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# Solid pseudopapillary tumor of the pancreas mistaken for gastrointestinal stromal tumor: A case report

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**Abstract**

Solid pseudopapillary epithelial neoplasms of the pancreas are rare entities, first described in 1959 by Frantz. These tumors represent less than 2% of pancreatic cancers and mainly affect young women. They can reach a significant size and its radiological features can lead to diagnostic pitfalls, such as gastrointestinal stromal tumors, which are rare soft-tissue sarcomas that can appear anywhere along the gastrointestinal tract. Clinicians and radiologists need to be aware of the existing diagnostic pitfalls between these two entities, because of their possible similarities. We report here the case of a 33-year-old woman with a solid pseudopapillary epithelial neoplasms of the pancreas initially misdiagnosed as an exophytic gastric stromal tumor.

**Keywords**

Solid pseudopapillary tumor, gastrointestinal stromal tumors, imaging pitfalls

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## Introduction

Solid pseudopapillary epithelial neoplasms of the pancreas (SPEN) are rare entities, first described in 1959 by Frantz.[1] These tumors represent less than 2% of pancreatic cancers, mainly affect young women and thought to generally affect people of Asian or African-American origin.[2,3] They can reach a significant size, with an average of 10 cm at the time of diagnosis,[4] while remaining poorly symptomatic or asymptomatic. Its size and its radiological features can lead to diagnostic pitfalls, particularly with gastrointestinal stromal tumors (GIST), which are rare soft-tissue sarcomas that can appear along the gastrointestinal tract. The most common site is the stomach, followed by the small intestine, colon, and rectum.[5] We report here the case of a 33-year-old woman with a SPEN initially misdiagnosed as an exophytic gastric stromal tumor.

## Case report

A 33-year-old Guinean woman, with a past medical history of diabetes, presented with a chronic left upper quadrant pain, with an abdominal mass, progressively increasing in size. The patient underwent a median laparotomy in her country 2 years ago, which resulted in a splenectomy (surgical procedure unknown; documents were missing).

On admission, clinical examination revealed a fixed mass deep in the left upper quadrant, extending beyond the costal margin. Tumor markers Carcinoembryonic antigen (CEA) and CA19-9 were normal.

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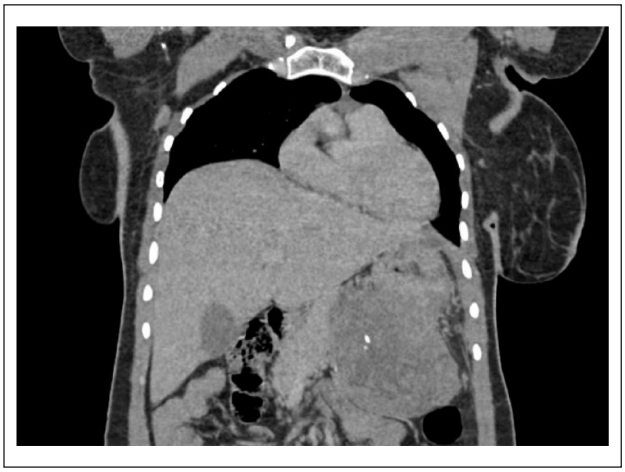

**Figure 1.** Coronal CT image showing rounded, well-limited lesion, appearing to be attached to the gastric wall which seems thickened. It has a double component cystic and tissular and it presents a few calcifications.

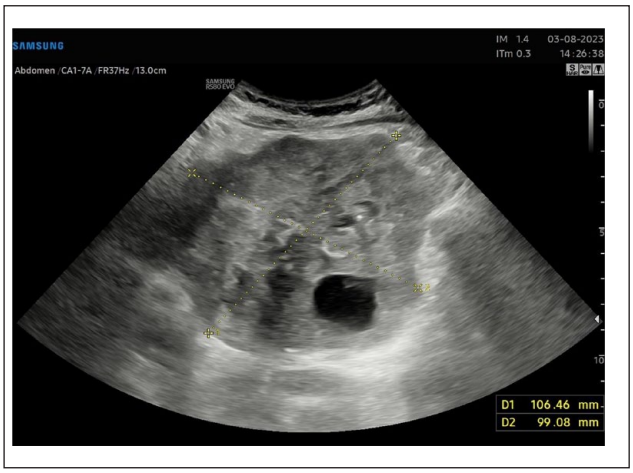


**Figure 2.** During biopsy, ultrasound showed a circumscribed, encapsulated mass spreading to the gastric region with a mixed, heterogeneous echogenicity and calcifications.

