Case Report

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Spontaneous ruptured solid pseudopapillary tumor of pancreas: a rare condition identified in a patient presenting with features of shock

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Introduction: Solid pseudopapillary tumor of pancreas are rare benign tumors of pancreas, that typically affects the young women. They are usually asymptomatic or minimally symptomatic, and are usually diagnosed by different imaging modalities.
Case presentation: We herein present a case of solid pseudopapillary tumor of pancreas in an 18-year-old female patient who presented to our emergency department with features of shock following its spontaneous rupture. Imaging showed features suggestive of solid pseudopapillary tumor of pancreas, and she was managed by exploratory laparotomy following initial resuscitation.
Discussion: Solid pseudopapillary tumors of pancreas are one of the rare exocrine pancreatic tumors. Their origin is not exactly clear, the most accepted theory states that these tumors originate from the multipotent primordial cells. Preoperative diagnosis can be made showing features distinct from other pancreatic solid/cystic neoplasms. Surgical management is the preferred treatment modality in these tumors.
Conclusion: These are rare entities of pancreas, and may present with vague abdominal symptoms. Sometimes, these tumors may undergo spontaneous rupture, presenting with features of shock.

Keywords: case report, pancreatic tumor, shock, solid pseudopapillary tumor of pancreas, spontaneous rupture

Introduction

Solid pseudopapillary tumors (SPT) of pancreas are relatively rare primary neoplasms of pancreas, relatively benign in nature, and typically affect young women[1]. These tumors are usually characterized by the presence of encapsulated mass, with low malignant potential[2]. These tumors are usually asymptomatic or minimally symptomatic and may present with vague and nonspecific abdominal symptoms[1,3]. Multiple imaging modalities like transabdominal ultrasonography (USG), computed tomography scan (CT scan), or magnetic resonance imaging (MRI) can be used to visualize these tumors, as well as to differentiate these tumors from other pancreatic pathologies[1]. Since these tumors are usually limited to the pancreas at the time of diagnosis, complete surgical excision is the curative treatment in most cases[4].

This paper, following the SCARE 2023 guidelines, describes a case of a solid pseudopapillary tumor of pancreas in an 18-year-old female patient who presented to our center with abdominal pain[5].

Case presentation

An 18-year-old female patient presented to our emergency department with complaints of generalized abdominal pain which was sudden onset, dull aching, and continuous without any relieving and aggravating factors. She also had 2-3 episodes of vomiting in the last 5 days. There was no history of fever, presence of blood in stool, or blood in vomitus. Her bowel and bladder habits were normal.

On examination, her vitals were as follows: blood pressure of 100/70 mm/Hg, pulse rate of 110 beats/minute, respiratory rate of 20 breaths per minute, spO2 94%, and body temperature of 36.5°C. She was conscious and her general condition was fair. She was hemodynamically unstable at the time of presentation with physical findings of pallor and dehydration. On per abdominal examination, generalized tenderness was present however there was no guarding and rigidity. Bowel sounds were sluggish on auscultation. The rest of the systemic examinations were normal.

Her baseline blood and urine examinations were done which revealed an increased total leucocyte count of 22 000/mm^3 (4000–11 000/mm^3) and decreased hemoglobin of 8.8 g/dL (12–15.5 g/dL for females). Other parameters were within normal limits.

On transabdominal ultrasonography, a well-defined heteroechoic mass was seen in the body and the tail of pancreas, suggestive of a solid pseudopapillary tumor of pancreas.

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On a CT scan of abdomen, a well-defined hypodense non-enhancing complex cystic lesion (HU 30–70) was noted in the body and tail of pancreas, with minimum surrounding mesenteric edema. The features were suggestive of a pseudopapillary tumor of pancreas. The computed tomography scan is shown in Fig. 1.

