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Indu B. Dubey
Vardhaman Mahavir Medical College and Safdarjung Hospital

Sajal Gupta
Vardhaman Mahavir Medical College and Safdarjung Hospital, sajal.dare@gmail.com

Shivani Paruthy
Vardhaman Mahavir Medical College and Safdarjung Hospital

Vimal Bhandari
Vardhaman Mahavir Medical College and Safdarjung Hospital

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Case Report

Solid pseudopapillary epithelial neoplasm of the pancreas: A case report

Sajal Gupta, Vimal Bhandari, Shivani Paruthy, Indu B. Dubey

Department of General Surgery, Vardhaman Mahavir Medical College and Safdarjung Hospital, New Delhi, Delhi, India

Abstract

Solid pseudopapillary epithelial neoplasm (SPEN) of the pancreas is a rare neoplasm of the pancreas accounting for only 0.17–2.7% of all pancreatic neoplasms, often detected initially on imaging. Despite advances in imaging, pseudocysts and other cystic neoplasms feature in the differential diagnosis. We report a case of solid pseudopapillary epithelial neoplasm in a young female who presented with a progressively increasing abdominal lump in whom the diagnosis was considered based on imaging studies. She underwent exploratory laparotomy, pancreaticoduodenectomy, pancreaticojejunostomy, hepaticojejunostomy, gastrojejunostomy, and made an uneventful recovery in the postoperative period. Histopathological examination and immunohistochemistry confirmed the diagnosis. This case highlights the need for a high index of suspicion and timely surgical intervention for optimal outcomes.

Keywords: Abdominal lump, immunohistochemistry, pancreatic neoplasm, surgery

Introduction

Solid pseudopapillary epithelial neoplasm (SPEN) of the pancreas is a rare cystic neoplasm representing 0.17–2.7% of all pancreatic cancers seen predominantly in women (female: male = 10: 1) [1,2].

The history of SPEN or Frantz tumor dates back to its first description in 1933 and official recognition in 1959 [2]. Although seen in significant numbers in the age group of the 30s and 40s, recently, this has become commoner in the younger-age group of 20s and pediatric population (≤20 years). The number has been on the rise in the last decade because of the advancements in immunohistochemistry and antibodies [2]. As against the adult prevalence rate of upto 3%, SPEN accounts for 8–12.5% of pancreatic neoplasms in children [3]. Upto 9000 cases have been reported in the literature so far [2].

The presentation is variable, ranging from completely asymptomatic to gradually enlarging lump producing symptoms like early satiety or obstructive jaundice on account of compression of the stomach, duodenum, or bile duct, intermittent abdominal pain, dyspepsia, loss of appetite, nausea, and vomiting [1,2].

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In the majority, the diagnosis is made incidentally when imaging is performed for upper-abdominal symptoms or lump on account of characteristic features on ultrasonography (USG) and computed tomography (CT) scan [4], necessitating the need of suspicion of this growing problem in patients presenting with abdominal symptoms, especially females.

We hereby report a case of a 23-year-old lady who presented to the surgical department with abdominal symptoms, and the diagnosis of SPEN was considered on the basis of imaging studies.

Case Report

A 23-year-old lady presented to the outpatient department with a history of abdominal lump and progressively

Correspondence to: Sajal Gupta, MS, Department of General Surgery, Vardhaman Mahavir Medical College and Safdarjung Hospital, New Delhi 110029, India.
Tel: +91 956 076 5429;
E-mail: sajal.dare@gmail.com

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increasing distension of 4-year duration. She also complained of occasional dull aching pain in the epigastrium, early satiety, and anorexia. Past history was significant for jaundice that resolved after medication from a local practitioner. Family history and general physical examination were unremarkable.

On abdominal examination, there was a well-defined intra-abdominal lump of size 15 × 15 cm occupying the epigastric, right hypochondriac, right lumbar, and umbilical region. The lump was nontender, nonpulsatile, firm in consistency, and mobile from side to side but not with respiration.

USG revealed a well-defined intra-abdominal heterogeneous lesion of size 12 × 15 × 17 cm anterior to the inferior vena cava and aorta. A large well-defined solid cystic mass lesion in the region of head, neck, and uncinate process of the pancreas causing compression and displacement of the surrounding structure and obstructive focal dilatation of the main pancreatic duct suggestive of SPEN was found on contrast-enhanced CT scan.