Abdominal computed tomography (CT) scan (Figure 1) showed a $104 \times 77 \times 44$ mm rounded, well-limited lesion, appearing to be attached to the gastric wall which seems thickened. It has a double component cystic and tissular, with calcifications. There were no signs of locoregional or systemic extent. These findings were suggestive of exophytic gastric stromal tumor. Ultrasound-guided biopsy was performed (Figure 2), and pathological findings (Figure 3) revealed the existence of tumor cells organized around a vascular axis and presenting cytoplasm that was clear and also eosinophilic. These findings were suggestive of a SPEN.

Immunohistochemistry showed that tumor cells were positive for CD 10, synaptophysin, beta-catenin, and progesterone receptor, in favor of a SPEN or Frantz tumor (WHO 5th edition). It showed also that CD 117 and DOG 1 were negative, excluding a GIST.

Further investigation with pancreatic magnetic resonance imaging (MRI) (Figure 4) showed a well-limited, encapsulated lesion that originates from the tail of pancreas, with a dual component: a cystic portion with hyposignal T1 and a distinct T2 hypersignal, and a solid portion with isosignal T1, intermediate T2 signal, restricting diffusion and heterogeneously enhanced after gadolinium injection. This examination also enabled us to exclude the presence of a ductal communication, locoregional invasion, or hepatic localization.

A caudal pancreatectomy was performed removing the splenic artery and vein, with uneventful post operative course (Figures 5 and 6). Diagnosis of pancreatic SPEN was subsequently confirmed on histopathology examination of surgical specimen.

## Discussion

SPEN is a very rare entity, representing less than 2% of exocrine pancreatic tumors and less than 5% of cystic pancreatic tumors,[6] with a reported incidence of 0.13%–2.7% of all pancreatic tumors.[7] It was first described by Frantz in 1959, who called it a "benign or malignant papillary tumor of the pancreas." The term pseudo-papillary solid tumor was finally adopted by the WHO in 1996. It generally affects women, with an average age of 28 and a sex ratio F/M of 10:1.[8] Some studies point to a possible predominance in Asian and African-American women.[9–11] In our case, the patient was a 33-year-old woman of African origin.

SPEN can involve the head, body, or tail of the pancreas, with a predilection for the corporeal caudal region (64% of cases).[11] This case involved the tail of the pancreas. Rare cases of extra-pancreatic localization have also been described (1%), namely retroperitoneal, duodenal, mesocolic, and hepatic.[12,13]

SPEN leads to metastasis in 10%–15% of cases (mainly hepatic, peritoneal, and pulmonary), although metastatic and advanced forms have a favorable overall survival.[14,15]

The exact origin of SPEN remains unclear. Several authors have looked for a possible correlation between SPEN and sex hormone receptors[16,17] without any clear evidence of their involvement, particularly estrogen.[18–22] Other pathways have been investigated, such as the involvement of the p53 and K-RAS genes, chromosomal abnormalities, or protein mutations.[23]

Immunohistochemistry can be very useful in difficult cases, notably to differentiate SPEN from pancreatic endocrine tumors. SPEN generally express α1-antitrypsin, α1-antichymotrypsin, neuron-specific enolase, vimentin, and the progesterone receptor.[24]

There is a consensus to classify SPEN as lesions of uncertain or low malignant potential.[8,25,26]

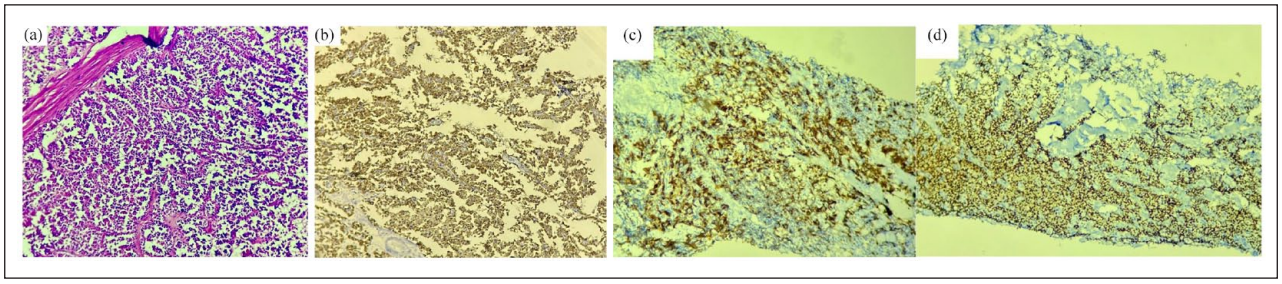


**Figure 3.** Microscopic images: (a) Section showing solid monomorphic polygonal cells with delicate vessels and a hyalinized or myxoid stroma and marked degenerative changes with pseudopapillae formation (×25) and (b) are showing nuclear expression of beta catenin (×25) (c) the tumor cells are positive for synaptophysin (×10) (d) and the progesterone receptor is strongly expressed (×10).

