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CASE REPORT

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Atypical solid pseudopapillary tumor of the pancreas in a 14-year-old

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Abstract

Diagnostic approach can be difficult in pediatric pancreatic masses. Our case shows that even though radiologic appearance was not conclusive, surgery remains the main treatment in resectable masses especially in children.

KEYWORDS

Frant'z tumor, pancreatic tumor, pancreatoduodenectomy, solid pseudopapillary tumor pediatric, surgery

1 | INTRODUCTION

Solid pseudo papillary tumor (SPT) of the pancreas is an uncommon malignancy in pediatric patients. A fourteen-years-old girl presented with obstructive cholestasis. Magnetic resonance imaging showed a pancreatic head mass. Whipple's procedure was performed. Histopathology confirmed SPT. The diagnostic approach can be difficult in pediatric pancreatic masses.

Pancreatic tumors are rare in children and only count for 0.2% of childhood malignancies.[1] Solid Pseudopapillary Tumor (SPT) of the pancreas or Frant'z tumor is a low-grade malignancy and the most common histologic subtype.[2] It is commonly an adult tumor but rarely occurs in the pediatric population.[3] We report an interesting case of SPT occurring in a child.

2 | CASE PRESENTATION

A 14-year-old girl presented to the out-patient clinics with a three-month history of diarrhea, intense pruritus, and weight loss. Upon hospital admission, her vital signs were blood pressure 110/60 mm Hg, pulse 82 beats/min, and temperature 37°C. Her body mass index was calculated at 41 kg/m^2 (>95th percentile). Physical examination of the abdomen was normal. Laboratory assessment showed an anicteric cholestasis: alkaline phosphatase of 480 U/L (N = 135 U/L), a total bilirubin of 9 mg/L (normal value 3-10 mg/L), and elevated liver enzymes: alanine aminotransferase of 180 U/L (N = 45 U/L), aspartate aminotransferase of 365 U/L (N = 45 U/L). An abdominal ultrasound showed intrahepatic and extrahepatic ductal dilation and a hypoechoic mass in the head of the pancreas measuring 3 cm. Upper endoscopy

and colonoscopy with duodenal, ileal, and colonic biopsies were normal. A computed tomography (CT) scan with contrast (Figure 1) showed a weakly enhancing lesion in the head of the pancreas with a central calcification measuring 36 × 30 mm, dilation of the biliary tree, a normal pancreatic duct, and three intrahepatic lesions recalling focal nodular hyperplasia (FNH). Magnetic resonance imaging (MRI) of the abdomen was ordered to further examine the pancreatic mass. It demonstrated the same biliary dilation upstream a nonencapsulated, well-defined 35 × 30mm lesion with solid and cystic components in the head of the pancreas. The tumor was hypointense on T1 compared to the rest of the pancreas and heterogeneously isointense on T2 sequence with mild peripheral contrast enhancement adjacent to second and third duodenum (Figure 2). The common bile duct measured 1.6 cm in diameter with an abrupt transition at the head of the pancreas. The MRI also confirmed FNH with central scarring and showed no invasion of the blood vessels or adjacent organs. The radiologist concluded that the mass features were suggestive of a pancreatoblastoma (PB). The tumor markers carcinoembryonic antigen, carbohydrate antigen, and alpha-fetoprotein were all within the normal range. Immunoglobulin G 4 levels were also within normal limits, and the hepatitis immune panel was negative. There was no evidence of distant metastatic spread.

This patient's case was then reviewed in a multidisciplinary board meeting. Initially, an endoscopic ultrasound was scheduled but later canceled because fine needle biopsie (FNB) was not available in our center. Laparoscopic biopsy was not feasible because of the interposition of digestive structures. One day, the patient reported intense epigastric pain as well as jaundice. A decision was promptly made to take her to the operating room for a Whipple's procedure. She underwent surgery 3 months after her initial presentation. At intraoperative exploration, surgeons discovered a white peritoneal effusion surrounding the pancreas suggesting lymphorrea which was confirmed by intraoperative cytology. A cholecystectomy and a cephalic duodenopancreatectomy were performed.

