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Solid pseudopapillary neoplasm of pancreas presenting as acute abdomen: A rare case report

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Abstract

Introduction: Solid pseudopapillary neoplasm (SPN) is a rare tumor, first described by Frantz in 1959. It commonly presents in a young female with abdomen pain or as a mass per abdomen. It may be asymptomatic. Spontaneous rupture of SPN is a rare presentation.

Case presentation: Here we present a case of acute abdomen due to rupture of SPN in a nineteen year old girl for its rarity. She underwent distal pancreatico-splenectomy. She had an uneventful postoperative period.

Conclusion: SPN is an uncommon neoplasm of young females. They usually present with mass per abdomen or abdomen pain. Rupture of SPN is a rare form of presentation.

Keywords: Solid pseudopapillary neoplasm; Frantz tumor; Cystic neoplasm of pancreas; Tumor rupture.

Abbreviations: SPN: Solid Pseudopapillary Neoplasm; CT: Computed Tomography; MRI: Magnetic Resonance Imaging.

Introduction

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare cystic neoplasm of the pancreas representing 1-2% of all pancreatic neoplasms [1]. It is usually characterized by an encapsulated mass with high chance of intratumoral hemorrhage and with low malignant potential. It occurs predominantly in young females during the second and third decade of life. The female to male incidence ratio is (9.78:1) [2]. These patients present with abdomen pain, distension and poor appetite. Spontaneous rupture is one of the rare presentations and here we would like to present one such case.

Case presentation

A 19 year old girl presented to the emergency room in our hospital with a history of abdominal pain for 3 days which suddenly increased in severity for 6 hours. The pain which was initially in the upper abdomen suddenly changed to involve the whole of the abdominal. On examination she was found to be pale, tachycardic and hypotensive. There was generalized abdominal tenderness and guarding. She was resuscitated with IV crystalloids and blood products as her hemoglobin was 7.7 g%. A contrast enhanced CT scan of abdomen was done which revealed a cystic mass lesion in the retroperitoneum abutting the pancreatic tail, spleen and stomach anteriorly with retroperitoneal hematoma, moderate intraperitoneal free fluid (Figure 1).

In view of the above findings a diagnosis of tumor rupture was made and emergency exploratory laparotomy was planned. On opening the abdomen around 1L of serous peritoneal fluid was drained. A cystic mass of size 16 x 8 cm was seen arising from the tail of the pancreas with splaying of the splenic vessels and closely adherent to the spleen (Figure 2). Hematoma was found along the posterior aspect of the lesion (Figure 3).

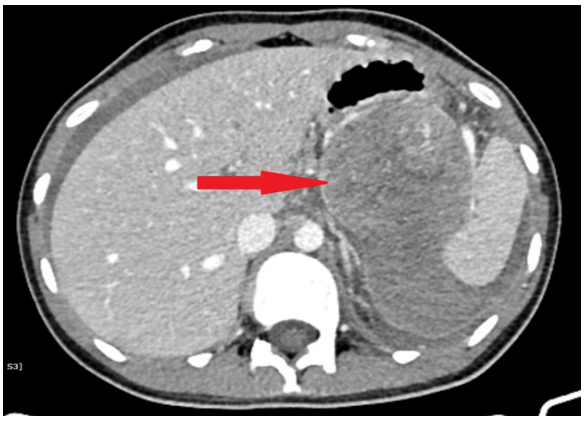

Figure 1: Axial CT images showing the lesion.

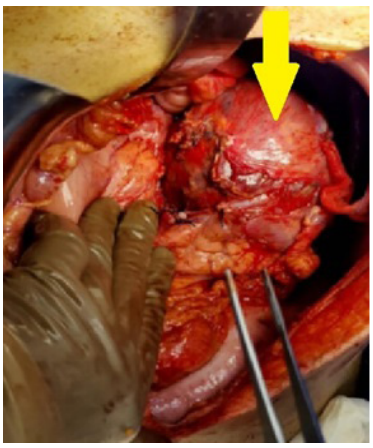

Figure 2: Intraoperative picture of the lesion arising from the pancreatic tail.

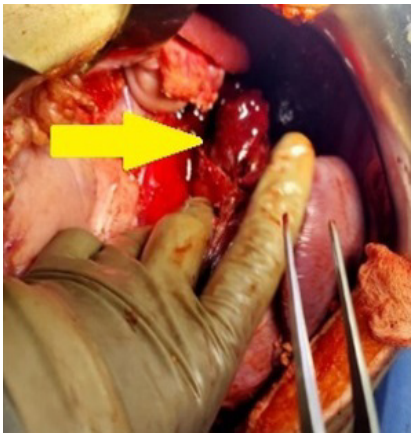

Figure 3: Hematoma seen when the lesion mobilized anteriorly.