After resuscitation in emergency department, she was planned for exploratory laparotomy. Necessary pre-operative investigations were done, and then the patient was operated under general anesthesia. Intraoperative findings of about 500 mL hemoperitoneum with 10 × 10 cm well-defined round mass with smooth surface arising from body and tail of pancreas with abutment of visceral surface of spleen were noted. There was a capsular tear of mass on superior surface sealed with clotted blood. Distal pancreatectomy with splenectomy was done, and abdominal drain was kept. The resected specimen was sent for histopathologic examination. The histopathologic images of the specimen in different magnifications are shown in Figs. 2 and 3. The patient was kept in the post-operative ward for 4 days. Drain amylase was not significantly raised in 1st, 3rd,

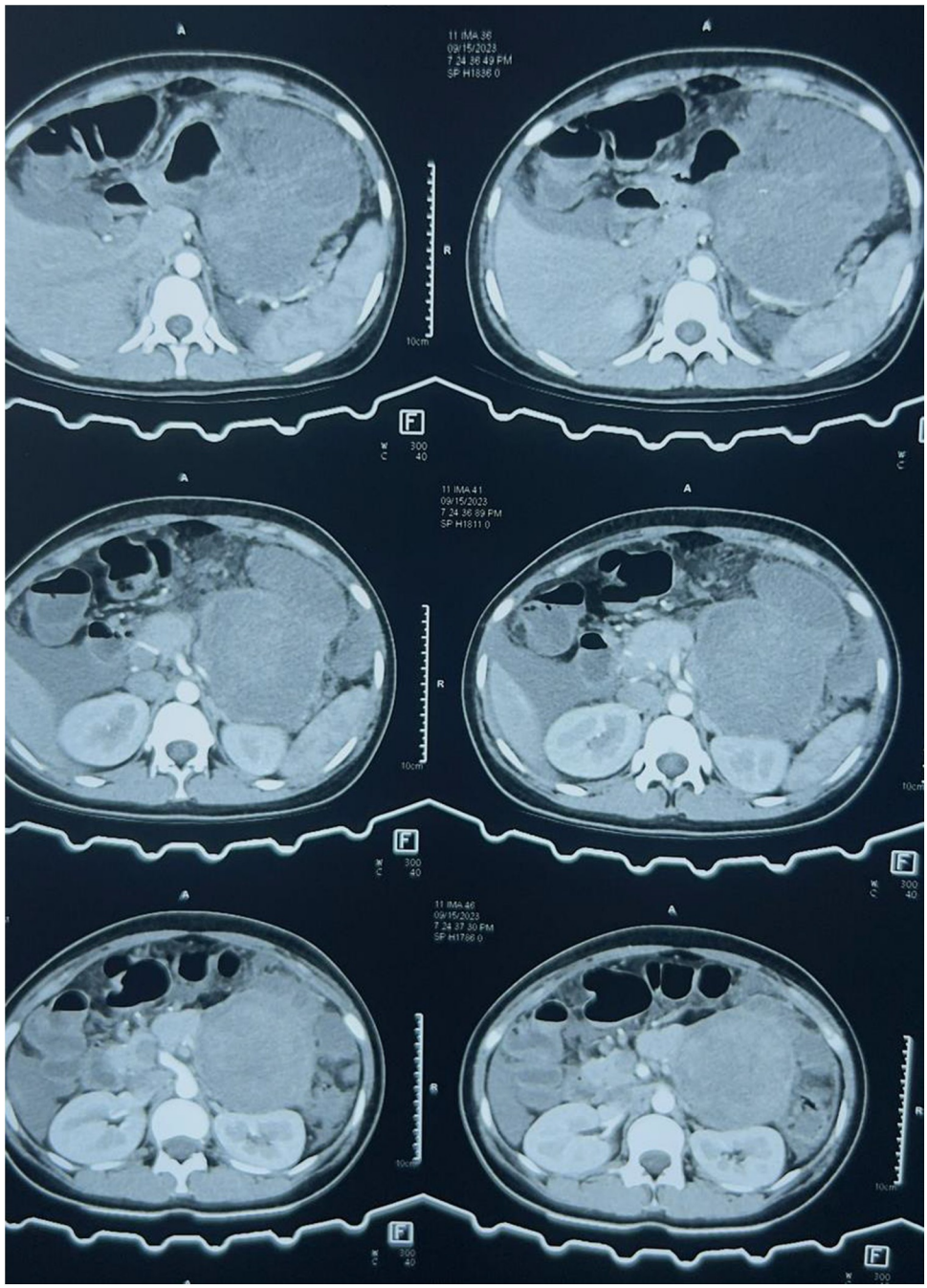

Figure 1. Computed tomography scan of abdomen showing solid pseudopapillary tumor of pancreas.