The macroscopic examination of the surgical specimen showed a tumor measuring 45 mm (Figure 3). The solid component was formed by uniform cells with round nuclei. These tumor cells were not cohesive in arrangement, but they were organized around a fibrous axis forming pseudopapillary structures. Perineural invasion and deep invasion of adjacent surrounding pancreatic tissue were also noted (Figure 4). The immunostaining of the tumor was positive for beta-catenin (nuclear and cytoplasmic), synaptophysin, CD56, and CD10 (Figure 5) but negative for chromogranin and CD99. Examination of eight lymph nodes was negative for tumor invasion.

The final pathology report concluded to a T3N0M0 with no lymph node invasion and no distant metastases pancreatic SPT with negative resection margins (R0) (according to exocrine pancreatic cancer TMN staging AJCC UICC 8th edition). Her postoperative course was uneventful. No adjuvant chemotherapy was indicated. No recurrence was observed on follow-up 6 months later.

3 | DISCUSSION

We report a case of SPT occurring in a pediatric patient. SPT is most common in adult women.[2–4] It does not have a typical clinical presentation.[5] The symptoms are nonspecific ranging from pruritus to acute or chronic abdominal pain; it can also be an incidental finding on radiological examination.[6] In our case, it was discovered through exploration of a chronic diarrhea and obstructive pancreatic head mass. Although an exocrine insufficiency was not ruled out, the diarrhea was a misleading symptom, and no other explanation was found.

Usually, SPTs are located in the head of the pancreas but can also develop in the tail.[7]

There are no pathognomonic anomalies on laboratory assessment and no specific tumor markers. The common methods used to diagnose SPTs are an abdominal ultrasound and CT scan, but MRI is the most sensitive method in detecting pancreatic lesions.[8,9] The typical radiological features of SPT are an encapsulated heterogeneous mass with well-defined

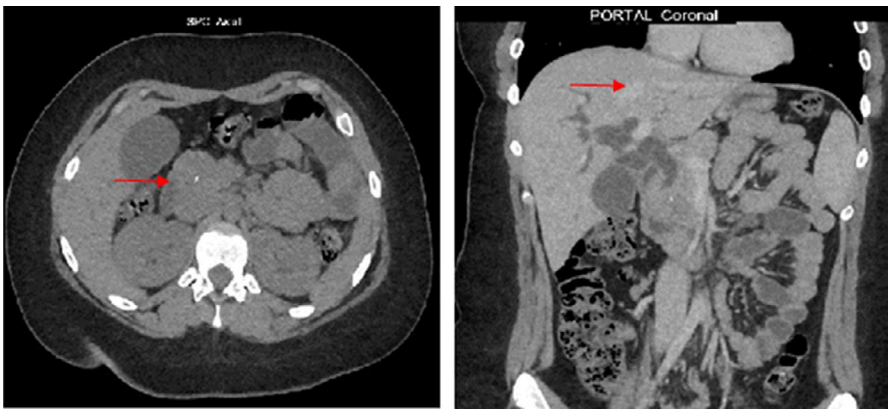

FIGURE 1 Computed tomography scan showing a cystic and solid mass 36 × 30 mass within the pancreatic head and hepatic lesions suggestive for nodular regeneration hyperplasia (arrows)

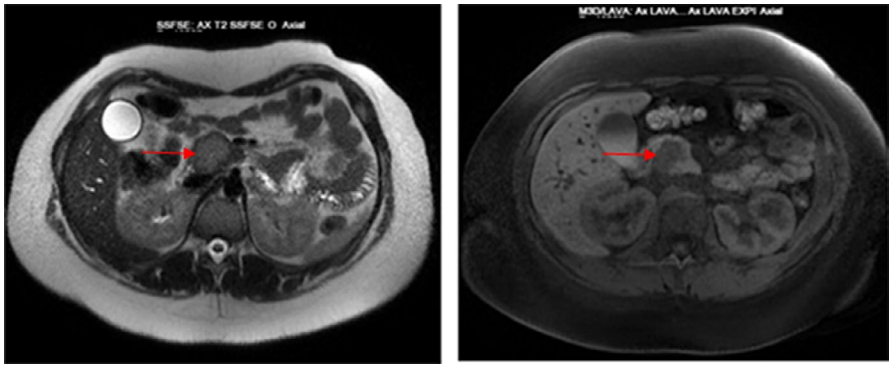

FIGURE 2 Magnetic resonance imaging shows that the tumor is a well-marginated solid and cystic mass (arrow)

