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Case Report
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A Case Report: Pancreatic Schwannoma, A Rare Benign Mimicker of Cystic Neoplasm of the Pancreas

Bhimsi Kandoriya[1], Hirdaya Hulas Nag[1], Pritesh Kumar N[1] and Surbhi Goyal[2]*

[1]Department of Surgical Gastroenterology, GB Pant Institute of Postgraduate Medical Education and Research, Delhi University, India

[2]Department of Pathology, GB Pant Institute of Postgraduate Medical Education and Research, Delhi University, India

Abstract

Background: Schwann cell tumours of the pancreas also known as Pancreatic Schwannoma (PS) is a very uncommon benign pancreatic space occupying lesion. Fewer than 80 cases of PS have been published in the English medical literature over the past forty years.

Case Presentation: A 53-year-old female with hypertension and hypothyroidism presented with a five-year history of dull, non-colicky upper abdominal pain radiating to the back. Abdominal examination was unremarkable. The ultrasound and Computerized Tomography (CT) imaging revealed a 48 mm × 35 mm well-defined, multiloculated cystic lesion involving the pancreatic head. The tumor markers (CA 19-9 and CEA) tested within the expected reference range. Magnetic Resonance (MR) imaging and Endoscopic Ultrasound (EUS) confirmed the cystic nature of the lesion without mural nodules and no vascular involvement or ductal communication. EUS-guided Fine Needle Aspiration Cytology (FNAC) showed the mesenchymal cells of Schwannian origin. The patient underwent open enucleation, with intraoperative frozen section analysis revealed the benign mesenchymal pathology. Postoperative recovery was uneventful, and the patient was discharged on postoperative day six. Postoperative histopathology report confirmed the diagnosis of schwannoma.

Conclusion: Pancreatic schwannomas are exceedingly uncommon benign lesions that deserves an inclusion in the differential diagnosis of the cystic pancreatic space occupying lesions. Diagnosis before surgery is challenging, but preoperative EUS-guided FNAC/Biopsy helps in diagnosis. Treatment of the pancreatic schwannomas requires enucleation or pancreatic resection, both associated with a good prognosis and no reported recurrences.

Keywords: Pancreatic schwannoma; Enucleation; Pancreatic resection; EUS; FNAC; Biopsy

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Correspondence:
Hirdaya Hulas Nag, Department of Surgical Gastroenterology, G B Pant Institute of Postgraduate Medical Education and Research, Delhi University, Room no 220, 2nd floor, Academic block, G. B. Pant Hospital, J. N. Marg, New Delhi, India, E-mail: Hirdayanag@gmail.com

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Introduction

Intra-abdominal and retroperitoneal schwannomas are rare types of peripheral schwannomas. The occurrence of schwannomas in the pancreas is exceptionally rare. Fewer than 80 cases have been reported globally, highlighting their rarity [1]. PS develop from Schwann cells lining the autonomic fibers of the vagus nerve that pass through pancreatic tissue [2]. Clinically, these tumors can present with a wide range of nonspecific symptoms such as abdominal or back pain, nausea, vomiting, jaundice, or weight loss. However, a significant number of patients remain asymptomatic, with the tumor being discovered on imaging done for unrelated concerns. This variability, along with overlapping features with other pancreatic lesions, makes preoperative diagnosis particularly challenging [3,4].

According to a study by Ma et al. [4], the pancreatic head is the most frequently involved part, approximately in the half of all cases. Over half of the PS exhibit tissue degeneration manifesting as cyst formation, calcification and internal haemorrhage. This cystic appearance often leads to diagnostic confusion with more common pancreatic pathologies, including other cystic lesions, pseudocysts, or Hamoudi tumors [2,5]. Radiologically, PS may appear cystic or solid-cystic, and thus deserves an inclusion in the differential diagnosis when evaluating atypical pancreatic masses with solid cystic appearance on imaging [4,5]. Surgery offers curative treatment for PS [3,4]. Here, we describe a case the pancreatic lesion that was ultimately diagnosed as a pancreatic schwannoma.

