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The first case reported in Southeast Asia of Huge Frantz's Tumor

El primer caso reportado en el Sudeste Asiático de tumor de Frantz enorme

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SUMMARY

Introduction: *Solid pseudopapillary pancreatic tumor (Frantz's Tumor) is a low-grade malignancy, that lacks clinical symptomatology. It was treatable with a favorable prognosis.*
Case presentation: *A young female patient presented with increasing abdominal pain three years ago. Abdominal examination revealed a lump palpable. Contrast-enhanced Computed Tomography abdomen reported a solid mass measuring 13.9 cm x 13.2 cm x 8.9 cm from the pancreas with an enhancing cystic component. Fine-needle aspiration biopsy yielded a cellular sample comprising pseudopapillary neoplasm of the pancreas. A distal pancreatectomy was performed; she was discharged in satisfactory condition.*
Conclusion: *A multidisciplinary team improves treatment accuracy and effective management. Prognosis over 95 % cure rate, so it is important to distinguish it from other pancreatic neoplasms.*

Keywords: *Solid pseudopapillary pancreatic tumor, Frantz's tumor, Diagnosis, Prognosis.*

RESUMEN

Introducción: *El tumor sólido pseudopapilar de páncreas (Tumor de Frantz) es una neoplasia maligna de bajo grado, sin sintomatología clínica. Era tratable con pronóstico favorable.*
Presentación del caso: *Una paciente joven se presentó con dolor abdominal cada vez mayor desde hace tres años. El examen abdominal reveló un bulto palpable. La tomografía computarizada de abdomen con contraste reportó una masa sólida de páncreas de 13,9 cm x 13,2 cm x 8,9 cm con un componente quístico realzado. La biopsia por aspiración con aguja fina arrojó una muestra celular que comprende una neoplasia pseudopapilar de páncreas. Se realizó una Pancreatectomía distal; la paciente se dio de alta con condiciones satisfactorias.*
Conclusión: *Un equipo multidisciplinario mejora la precisión del tratamiento y el manejo efectivo. Pronóstico superior al 95 % de tasa de curación, por lo que es importante distinguirlo de otras neoplasias pancreáticas.*

Palabras clave: *Tumor sólido pseudopapilar de páncreas, tumor de Frantz, Diagnóstico, Pronóstico.*

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INTRODUCTION

A solid pseudopapillary pancreatic tumor (SPPT) also known as 'Frantz tumor' is a very rare neoplasm of the pancreas. It was first described in 1959 by pathologist Virginia Kneeland Frantz (1). The term SPPT was introduced in 1996 by the World Health Organization (WHO) for the International classification of tumors of the exocrine pancreas (2). Although SPPT classification as an epithelial pancreatic tumor, in many cases.

Commonly misdiagnosed as a pancreatic pseudocyst and curative with total surgical resection. However, it was not recognized by the WHO as a low-grade epithelial malignant neoplasm until 2010 (2). Although the 5-year survival rate of more than 90 %, some cases may be locally aggressive; infiltrative with local recurrence, or distant metastases to the liver, lung, and skin (3).

Degenerative cystic changes and hemorrhagic areas are typical. On histological examination, the tumor is a solid mass with pseudopapillary and pseudocystic structures with rich microvascular in variant proportion (4). With the widespread availability of high-quality imaging systems and a better understanding of its pathology, the number of cases reported in the literature has been steadily increasing in recent years (5). Here, we reported the first case for Southeast Asia (Indonesia) of a Huge Frantz tumor in a young female patient who presented a rare indolent tumor.