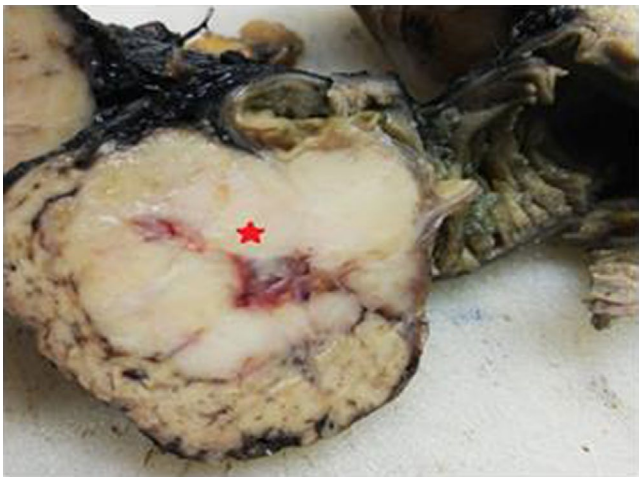

FIGURE 3 Macroscopic appearance of the resected specimen showing the solid and cystic component with hemorrhagic areas of the pancreatic head mass (star)

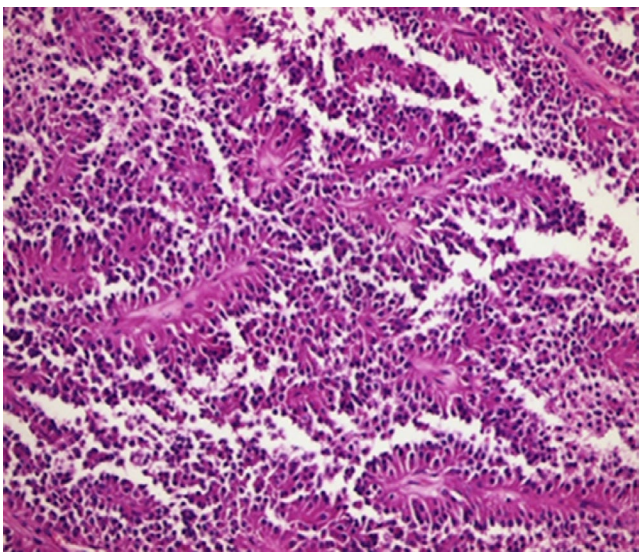

FIGURE 4 Histological appearance of solid pseudopapillar tumors. Solid pseudopapillar tumors exhibit a pseudopapillary pattern

borders, a prominent capsule, solid and cystic components, as well as hemorrhage.[2] Abdominal MRI or CT shows these features in 60%.[6]

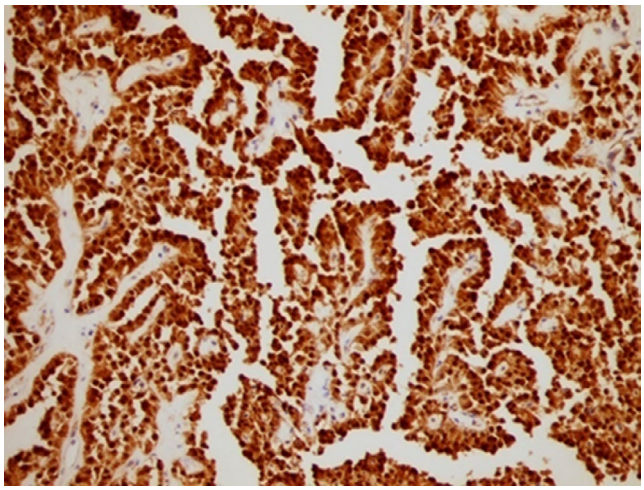

FIGURE 5 Solid pseudopapillary neoplasm: nuclear and cytoplasmic β-catenin labelling (Immunohistochimistry ×200)

