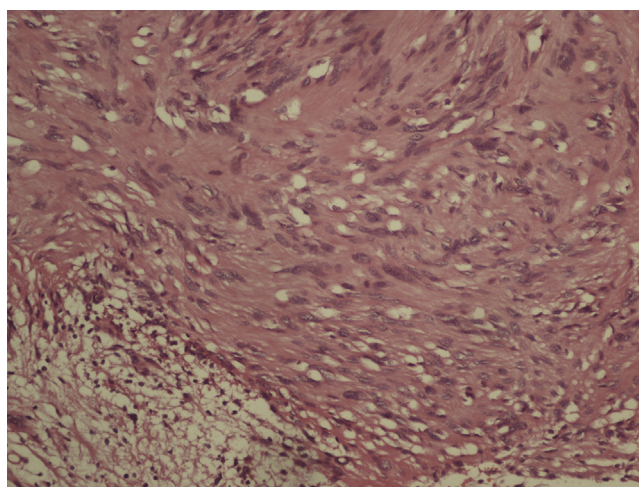
components with no distant metastasis or intraabdominal lymphadenomegaly (Fig. 1). An explorative laparotomy was planned following the anesthesia consultation, which determined her physical status as ASA-2 (American Society of Anesthesiologists). Following a bilateral subcostal incision, a tumoral mass measuring approximately 5 cm in diameter with well-circumscribed, encapsulated hard wall in consistency, involving head of the pancreas and uncinate process was found. The tumor mass did not compress the main biliary tract and there was no dilatation in the pancreatic canal (approximately 2 mm). A standart Whipple procedure was performed. In the postoperative period, biliary leakage had been developed which resolved under conservative treatment. Histopathological examination of the resected specimen revealed a cystic lesion that included an organized hematoma with indistinct some solid fields in the peripheral areas. Microscopically, diffuse signs of degeneration like hemorrhage, organized hematoma, edema, stromal hyalinization, dystrophic calcification, and hyalinization in vascular walls was detected. The tumor had a thick fibro-hyaline capsule which was well separated from the surrounding pancreatic tissue. In immunohistochemical staining, the tumor was diffusely and severely stained with S-100 and vimentin, but it was not stained with CD117 (C-kit), CD34, DOG-1, actin and desmin (Figs. 2–4). Her general condition is now well and she receives regular follow-up care at the Oncology and General Surgery departments.

### 3. Discussion

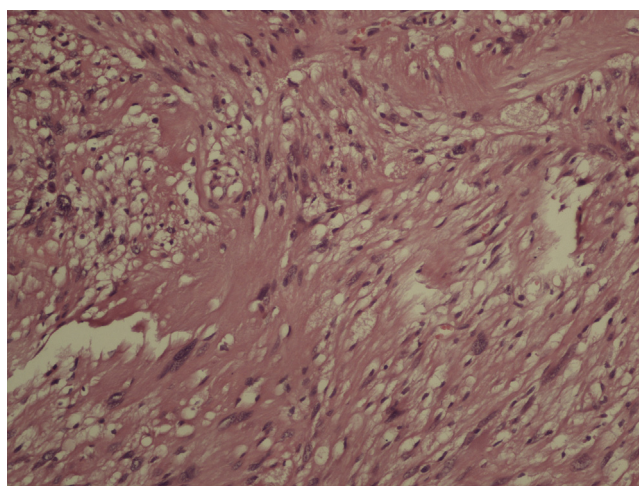
Pancreatic schwannoma also known as neurilemoma or neuroma is a slowly growing, encapsulated, mostly benign tumor with smooth well-delineated margins that originates from myelin producing schwann cells located on the nerve sheath of the peripheral epineurium of either the sympathetic or parasympathetic autonomic fibers [13,14]. In 1910, it was firstly named 'neuroma' by Verocay [1] and then Stout [2] changed its name 'neurilemmo' and finally it was demonstrated that schwann cells proliferated in tumor in electron microscopy. Schwannomas are usually benign, smooth-limited, encapsulated and slowly growing tumors [1–3]. They are frequently seen at the 3th and 5th decades and equally affect both genders [2,3]. Moriya et al. [3] with their 47 schwannoma cases, reported the mean age as 55.7 years (20–87) with a being higher female ratio (female/male = 26/21) [3]. In patients with von Recklinghausen disease, schwannomas are seen more often with an increased malignant transformation [5]. Our patient was



**Fig. 2.** The spindle cells with composed of eosinophilic cytoplasm and indistinct boundaries (Hematoxylin and Eosin) (H&E).