Thorough peritoneal lavage given and distal pancreato-splenectomy was performed. Patient had an uneventful postoperative period and was discharged on postoperative day 5.

Final histopathological examination was suggestive of a diagnosis of SPN (Figure 4). No evidence of malignancy was noted. The patient is on regular follow up every 6 months where we are evaluating her with ultrasound imaging of the abdomen to look for recurrence.

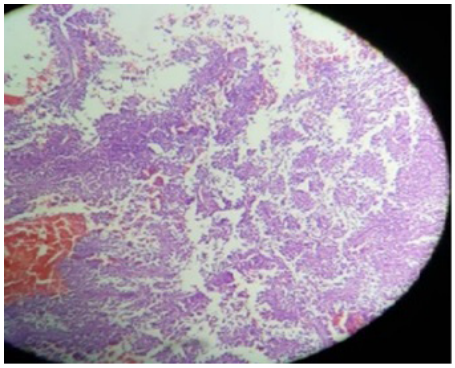

Figure 4: Histopathological picture of the lesion.

Discussion

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare pancreatic tumor, which represents less than 4% of all resected tumors of the pancreas [3]. It was first described by Frantz in 1959. SPN was called by different names like papillary cystic neoplasm of pancreas, Hamoudi tumor until it was defined by the World Health Organization (WHO) in 1996 as a "solid pseudopapillary tumor" of the pancreas [4]. The origin of solid pseudopapillary tumors is unclear. Many investigators favor the theory that SPNs originate from multipotent primordial cells, whereas others suggest an extrapancreatic origin, from genital ridge angle-related cells [5]. Its pathogenesis involves the activation of Wnt-β-catenin, Hedgehog and Androgen signaling pathway. There is also an increased expression of CTNNB1 protein and other proteins that belong to the Wnt-β-catenin pathway (i.e., DKK4, Fine needle 1, SELENBP1, DDX5, YWHAZ, NONO and Fused in sarcoma) [6,7].

This rare tumor seems to have a predilection for young Asian and African-American women [8]. The mean age at presentation is 22 years. It is frequently asymptomatic or minimally symptomatic [9]. The most common symptom is abdominal pain, occurring in 65% of cases. Other symptoms can be in the form of nausea, vomiting or a palpable abdominal mass. Jaundice is seen in 10.3% of patients [10]. About 15%- 19% of cases are asymptomatic [2,11] and discovered incidentally on imaging for other clinical reasons. The presence of cystic lesions with solid components and hemorrhage without internal septation on computed tomography (CT) or magnetic resonance imaging (MRI) is characteristic for SPN [12].

Complete resection is curative in most cases [2]. Despite the locally aggressive features and acute presentation, the tumor has a low-grade malignant potential and tends to have a favorable prognosis, even in the presence of metastatic disease. Overall 5-year survival is as high as 97% in patients undergoing surgical resection [13]. Neither vascular, or perineural invasion has been a factor for predicting tumor recurrence or overall survival of patients [14]. Surgery is the treatment of choice, even in the case of distant hepatic metastasis or local recurrence [15].

Spontaneous rupture of SPN is a rare presentation which has been reported in the literature. Takanori K had published a case of successful resection of recurrent SPN which had ruptured in the index surgery [16]. Shih-Chiang Huang had published a case report of a ruptured SPN during pregnancy [17]. Susumu T had published a report of spontaneous rupture of SPN which was

resected by laparoscopy [18]. Xiaofeng Xu had also reported a case of spontaneous rupture of SPN [19]. Barbara R had reported a case of ruptured SPN and elaborated on 16 cases of ruptured SPN from 1984 to 2017 [20].

Does the rupture of SPN lead to increased risk of recurrence? High risk features like large tumor size, lymphatic and vascular invasion were found to be associated with high recurrence [21,22]. Takanori K reported a case of recurrence six years after SPN rupture which was successfully resected [16]. There is no conclusive evidence from large volume studies to say that SPN rupture is being associated with increased risk of recurrence, however being a rare form of presentation we suggest close follow up of these patients in order to identify and treat recurrence early.

Conclusion

Pancreatic pseudopapillary neoplasms are rare with malignant potential. They may be asymptomatic or present as an abdominal mass or rarely, with jaundice. Spontaneous rupture is not a common presentation which requires emergency resection to improve long term outcome. These patients should be under regular follow up so that early identification of recurrence can offer complete resection.

Declarations

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Conflict of interests: The authors declare that there is no conflict of interest regarding the publication of this article.

Informed Consent: Patient has been informed regarding publication of this article and consent has been obtained for the same.

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