In our case, we came across an obstructive mass of the head, but diagnostic strategy of pancreatic head masses was not standardized as it was in adults.[1] The resection without previous histological diagnosis is very common in adults because of the high probability of malignancies. The differential diagnosis is different in pediatric patients as adenocarcinoma is very rare. With the exception of pancreatic tumors such as PB which are the most common type in children, benign lesions of the head of the pancreas such as autoimmune chronic pancreatitis with mass effect are more likely to cause biliary obstruction than neoplasms in children.[10] Old reports suggest that children should undergo biopsy and biliary diversion before aggressive surgical treatment.[1] However, the benefits of preoperative histological proof of malignancy is still controversial. Some reports that preoperative FNB, for example, can guide the surgical management of low malignant tumors such as PB or SPT as opposed to adenocarcinoma.[11] Other studies report that even a benign histology reported on an intraoperative biopsy does not exclude the presence of malignancies in 35% of the cases.[1] Recently, there are reports supporting that the diagnostic approach of SPTs should rely more on radiologic findings because MRI or CT can be conclusive. Hence, biopsy is not required for diagnosis before

surgery especially for large masses.[12] Moreover, FNB can be dangerous, and there was a reported case of ruptured SPT through FNB.[12] Some studies suggest guidelines supporting this approach.[13] In our case, AIP with a mass effect was suspected, but knowing that AIP can coexist with a pancreatic tumor,[1] the absence of the irregular pattern of biliary duct on MRI and the normal levels of IgG4 levels helped with the differential diagnosis.

Surgical resection of pancreatic tumors is the standard treatment for children as for adults.[14] Even though SPTs are of low malignant potential and generally have a good prognosis, a complete surgical resection should be performed in patients presenting with locally invasive SPTs.[15] The exact extent of resection is less defined, and organ preservation is preferred.[16] Over time, more conservative procedures and less invasive approaches have been used. Generally, distal pancreatectomy with spleen preservation is recommended for SPT located in corpus and tail of the pancreas, and Whipple's procedure is recommended for SPT located in the head of pancreas.[16,17]

The final pathological examination confirms the tumor and usually requires immunohistochemistry that shows beta-catenin staining in 100% cases as it is considered the most sensitive and specific marker for SPT.[18] The malignant potential of the tumor is defined by histopathological criteria such as an elevated mitotic rate, high-cellular pleomorphism, a large tumor size (diameter >5 cm), lymphovascular invasion, perineural infiltration, lymph node metastasis, and positive surgical margins, as well as clinical criteria such as metastatic spread and recurrence.[15,19]

Although SPT is usually not a high-grade tumor, it can be locally aggressive invading the spleen and the duodenum. The common metastatic sites include the liver, lymph nodes, and peritoneum.[7] One study suggests that there is no evidence of ascites even in SPT with peritoneal metastasis.[20] In fact, tumor rupture is the main cause of peritoneal metastasis in low-grade SPTs.[20] Hence, the peritoneal effusion found around the pancreas in this case was not carcinomatosis and was confirmed on cytology as lymphorrea. As for lymph node invasion, it is also rare, and therefore, no lymphadenectomy is needed.[20]

Finally, SPT has an overall favorable outcome. Therefore, curative surgery is performed when possible.[21] There should be a minimum of a 5-year follow-up to detect recurrence.[22] As for tumors with aggressive features, a prolonged follow-up is essential.[19]

4 | CONCLUSION

SPT is rare in the pediatric population. Clinical, laboratory, imaging findings are not specific. Surgery remains the essential treatment. Prognosis is good if surgery is performed.

ETHICS STATEMENT

Personal data of the patient were respected during the study.

CONFLICT OF INTEREST

Authors report no conflicts of interest for this study.

AUTHOR CONTRIBUTIONS

Sabbah Meriam and Briki Ines: wrote the paper. Ben Farhat Fatma: changed the required modifications by the reviewers. Khanchel Fatma and Helal Imen: read the histology and give pictures. Haddad Dhafer: performed the surgery. Chammakhi Chiraz: performed the CT scan and MRI. Bellil Nawel: managed the case in the gastroenterology department Dalila. Gargouri: reviewed the paper.

DATA AVAILABILITY STATEMENT

No data are available.