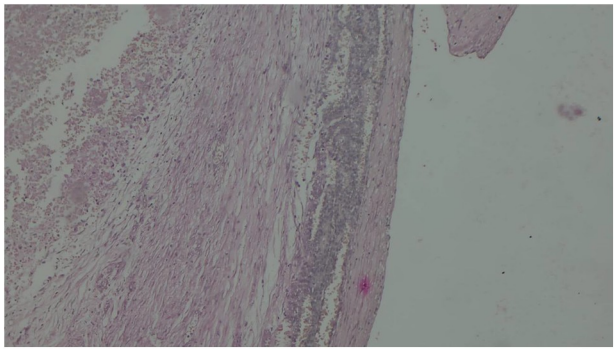

Figure 2. Histopathologic image of the resected specimen (solid pseudopapillary tumor of pancreas) at low power.

and 5th post-operative day. She was shifted to general ward on 5th postoperative day. Dressing of the incision site was done on every alternate day, and the abdominal drain was removed on 10th postoperative day.

The resected specimen was sent for histopathologic examination. On gross examination, the specimen was part of pancreas (body and tail) and the spleen. Cut section of pancreas showed gray brown to dark brown solid fragile mass measuring 10 × 6 × 5.8 cm. On microscopic examination, sections examined from the pancreas show an encapsulated tumor mainly showing hemorrhage and large area of necrosis. Locally viable tumors are composed of tumor cells arranged in solid sheets and papillae. Tumor cells have moderate amount of eosinophilic cytoplasm, round to oval nucleus, and granular chromatin. Tumor cells exhibit a mild degree of nuclear pleomorphism. Mitotic figures are infrequent. The overall histopathologic diagnosis was made as a solid pseudopapillary neoplasm of pancreas, of size 10 cm, present at the body and tail of pancreas. There was no evidence of lymphovascular invasion and perineural invasion, and the resection margin was free of tumor. The TNM staging was pT2N0.

A 6 months follow-up of the patient was ensured well, and she showed a satisfactory recovery with no significant post-operative complications and stable vital signs. She steadily improved in overall health, due to regular monitoring and diligent care, leading to a smooth transition from the post-operative ward to the general ward and her eventual discharge in good condition.

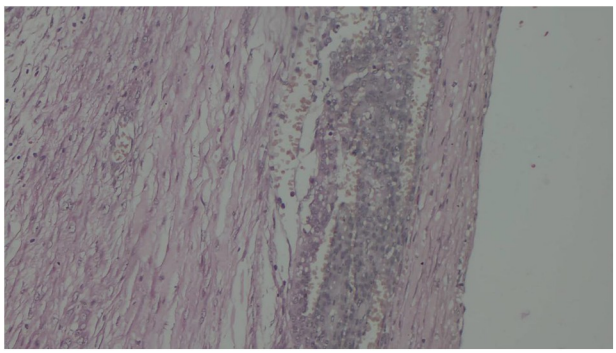

Figure 3. Histopathologic image of the resected specimen (solid pseudopapillary tumor of pancreas) at high power.

Discussion

Solid pseudopapillary tumors of pancreas are also known as the "Frantz tumor," named after the author who first described these in 1959, under the name "papillary tumors of pancreas: benign or malignant"[2,6]. These tumors were included in classification of pancreatic tumors by WHO in 1996 under the name "solid pseudopapillary tumors (SPT) of pancreas"[6]. Solid pseudopapillary tumors of pancreas are the rare exocrine pancreatic tumors that accounts for only about 1% of all the tumors of pancreas[1].

Though their origin is not exactly clear, the most accepted theory states that these tumors originate from the multipotent primordial cells, whereas some theories suggest extra pancreatic origin (from the genital ridge angle-related cells)[7]. It typically originates from the body and tail of the pancreas, as observed in our patient. Although it is generally benign, approximately 10% of solid pseudopapillary tumors can be malignant, potentially showing local invasion and distant metastasis to organs like the liver and lungs at the time of diagnosis[8].

These tumors have predilection for young Asian and African-American females, and the male-to-female ratio is 1:10. These tumors commonly occur between 2nd and 3rd decade of life, and the mean age of presentation is 22 years[2,9].