Case Presentation

A 53-year-old female with systemic hypertension and hypothyroidism presented with a 5-year

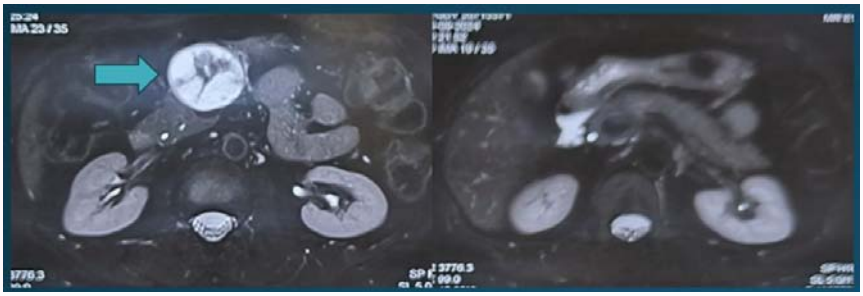

Figure 1: Axial MR Abdomen images showing well defined, mixed intensity lesion (arrow) involving head of the pancreas with central hypointense and peripheral hyperintense area, rest of the pancreas normal.

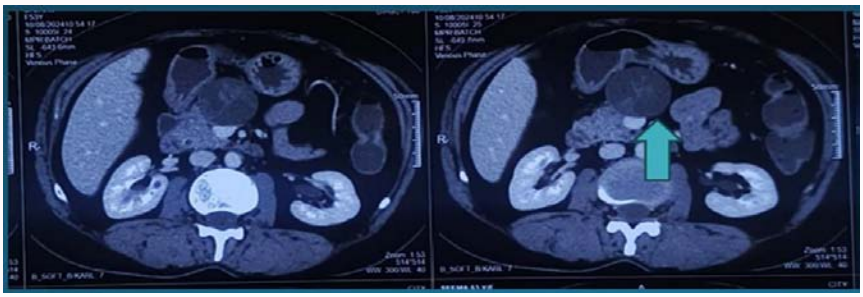

Figure 2: Axial CT scan image showing well defined round solid cystic lesion (arrow) noted involving head of the pancreas.

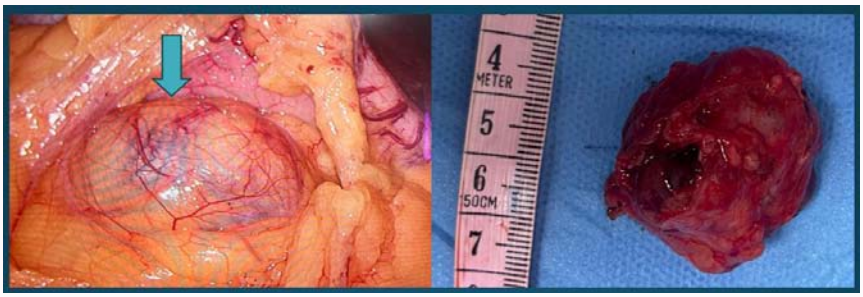

Figure 3: Intra operative image (image on left) showing lesion involving head of the pancreas (arrow), Postoperative image (image on right) showing enucleated schwannoma specimen measuring 4 cm × 4 cm.

history of non-colicky, mild to moderate upper abdominal pain radiating to the back. There were no associated jaundice, fever, or gastrointestinal symptoms. The physical assessment showed no significant findings, and the family history was unremarkable. Laboratory work up, including complete blood picture, liver and renal function tests, revealed no abnormalities.