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REFERENCES

1. Park J, Dunn JCY, Atkinson JB. Management of children with pancreatic head mass. *J Pediatr Surg*. 2006;41:e1-e4.
2. Mylonas KS, Doulamis IP, Tsilimigras DI, et al. Solid pseudopapillary and malignant pancreatic tumors in childhood: A systematic review and evidence quality assessment. *Pediatr Blood Cancer*. 2018;65:e27114. https://doi.org/10.1002/pbc.27114
3. Tan HL, Syn N, Goh BKP. Systematic review and meta-analysis of minimally invasive pancreatectomies for solid pseudopapillary neoplasms of the pancreas. *Pancreas*. 2019;48:1334-1342.
4. Martin RCG, Klimstra DS, Brennan MF, Conlon KC. Solid-pseudopapillary tumor of the pancreas: a surgical enigma? *Ann Surg Oncol*. 2002;9:35-40.
5. Bochis OV, Bota M, Mihut E, Buiga R, Hazbei DS, Irimie A. Solid pseudopapillary tumor of the pancreas: clinical-pathological features and management of 13 cases. *Clujul Medical*. 2017;90:171.
6. Yagcı A, Yakan S, Coskun A, et al. Diagnosis and treatment of solid pseudopapillary tumor of the pancreas: experience of one single institution from Turkey. *World J Surg Oncol*. 2013;11:308.
7. Kang CM, Kim KS, Choi JS, Kim H, Lee WJ, Kim BR. Solid pseudopapillary tumor of the pancreas suggesting malignant potential. *Pancreas*. 2006;32:276-280.
8. Jiang L, Cui L, Wang J, Chen W, Miao L, Jia J. Solid pseudopapillary tumors of the pancreas: Findings from routine screening sonographic examination and the value of contrast-enhanced ultrasound. *J Clin Ultrasound*. 2015;43:277-282.
9. Yu CC, Tseng JH, Yeh CN, Hwang TL, Jan YY. Clinicopathological study of solid and pseudopapillary tumor of pancreas: Emphasis on magnetic resonance imaging findings. *World J Gastroenterol*. 2007;13:1811-1815.
10. Friedlander J, Quiros JA, Morgan T, et al. Diagnosis of autoimmune pancreatitis vs neoplasms in children with pancreatic mass and biliary obstruction. *Clin Gastroenterol Hepatol*. 2012;10(1051–1055):e1.

11. Master SS, Savides TJ. Diagnosis of solid-pseudopapillary neoplasm of the pancreas by EUS-guided FNA. *Gastrointest Endosc*. 2003;57:965-968.
12. Virgilio E, Mercantini P, Ferri M, et al. Is EUS-FNA of solid-pseudopapillary neoplasms of the pancreas as a preoperative procedure really necessary and free of acceptable risks? *Pancreatology*. 2014;14:536-538.
13. Lanke G, Ali FS, Lee JH. Clinical update on the management of pseudopapillary tumor of pancreas. *World J Gastrointest Endosc*. 2018;10:145-155.
14. Mylonas KS, Nasioudis D, Tsilimigras DI, Doulamis IP, Masiakos PT, Kelleher CM. A population-based analysis of a rare oncologic entity: malignant pancreatic tumors in children. *J Pediatr Surg*. 2018;53(4):647-652.
15. Campanile M, Nicolas A, LeBel S, Delarue A, Guys JM, de Lagausie P. Frantz's tumor: is mutilating surgery always justified in young patients? *Surg Oncol*. 2011;20:121-125.
16. de Castro SMM, Singhal D, Aronson DC, et al. Management of solid-pseudopapillary neoplasms of the pancreas: a comparison with standard pancreatic neoplasms. *World J Surg*. 2007;31:1130-1135.
17. van den Akker M, Angelini P, Taylor G, Chami R, Gerstle JT, Gupta A. Malignant pancreatic tumors in children: a single-institution series. *J Pediatr Surg*. 2012;47:681-687.
18. Nguyen NQ, Johns AL, Gill AJ, et al. Clinical and immunohistochemical features of 34 solid pseudopapillary tumors of the pancreas. *J Gastroenterol Hepatol*. 2011;26:267-274.
19. Gao H, Gao Y, Yin L, et al. Risk Factors of the recurrences of pancreatic solid pseudopapillary tumors: a systematic review and meta-analysis. *J Cancer*. 2018;9:1905-1914.
20. Guimarães LS, de Melo AM, Ruiz MR, Viana Jdos S, da Silva Junior RA. Solid pseudopapillary tumor of the pancreatic: evaluation of clinical, radiological and surgical profiles. *Rev Col Bras Cir*. 2013;40:398-403.
21. Marchegiani G, Crippa S, Malleo G, et al. Surgical treatment of pancreatic tumors in childhood and adolescence: uncommon neoplasms with favorable outcome. *Pancreatology*. 2011;11(4):383-389.
22. Law JK, Ahmed A, Singh VK, et al. A systematic review of solid-pseudopapillary neoplasms: are these rare lesions? *Pancreas*. 2014;43:331-337.

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