On MRCP, a well-defined encapsulated solid cystic mass lesion of size 16.6 × 14 × 10 cm was seen in the right hypochondriac region compressing the second part of the duodenum, causing posterior displacement of suprarenal and infrarenal inferior vena cava and anterolateral displacement of the superior mesenteric vein and portal vein. The pancreas was not visualized separately. The gall bladder was appreciated separately from the mass lesion. The common bile duct was seen anterolateral to the mass lesion and is separate from the mass lesion (Figs. 1 and 2).

Surgical exploration was performed in view of a possible neoplasm following informed consent. Intraoperatively, a 15 × 20-cm mass was seen arising from the head and uncinate process of pancreas with mixed solid and cystic components (Fig. 3). Dense adhesions were found with adjoining structures. Pancreaticoduodenectomy with resection of the lesion was accomplished. Gastrointestinal and hepatobiliary continuity was established by pancreaticojejunostomy, hepaticojejunostomy, and pylorus-preserving retrocolic gastrojejunostomy.

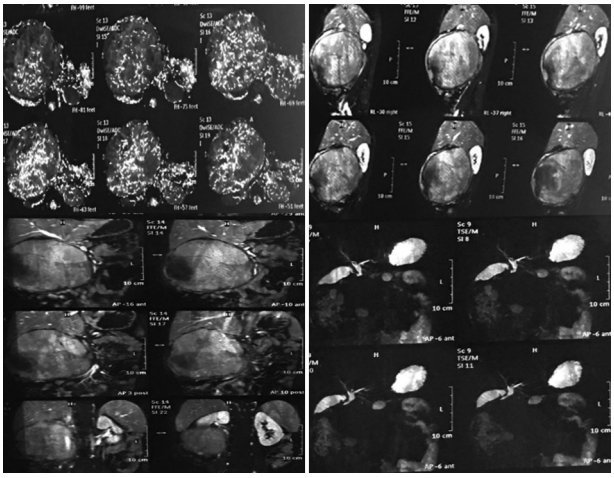

Figure 1: Well-defined encapsulated solid cystic mass lesion of size 16.6 × 14 × 10 cm. GB is seen separate from the mass lesion. The RHD, LHD, CHD, and CBD appear mildly prominent with central IHBRD. CBD is seen anterolateral to mass lesion and is separate from mass lesion. GB: gall bladder, RHD: Right hepatic duct, LHD: Left hepatic duct, CBD: Common bile duct, CHD: Commmon hepatic duct, IHBRD: Intra Hepatic Biliary Radical Dilatation.

On gross examination of the resected specimen, both solid and cystic areas containing brownish necrotic material were seen (Figs. 4 and 5). Histopathological examination revealed cells arranged as solid sheets in the pseudopapillary pattern (Fig. 6). Immunohistochemistry showed strong positivity for vimentin, progesterone receptor, CD-10, and CD-99 of tumor cells, thereby establishing the diagnosis of SPEN pancreas. The patient made an uneventful recovery in the postoperative period and has not shown any recurrence during 6 months of follow-up.

Discussion

Our patient was a young-aged female, which corroborated with the previous reports of young-age presentation in the case series by Cantisani *et al*. [5] (median age, 20 years) and case reports by Mujtahedi *et al*. [1] (18-year female), Hegde *et al*. [6] (16-year-old female), and Rivera *et al*. [4] (14-year-old female). Besides, studies have also shown predominance of pediatric-age group (median age, 17 years) [7] and elderly age group of 50 [8] and 63 years [9].

Although these tumors usually have a torpid benign course, 10–15% of all cases may be malignant with an aggressive potential [10,11].

Aggressive behavior of malignancy may be set by features like size more than 5 cm, vascular or perineural infiltration, nuclear atypia, invasion of surrounding structures, and high proliferation rate. In terms of age, pediatric-age group (≤21 years) has shown better prognosis as compared with adults with elderly showing the worst prognosis with increasing metastatic potential [1,9,12].

There are many theories on what causes these tumors, however, the tumor biology remains unpredictable and still an enigma [12]. Some say multipotent primordial cells cause SPEN; other opinionated tumors have an extrapancreatic origin from cells related to genital ridge angle [12]. It is suggested that the pancreatic pluripotent embryonic cells with multipotent differentiation are responsible for its genesis. However, there is no confirmation for terminal differentiation for both the endocrine and acinar cells. The origin from primitive ovarian cells or stem cells within the pancreatic parenchyma are examples of many other theories. There are two opponents of the theory of stem-cell origin. One is low malignant potential, and the other is slow growth. Men presenting with these tumors suggest a flaw in the idea that it arises from primitive ovarian cells [12].