An abdominal ultrasound showed a clearly defined, hypoechoic mass located in the pancreatic head, measuring 4.2 cm × 4.4 cm. The mass contained a cystic area within and demonstrated no significant intralesional vascularity on doppler imaging. Contrast CT scan of the abdomen (CECT) demonstrated a 48 mm × 35 mm-sized, round, multiloculated cystic lesion with internal septations. Magnetic Resonance Cholangiopancreatography (MRCP) scan of the abdomen revealed a lesion in the head and neck of the pancreas, with thick T2 hyperintense septa but no mural nodules or ductal communication. CA 19-9 (4.7 U/mL) and CEA (<0.31 ng/mL) were within the reference range. EUS confirmed a multiloculated cystic lesion involving the head of pancreas without ductal, vascular involvement or mural nodules. EUS-guided FNAC was performed using a 19-G needle; straw-coloured fluid was aspirated, and cytology of which revealed a mesenchymal lesion with Schwann cell origin.

The patient was taken for open surgery during which a 4 × 4 cm-sized cystic lesion noted involving the pancreatic head. Rest of the pancreas was normal. The lesion was enucleated and sent for intraoperative frozen section analysis which revealed a benign lesion of mesenchymal origin. Postoperative course was uneventful. The abdominal drain was removed on post op day three (POD3) and the patient was discharged on POD4. Final histopathology reports showed spindle-shaped tumour cells with hypercellular (Antoni A) and hypocellular areas (Antoni B). On immunohistochemistry, the tumour cells are stained positive for S-100 and negative for CD117, confirming the diagnosis of schwannoma.

Discussion

PS are exceedingly rare peripheral nerve sheath tumors making up less than 1% of all schwannomas [5]. PS arise from autonomic nerve fibers traversing the pancreas via branches of the vagus nerve [2,3]. They are most frequently located in the pancreatic head, followed by the body and tail, which are involved in approximately 40%, 20%,

and 15% of cases, respectively. Involvement of the uncinate process and neck is uncommon [4,6]. In PS degenerative changes are seen in more than 50% cases, this includes cystic changes, intralesional bleed, and necrosis. This degenerative change leads to cystic appearance of tumors on imaging. This variability often leads to these tumors being confused with other pancreatic space occupying lesions such as cystic neoplasms, pseudocysts, neuroendocrine tumors or Hamoudi tumors [7].

Patients present with abdominal discomfort and nonspecific symptoms. About a one third cases are completely asymptomatic. According to the study conducted by Moriya et al. [3], abdominal pain was the most commonly reported symptom. Larger lesions (especially those >6-7 cm), evidence of vascular encasement, or loss of fat plane with nearby viscera should raise concern for malignant transformation as reported by Ma et al. [4]. The cross-sectional imaging is crucial for establishing the preoperative diagnosis of PS. CT scan typically reveals well-defined, round, hypoattenuating, cystic masses with variable enhancement. MRCP shows cystic lesion with T1 hypointensity, T2 hyperintensity and give information on ductal communication [8]. Endoscopic Ultrasound (EUS) identifies PS as a well circumscribed, homogeneous, anechoic or hypoechoic mass; EUS FNA may reveal spindle cell morphology and confirm the tumor's Schwann cell origin with strong S 100 staining [9]. Serum CEA and CA19-9 level helps in ruling out pancreatic malignant lesions, narrowing the differential diagnosis [7].

Treatment depends on clinical and radiological risk factors. The patient can be managed with a wait-and-see policy if a patient is asymptomatic and preoperative diagnosis is established by EUS FNAC, as highlighted by Bruno et al. [10]. Because most pancreatic schwannomas are benign and encapsulated, enucleation is often sufficient especially if preoperative diagnosis is done via EUS FNA and carries an excellent prognosis with rare recurrence [3,4,11]. More extensive pancreatic resections are reserved for tumor close to the main pancreatic duct, larger tumors, or suspected vascular involvement [3,4]. Following the tumour removal, the long-term outcome is excellent and recurrence is very rare [6].

Conclusion

Pancreatic schwannomas are exceedingly uncommon benign lesions that deserves an inclusion in the differential diagnosis of the pancreatic cystic space occupying lesions. Diagnosis before surgery is challenging, but preoperative EUS-guided FNAC/Biopsy helps in diagnosis. Treatment of the pancreatic schwannomas requires enucleation or pancreatic resection, both associated with a good prognosis and no reported recurrences.

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