**Fig. 3.** Compact hypercellular and edematous hypocellular areas in pancreatic schwannoma (H&E).



**Fig. 4.** The degenerative nuclear atypic areas (ancient modification) (H&E).

63 years old. PSs are frequently seen in the head and neck areas (25–48%) and they are less detected in the surface of the flexor extremities, posterior mediastinum, pelvis, rectum, and retroperitoneum [4–6]. They are unilaterally located without abbreviations and the most common involved localization is intracranial area. Majority of schwannomas are originated from VIII. cranial nerve (acoustic neuroma) and less frequently involved cranial nerves are the 5th cranial nerve (trigeminal neuroma), II., IX., X. and XII nerves, respectively. PS is extremely rare and made up less than 1% of all schwannomas [3–5]. PS's are derived from autonomic sympathetic and parasympathetic fibers or branch of nervous vagus extending to the pancreas [4–6]. According to the location of PS's, they are mostly located in the head (40%), followed by corpus (21%), neck (6%), tail (15%), and uncinata process (13%), respectively. Two-thirds of the tumors are cystic [6,12]. Because of their slowly growing behaviour, degeneration like hemorrhage, cyst formation, necrosis and calcification may occur and this condition is more often seen in schwannomas. This type of schwannomas is referred as "degenerative or ancient schwannoma" which can often mimic pancreatic cystic neoplasms, neuroendocrine tumors, cystadenoma, cystadenocarcinoma, intraductal papillary mucinous tumor, lymphangioma and pancreatic pseudocyst [7,15]. The present case has features of ancient PS. The classic schwannomas histopathologically include two basic tumoral patterns; Antony A and Antony B [3,15]. The Antony A pattern has a spindle-long cell which shows a tight fence and higher cells with nuclear double-sequence field (palisading) (Figs. 2–4). The Antony B pattern has a loose distribution of the cells with varying degrees of lipidization (Fig. 2). In Antony A pattern, the tumor cell nuclei are in certain order sequence line as long axes to be parallel to each other (the nuclear palisading) and these areas are called 'Verocay bodies' (Fig. 2). The roof of vessel is typically thick-walled containing hyaline. The ancient schwannomas have intranuclear cytoplasmic inclusions and bizarre cells containing nuclear pleomorphism (Fig. 4). The cellular schwannomas consist completely or dominated from Antony A areas where Verocay bodies may not be observed (Fig. 3). The most common locations of cellular schwannomas are the paravertebral areas in pelvis, retroperitoneum and mediastinum [3,7,15]. The flexiform and melanotic schwannoma are other defined histological types. Tumor cells are widely and strongly stained with S-100, Leu-7 and Calretinin by immunohistochemical staining [3,8,15]. The cytogenetic studies have showed that schwannomas have monosomy 22 or loss of material 22q [12,16]. In the present case, we observed wide areas of Antony A patterns which are rich from Verocay bodies and poor areas of Antony B patterns containing smaller number of cells with lipidization (Figs. 2 and 3). Additionally, the tumor cells were stained with S-100 by immunohistochemical staining (Fig. 5). The clinical signs and symptoms are related with the location and size of the tumor leading to motor or sensory dysfunction depending on the tumor involvement of the nerves. Abdominal pain, nausea and vomiting, weight loss, melena, jaundice, back and leg pain, may be seen in intraabdominal localized schwannomas [3,12,15]. In the series by Moriya et al. [3], abdominal pain (57%) was the most observed symptom. Thirty percent of the patients were asymptomatic. Weight loss (13%), back pain (6%), nausea and vomiting (4%), melena (4%) and jaundice (4%) were the other symptom and signs. The mean tumor diameter was  $6.2 \pm 5.1$  cm (range 1–20 cm). The present case was asymptomatic with a tumor size of  $65 \times 55 \times 47$  mm. Diagnosis of asymptomatic PS patients is difficult and CT is helpful to delineate the anatomic relationship between the tumor and surrounding tissues. Suzuki et al. [9] demonstrated in CT images the cystic degenerative areas and reflection of Antoni B cystic components. Additionally, CT can show degenerations depending on the tumor growth such as hemorrhage, cyst formation, necrosis and calcification. MRI shows the schwannomas as hypointense on T1-weighted images and hyper-