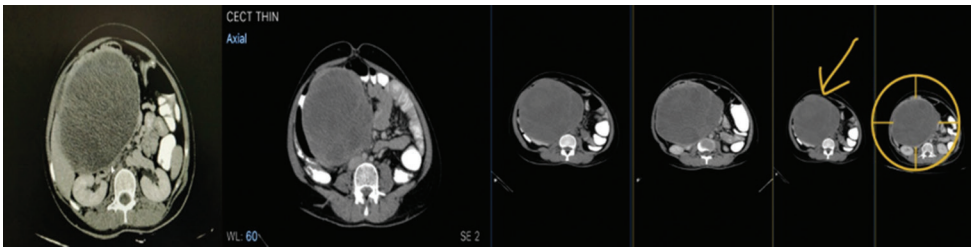

Figure 2: Well-defined solid cystic mass lesion in the region of head, neck, and uncinate process of pancreas.

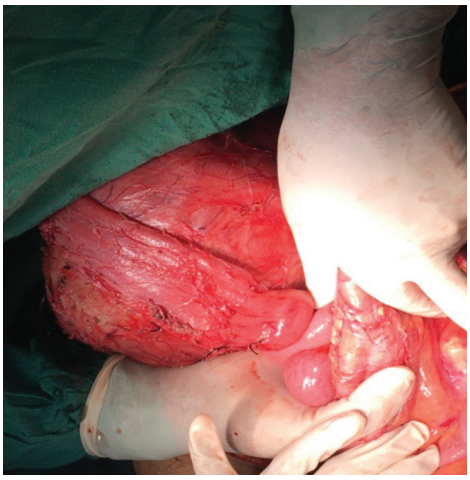

Figure 3: Intraoperative photograph.

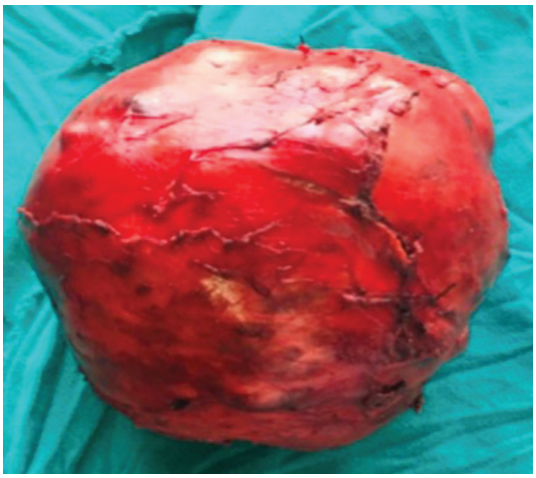

Figure 4: Resected lump.

These tumors mainly occur in the body and tail of the pancreas (55–60% cases) with the other common site being head of pancreas (35–40%). Seldom, these tumors are seen in the mesentery, left adrenal gland, and behind the peritoneum [3,13].

Steadily increasing abdominal mass, recurrent pancreatitis, and abdominal pain are standard clinical features. The symptoms vary depending on the site of compression by large tumor, like compression on the stomach causes abdominal pain, vomiting, and early satiety, whereas bile-duct compression leads to obstructive jaundice. Abdominal pain (present in 80% of patients) is a standard and nonspecific symptom [1,2], which was seen in the present case.

The diagnosis is usually suspicioned on imaging and confirmed on histopathology [14]. Biomarkers like amylase, CA19-9, CA 242, CEA, and CA125 remain nonspecific [2].

The heterogeneous appearance is caused by cystic and solid areas in the tumor, which look like encapsulated lesions with cystic (centrally located) and solid (peripherally located) elements on CT. Neoplasms appear to show hypointense fibrous capsules on high-intensity T1-weighted scans, which is the chief MRI feature. MRI is preferred over CT to demonstrate the existence of a solid, capsule, and cystic degeneration. Also, bleeding without apparent internal septum (a strong indication of solid pseudopapillary neoplasm) is better in MRI. Interventions like endoscopic USs, preoperative fine-needle aspiration cytology, and endoscopic retrograde cholangiopancreatography are rarely performed for the diagnosis. Positron-emission tomography is not required in these cases because of its nonmalignant nature mostly [12].

The differential diagnosis includes pancreatic cyst/pseudocyst, adenocarcinoma, cystadenocarcinoma, neuroendocrine tumor, cystadenoma, islet-cell tumor, or teratoma, which poses difficulties in making a preoperative diagnosis based on imaging alone. Histopathology and immunohistochemistry are the final stages for confirmation [15].