CASE PRESENTATION

An 18 year old female patient presented with a dump upper abdominal mass in the outpatient clinic of our hospital, which was started 3 years ago. The patient noticed abdominal distension increasingly lasted one month before (along with weight loss of > 10 kg). At that moment, she denied any history of vomiting, change in bowel habits, weight loss, night sweats, scleral or urine discoloration, and menstrual irregularity. She was afebrile with a normal pulse and blood pressure. There was a palpable abdominal tenderness occupying most of her abdomen. No history of hepatic-biliary or pancreatic disease or other malignancy in her family was noted. Laboratory data were within normal limits. Due to prolong epigastric pain, an upper gastrointestinal study was performed in January 2022 with the conclusion of narrowing duodenal bulbus due to extralumen mass (Figure 1). Contrast-enhanced Computed Tomography (CT) abdomen (Figure 2) reported a large solid mass with enhancing cystic component and necrotic area, measuring 13,9 cm x 13,2 cm x 8,9 cm from corpus pancreas. A large mass was compressing the stomach superiorly, also compressing the portal vein, inferior cava vein, and abdominal aorta without any evidence of thrombosis. There was no evidence of distant metastases. Fine-needle aspiration biopsy yielded a cellular sample comprising pseudopapillary neoplasm of the pancreas. Intraoperatively was noted a rounded tumor, which revealed a huge mass occupying most of the abdominal cavity and displacing the stomach. A radical resection of the distal portion of the pancreas was performed. The patient was discharged in satisfactory condition. Post-1-year follow-up patient is doing well, with no signs of tumor recurrence locally and distance.

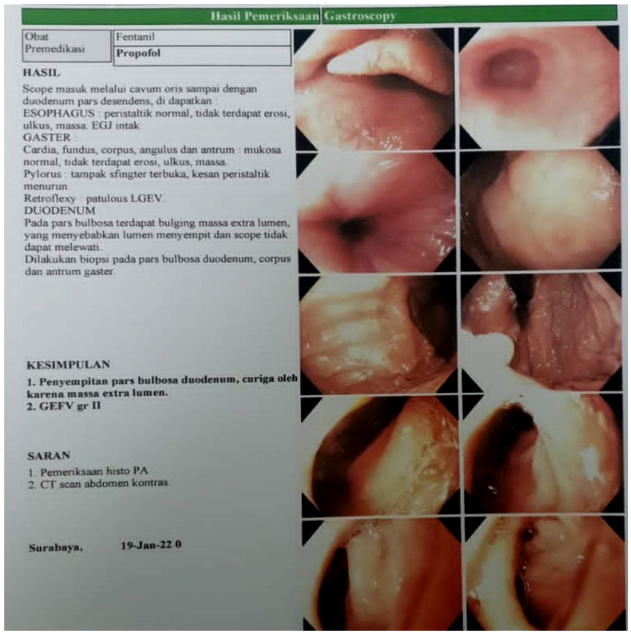

Figure 1. Upper gastrointestinal finding in endoscopy found narrowing duodenal bulbus due to extra luminal mass.

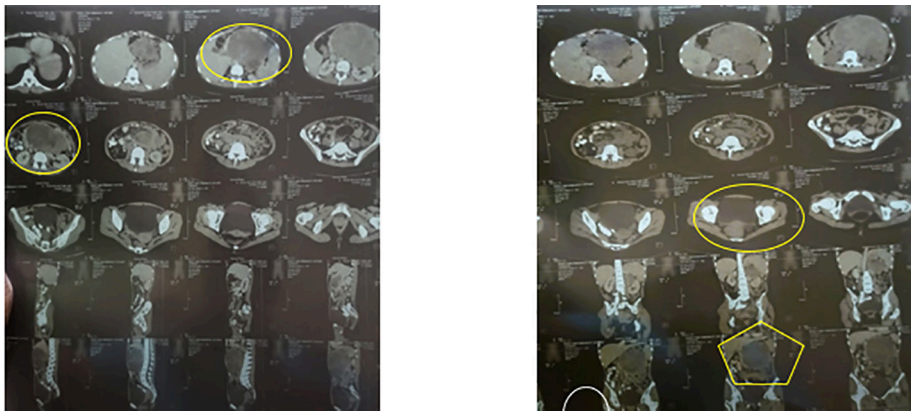

Figure 2. CT scan abdomen shows an enlargement mass from the pancreas compressed superiorly to gaster, well-marginated, encapsulated, solid and cystic mass.