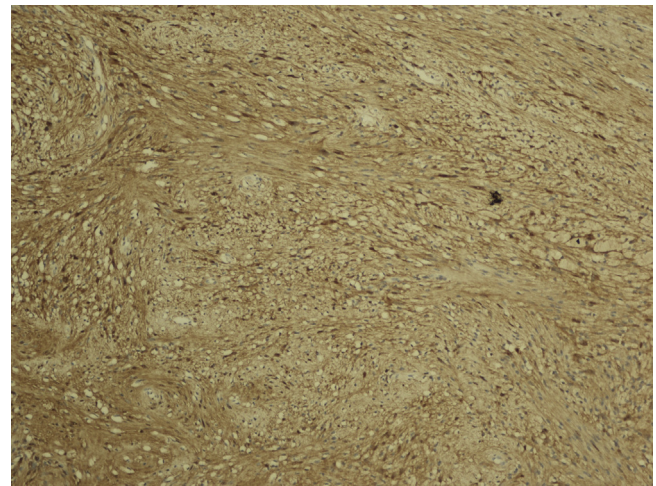


Fig. 5. The S-100 positivity (H&E).

intense on T2-weighted images. But it can be confused easily with other pancreatic tumors. Therefore, it should be considered in the differential diagnosis [17]. EUS-FNA is valuable in diagnosing PS. In cytological examination, schwannomas are characteristically composed of spindle-shaped cells which formed vague cytoplasmic boundaries and embedded a fibrillar wavy nucleus and sometimes has myxoid or collagen matrix. Antony A (adhesive clusters of cells) and Antoni B (loose adherent or poorly cellular page) fields is sometimes present. Immunohistochemical staining is a usually required for accurate diagnosis [10,16]. S-100 protein positivity is an important marker in the diagnosis of schwannoma. Li et al. [10] used EUS-FNA and immunohistochemical staining together in the diagnosis of PS. EUS-FNA is usually used in our clinic following multidetector triphasic contrast enhanced CT according to the clinical situation. In the present case, EUS-FNA could not be done at that time because of the breakdown of the endosonography device.

Enucleation is an effective treatment method because schwannomas are usually encapsulated, slowly growing benign tumors [3,18]. There is 10% documented recurrence in incomplete resections and continuous follow-up is recommended [19,20]. In patients with PS, open (anterior, posterior or combined-methods) or laparoscopic surgery can be used and the anterior approach is generally suggested in retroperitoneal schwannoma. The arterial or venous hemorrhages can be directly prevented with the anterior approach. On the other hand, the posterior approach is frequently used for intrasacral schwannomas [21]. Whipple procedure or distal pancreatectomy can be performed in PS in which malignant transformation could not be preoperatively ruled out. In the series by Moriya et al. [3], 15 patients underwent pancreaticoduodenectomy (32%), 11 patients underwent distal pancreatectomy (23%), 7 patients underwent enucleation (14%), and two patients underwent resection (4%). The remaining patients were usually unresectable or could not be operated [3,21]. Moriya et al. [3], pointed out that malignant formation and cystic formation is strongly associated with increase in tumor diameter. The high suspicion of malignancy in the present case with a past history of malignant breast cancer was the reason why we choose Whipple procedure in the management of this patient.

#### 4. Conclusion

PS is a rare tumor which should be taken into consideration in the differential diagnosis of pancreatic cystic tumors. Surgical resection is the treatment of choice and continuous follow-up remains the standard of care in the management of PSs.

**Conflict of interest**

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**Ethical approval**

None.

**Consent**

The patient's consent was obtained.

**Author contribution**

Metin Ercan→Study design and concept.

Mehmet AZİRET→Data collection and write paper.

Ali Bal→Data collection.

Adem Şentürk→Interpretation.

Kerem Karaman→Design and study concept.

Zeynep Kayhaoğlu→Data collection and analysis.

Erdal Birol Bostancı→Study concept.

Musa Akoğlu→Design and study concept.

**Guarantor**

None.

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