Though majority of the patients with solid pseudopapillary tumors of pancreas are asymptomatic, some may present with vague abdominal pain/discomfort, or a gradually enlarging abdominal mass. Some patients may present with obstructive features if the tumor is large enough to compress the adjacent structures[2]. Nausea, vomiting, and jaundice, though rare, can be the initial signs of a solid pseudopapillary tumor of the pancreas, occasionally accompanied by spontaneous tumor rupture[8]. In our case, the patient presented to emergency department, with abdominal pain following blunt abdominal trauma.

On imaging, these tumors usually present as a large cystic mass (larger, as compared to other pancreatic neoplasm), and they are often confused with a pancreatic pseudocyst or an intraabdominal cyst[2]. Evaluation of large SPTP on imaging often shows large well-circumscribed mass, with central cystic degeneration, and sometimes the capsular calcification. Endoscopic ultrasonography can allow better visualization of these tumors, but the findings are not specific[4]. In our patient, transabdominal ultrasonography, and computed tomography scan were done. However, according to Cantisani *et al*, MRI is superior to CT for differentiating specific tissue characteristics, including hemorrhage, cystic degeneration, and the presence of a capsule. This may be evidenced by high signal intensity on T1-weighted images and heterogeneous peripheral contrast enhancement, which becomes more pronounced after gadolinium administration during dynamic imaging[10].

Given the good prognosis of the disease, it is important to make the diagnosis preoperatively if possible so that adequate resection will be undertaken. Preoperative diagnosis of these tumors can be made by ultrasonography (transabdominal or endoscopic) or CT-guided fine needle aspiration cytology, which shows features distinct from other pancreatic solid/cystic neoplasms. Histological pictures include numerous isolated capillaries, foam cells, cellular debris, psammoma bodies, and uniform arrangement of neoplastic

cells around small blood vessels in a manner similar to papillary structure, so-called "pseudopapillary pattern"[2].

On immunohistochemistry, solid pseudopapillary tumors of pancreas are usually positive for alpha-1 antitrypsin, alpha-1 antichymotrypsin, neuron-specific enolase, vimentin, and progesterone receptor. These tumors are usually negative for hormone peptides, and chromogranin. Though these tests can be used to differentiate solid pseudopapillary tumor of pancreas from other pancreatic tumors, this investigation was not done in our patient, because of limited resources, and financial constraints[2].

SPT can develop in any part of the pancreas and generally have a favorable prognosis following various treatment options, as they exhibit characteristics similar to a very low-grade malignancy[11]. Surgical management is the preferred treatment modality in these tumors, even in those having distant hepatic metastasis or local recurrences[12]. The type of surgery performed will be determined by the tumor's anatomical location and its extent. Possible procedures include tumor enucleation, partial pancreatectomy, or pancreaticoduodenectomy, depending on the site, size, and characteristics of the tumor[8]. Distal pancreatectomy should be recommended only when the tumor occurs in the body or tail with invasion into the adjacent parenchyma as in our patient[11].

SPT is considered to be a low-grade malignancy and a good prognosis is expected after the surgery. However, our patient who is alive without recurrence after surgery for 6 months should continue to be followed up carefully.

Conclusion

Solid pseudopapillary tumors of pancreas are rare entities that may be asymptomatic or present with vague abdominal symptoms. These tumors can spontaneously rupture and thus present with the features of shock. If identified early, complete surgical excision is the preferred treatment modality.

Ethical approval

Since this is a case report, our Institutional Review Board of Institute of Medicine, Tribhuvan University has waived the requirement for ethical approval

Consent

Written informed consent was obtained from the patient's parents 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.

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Author's contribution

Concept of study, data acquisition: S.D., K.A., N.P.; Literature review and manuscript preparation: A.S., K.A.; Performing the surgical procedure and management of the patient: S.D., A.B., P. K. All the authors individually did the final proof-reading of the manuscript before submission.

Conflicts of interest disclosure

All the authors declare to have no conflicts of interest relevant to this study.

Research registration unique identifying number (UIN)

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Data availability statement

The datasets used during this study will be available from the corresponding author upon reasonable request.

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