DISCUSSION

SPPT can be found asymptomatic, and slow growing with an indolent course. Characterized by a long asymptomatic course and non-specific symptoms. The clinical presentation of the tumor is a palpable abdominal mass with uncharacteristic abdominal pain, and epigastric pain. Most commonly located in the body and tail (7). This case describes an indolent course from three years before admission because of non-specific symptoms and came to the hospital because palpable huge abdominal mass. SPPT is usually a well-demarcated tumor with a diameter ranging from 1,5 from 30 centimeters (cm) (average 10 cm). In our cases were 13.9 cm x 13.2 cm x 8.9 cm. Contrast Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) is superior in identifying capsule (8,9). SPPT is well-encapsulated from the pancreas, with large spongy areas of the hemorrhage on its cut surface alternating with both solid and cystic degeneration. The tumor contains a mixture of solid, cystic, and pseudopapillary patterns in various proportions. Both capsule and intratumoral hemorrhage are important clues to the diagnosis because these features are rarely found in another pancreatic tumor (10). Park et al. studied CT imaging features of SPPT in males and females which resulted in a lobulated shape that is more common among males and an oval shape in females (12,13)35.0 years.

Immunohistochemically, SPPT is typically positive for vimentin (Vim), a-1-antitrypsin (AAT), a-1-antichymotrypsin (AACT), and neuron-specific enolase (NSE) (14,15) which is consistent with the finding. However, the unique immunohistochemical features with an expression of CD56 and CD10 have not reached an agreement in a recent study (15). SPPT cells may also reveal focal immunoreactivity for cytokeratin (CK) and synaptophysin (Syn), abnormal nuclear location of β-catenin, and presence of progesterone receptors (PR), and may express galectin-3, all of which are useful in differentiating SPT from the endocrine pancreatic tumor (16,17).

The pathophysiology behind the development of SPN and its cellular origin is still a matter of debate with multiple purposed and hypotheses (7,18). One of the theories that have been suggested is the role of sex hormones in the pathogenesis of this tumor. It was proposed based on the higher female predominance rate that has been reported, particularly during the reproductive age, along with the fact that SPPT was usually positive for these receptors (19). During normal pancreas development, beta-catenin signaling within the beta-catenin/Wnt pathway is necessary and in the adult organ, this pathway is usually downregulated (20). The majority (85 %-90 %) of SPPT have exon-3 mutations and 10-15 % of mutations are present in other exons. The aberrant protein expression in SPPT is strongly

correlated with mutations in the beta-catenin gene (19,21). Mutations in beta-catenin gene exon-3 lead to Wnt signaling activation which plays an important role in the development of SPPT. Cell cycle-associated proteins like cyclin D1 and cyclin D3 are overexpressed in SPPT because of the deregulation of the cell cycle (22). The low tumor growth rate in SPPT is explained by the role of cyclin-dependent kinase inhibitors P^{21} and P^{27} in controlling the activated Wnt/beta-catenin signaling pathway (19,21,22). SPPTs are considered hormone sensitive because they express progesterone receptors (23). P^{21}, P^{27}, and cyclin D1 expression are influenced by estradiol and progesterone (24,25). At pathology, a solid pseudopapillary tumor is usually large and encapsulated and is composed of a mixture of cystic, solid, and hemorrhagic components. Both capsule and intratumoral hemorrhage are important clues to its diagnosis because they are rarely found in other pancreatic neoplasms.

Surgical management with free surgical resection margins is the mainstay of treatment even with metastasis and vascular invasion, surgical excision should be performed whenever feasible (26). In this case, attention to complications due to total resection probably wound infection; consider the patient at a young age. The recurrence rate after surgical resection has been reported to be 3 %-9 % (27). Regardless, the patient should be promptly followed up due to the risk of potential recurrence or emergence of metastatic lesions with serial imaging examination as the best recommendation. The prognosis of SPPT is generally excellent with a 95 % cure rate following complete surgical resection. In this case, abdominal discomfort with a palpable mass starts at an early puberty age (16 years old) when progesterone hormone is still low. From the literature 700 reported cases show a range of ages starting from 20 years old. All data show resection (even in the metastatic stage) preserves a good prognosis until 15 years old.