Pseudopapillary appearance and cellular degeneration are characteristic microscopic features. A tissue tumor section consists of epithelial cells (with minimal atypia forming pseudorosettes) and pseudopapillae (with cystic breakdown). SPEN presents nuclear and cytoplasmic ß-catenin immunoreactivity, and loss of membrane staining for E-cadherin (due to activation of the Wnt-signaling pathway) in almost all of the cases. Besides, there is progesterone receptor+, androgen receptor + (80% cases), and estrogen-receptor negativity. There may be a significant overlay in other markers like α1-antichymotrypsin, NSE, α1-antitrypsin, synaptophysin, carcinoembryonic antigen, pan CK, vimentin, cyclin D, CD-10, and CD56 [12]. To further elaborate the IHC profile, TFE3 is suggested, which is positive in 94% cases [12].

American Joint Committee on Cancer (8th edition) is the gold standard used for pancreatic tumor staging. However, none

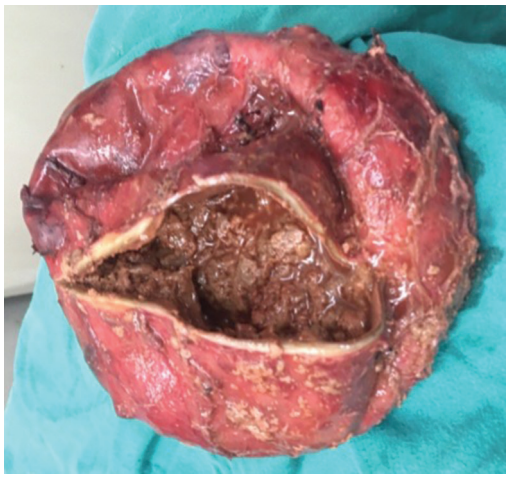

Figure 5: Cut specimen showing solid and cystic areas.

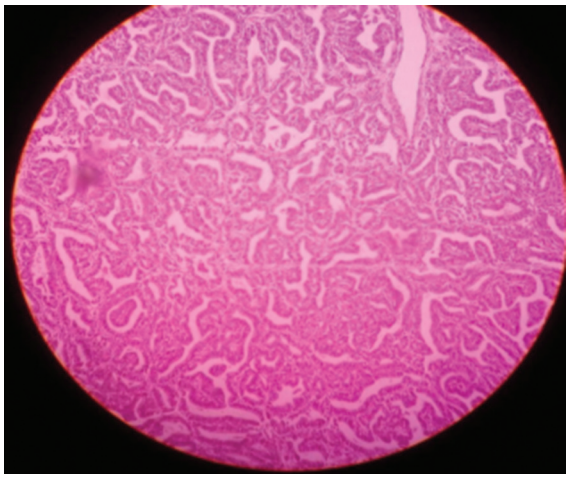

Figure 6: Histopathological view showing pseudopapillary pattern with pseudorosettes and pseudopapillae with areas of cystic breakdown pointing toward SPEN. SPEN, solid pseudopapillary epithelial neoplasm.

of the staging methods have been approved to date, mainly because the infiltration of the superior mesenteric artery or portal vein (constitutes T4 stage and is the criteria for pancreatic tumors not resectable) is impossible for pancreatic solid pseudopapillary tumors. Furthermore, it is unfeasible to compare American Joint Committee on Cancer staging for prognosis and survival rates for these tumors (because of its rarity) [16].

Unvariably, extensive surgical resection is advised in all the cases of SPEN, leading to a disease-free survival rate of 95% [1,2]. Only 5% of the cases have shown metastatic potential, due to which surgical resection is advised. Pancreaticoduodenectomy or distal pancreatectomy is commonly performed, with en bloc resection of involved adjacent organs. About 10–15% of patients already have metastases before the initial diagnosis or may develop them in the future [9]. Typical metastatic sites are the lymph nodes, peritoneum, liver, and mesentery [2]. Monitoring of recurrence in such cases demands long-term follow-up like upto 13 years in a study by Gurzu *et al.* [2]. In cases with metastasis, chemoradiotherapy with 5-fluorouracil, gemcitabine, and cisplatin, has been used, but the outcomes remain grave after metastasis [17].

Conclusion

This case highlights the need of high index of clinical suspicion to diagnose SPEN, especially in young females presenting with abdominal complaints. Although imaging studies (USG, CT scan, and MRI) provide a clue toward the diagnosis in the preoperative period, but definitive diagnosis rests on histopathological and immunohistochemical analysis. Surgical excision not only helps the pathologist and surgeon in reaching the definitive diagnosis, but also offers the best chance for cure to the patient and should always be attempted, irrespective of the magnitude of resection involved.

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Conflicts of interest

There are no conflicts of interest.

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