CONCLUSION

Solid pseudopapillary tumor of the pancreas is a rare primary neoplasm with unknown etiology, characterized by a paucity of clinical symptomatology, and can reach a large size. Radically curable with complete resection, distinguished from other tumors with a similar location because of its characteristic clinical and histopathologic features. The literature review supports the concept surgical resection offers an excellent prognosis.

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None.

Conflict of Interest

The authors declare no conflict of interest.

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Patient Consent

Obtained.

REFERENCES

1. Frantz VK. Tumors of the pancreas. In: Atlas of tumor pathology. Washington, DC: Armed forces Institute of Pathology. 1959:32-33.
2. Kloppel G, Solcia E, Longnecker DS, Capella C, Sobin LH. Histological typing of tumors of the exocrine pancreas. In: World Health Organisation. International histological classification of tumors. 2nd edition. Berlin Heidelberg New York: Springer; 1996.p.120-128.
3. Palnaes Hansen CP, Skaarup Kristensen TS, Stockholm JH, Hartnack Federspiel B. Solid pseudopapillary neoplasm of the pancreas: Clinical-pathological features and management, a single center experience. Rare Tumors. 2019;11:2036361319878513.
4. Sandlas G, Charu Tiwari C. Solid pseudopapillary tumor of pancreas: A case report and review of literature. Indian J Med Paediatr Oncol. 2017;38(2):207-209.
5. Yu PF, Hu ZH, Wang XB, Guo JM, Cheng XD, Zhang YL, et al. Solid pseudopapillary tumor of the pancreas: A review of 553 cases in Chinese literature. World J Gastroenterol. 2010;16(10):1209-1214.

6. Torres OJM, Rezende MB De, Waechter FL, Neiva RF, Moraes-Junior JMA, Torres C, et al. Pancreatoduodenectomy for solid pseudopapillary tumour of the pancreas: A multi-institution study. Arq Bras Cir Dig. 2019;32:e1442.
7. Lanke G, Ali FS, Lee JH. Clinical update on the management of pseudopapillary tumor of pancreas. World J Gastrointest Endosc. 2018;10(9):145-155.
8. Madjov R, Chernopolsky P, Bozhkov V, Madjova V. Solid pseudopapillary pancreatic tumor – Tumor of Frantz. Surg Gastroenterol Oncol. 2019;24(5):259-264.
9. Tang LH, Aydin H, Brennan MF, Klimstra DS. Clinically aggressive solid pseudopapillary tumors of the pancreas: A case report of two cases with component of undifferentiated carcinoma and a comparative clinicopathologic analysis of 34 conventional cases. Am J Surg Pathol. 2005;29:512-519.
10. Geers C, Moulin P, Gigot JF, Weynand B, Deprez P, Rahier J, et al. Solid pseudopapillary tumor of the pancreas-review and new insights into pathogenesis. Am J Surg Pathol. 2006;30:1243-1249.
11. Lam KY, Lo CY, Fan, ST. Pancreatic solid-cystic-papillary tumor: Clinicopathologic features in eight patients from Hongkong and review of the literature. World J Surg. 1999;23:1045-1050.
12. Park MJ, Lee JH, Kim JK, Kim YC, Park MS, Yu JS, et al. Multidetector CT imaging features of solid pseudopapillary tumours of the pancreas in male patients: distinctive imaging features with female patients. Br J Radiol. 2014;87:20130513.
13. Cai H, Zhou M, Hu Y, He H, Chen J, Tian W, et al. Solid pseudopapillary neoplasms of the pancreas: Clinical and pathological features of 33 cases. Surg Today. 2013;43:148-154.
14. Lee WY, Tzeng CC, Chen RM, Tsao CJ, Tseng JY, Jin YT, et al. Papillary cystic tumours of the pancreas: Assessment of malignant potential by analysis of progesterone receptor, flow cytometry, and ras oncogene mutation. Anticancer Res. 1997;17(4A):2587-2591.
15. Tieman K, Heitling U, Koshmal M, Kloppel G. Solid pseudopapillary neoplasm of the pancreas show an interruption of the Wnt-signalling pathway and express gene product of 11q. Mod Pathol. 2007;20:955-960.
16. Marikawa T, Onogawa T, Maeda S, Takadase T, Shirasaki K, Yoshida H, et al. Solid pseudopapillary neoplasms of the pancreas: an 18-year experience at a single Japanese Institution. Surg Today. 2013;43:26-32.
17. Machado MC, Machado MA, Bacchella T, Jukemura J, Almeida JL, Cunha JE. Solid pseudopapillary neoplasm of the pancreas distinct pattern of onset, diagnosis, and prognosis for male versus female patients. Surgery. 2008;143:29-34.
18. Heiser PW, Cano DA, Landsman I, Kim GE, Kench JG, Klimstra DS, et al. Stabilization of beta-catenin induces pancreas tumor formation. Gastroenterol. 2008;135:1288-1300.
19. Papavradimis T, Papvradimis S. Solid pseudopapillary tumors of the pancreas. Review of 718 patients reported in English literature. J Am Coll Surg. 2005;200:965-972.
20. Serra S, Chetty R. Revision 2: An immunohistochemical approach and evaluation of solid pseudopapillary tumour of the pancreas. J Clin Pathol. 2008;61:1153-1159.
21. Wrba F, Chott A, Ludvik B, Schratter M, Spona J, Reiner A, et al. Solid and cystic tumor of the pancreas: A hormonal-dependent neoplasm? Histopathology. 1988;12(3):338-340.
22. Tanaka Y, Kato K, Natohara K, Hojo H, Ijri R, Miyake T, et al. Frequent beta-catenin mutation and cytoplasmic/nuclear accumulation in pancreatic solid-pseudopapillary neoplasm. Cancer Res. 2001;61:8401-8404.
23. Maimune A, Luigiano C, Baccarini P, Fornelli A, Cennamo V, Polifemo A, et al. Preoperative diagnosis of a solid pseudopapillary tumor of the pancreas by Endoscopic Ultrasound Fine Needle Biopsy: A retrospective case series. Dig Liver Dis. 2013;45:957-960.
24. Nishihara K, Tsuneyoshi M, Ohsima A, Yamaguchi K. Papillary Cystic Tumor of the pancreas: Is it a hormone-dependent neoplasm? Pathol Res Pract. 1993;189:521-526.
25. Zamboni G, Bonetti F, Scarpa A, Pelosi G, Doglioni C, Iannucci A, et al. Expression of progesterone receptors in solid-cyst tumour of the pancreas- A clinicopathological and immunohistochemical study of ten cases. Virchow Arch A Pathol Anat Histopathol. 1993;423:425-431.
26. Mirminachi B, Farrokhzad S, Sharifi AH, Nikfam S, Nikmanesh A, Malekzahed R, et al. Solid pseudopapillary neoplasm of Pancreas; A case series and review literature. Middle East J Dig Dis. 2016;8:102-108.
27. Sperti C, Berselli M, Pasquali C, Pastorelli D, Pedrazzoli S. Aggressive behaviour of solid pseudopapillary tumor of the pancreas in adults: A case report and review of literature. World J Gastroenetrol. 2008;14:960-965.
28. Wang YM, Zhang SH, Sun CD, Hu WY. Diagnosis of solid pseudopapillary tumor of the pancreas: A case report of 16 cases. Qilu Yixue Zashi. 2008;23:112-114.
29. Abdullah SA, Hassan MA, Alaa AA, Muna A, Ahmed AA, Mohammed SA. Huge solid pseudopapillary tumor of the pancreas 'Frantz tumor': A case report. J Gastrointest Oncol. 2020;11(5):1098-1104.