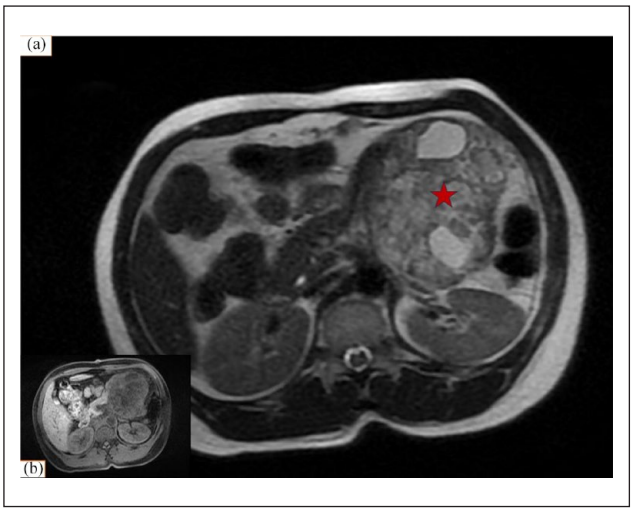


**Figure 4.** Pancreatic MRI: (a) Axial T2-weighted section showing a well-limited, encapsulated mass in the tail of the pancreas, with a dual component: cystic in frank hypersignal and tissular in intermediate signal. (b) T1FS axial sections without and with gadolinium injection, showing enhancement of the fleshy portion of the mass.

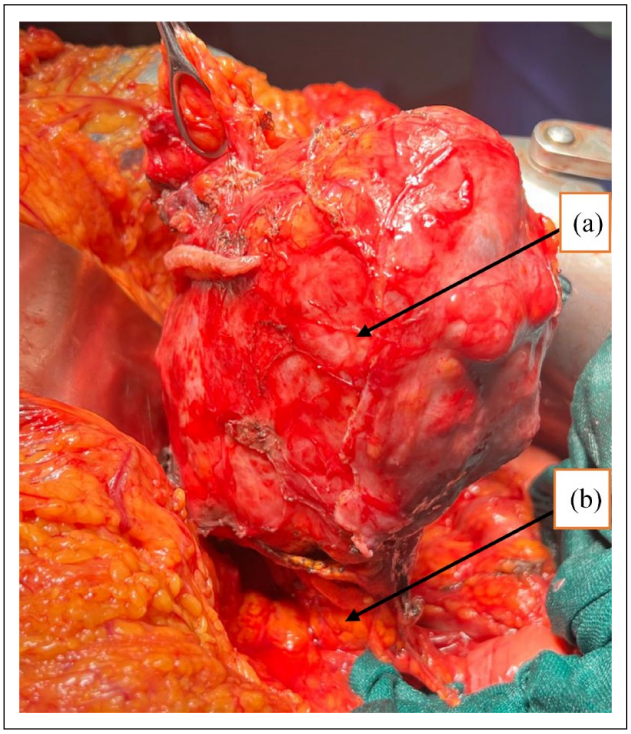


**Figure 5.** Operative view of the mass of the tail of the pancreas protruding into the transverse mesocolon. (a) Tumor mass (b) Body of the pancreas.

Clinical features of SPEN vary widely, and it is very often an incidental finding. If the tumor is symptomatic, it may be revealed by abdominal pain, more precisely supraumbilical or epigastric as in our case, or by palpation of an abdominal mass. SPEN may also be revealed, uncommonly, by a complication such as rupture or intra-tumoral hemorrhage.[27]

Biological tests are often normal, with no abnormalities in pancreatic tests or tumor markers.

Differential diagnosis of SPEN includes all heterogeneous pancreatic or peripancreatic masses with both solid and cystic components. It comprises entities such as neuroendocrine tumor, leiomyoma, leiomyosarcoma, liposarcoma, some cystic neoplasms such as serous microcystic adenoma, and rarely gastric stromal tumor. Although some of these lesions have a typical appearance on CT and MRI, there are several similarities and pitfalls leading to misdiagnosis.[28]

GIST are rare soft-tissue sarcomas that can arise anywhere along the gastrointestinal tract. The most common site is the stomach, followed by the small intestine, colon, and rectum.[5] They may remain asymptomatic for a long time, until they reach a certain volume and cause unspecific abdominal pain, leading to the appearance of a palpable mass and complications such as perforation or bleeding.[29–31]

There is a similarity with SPEN in the circumstances of discovery, which can compromise their distinctions.

CT scan is the most commonly used test for the diagnosis of GIST in the case of large tumors.[5] It reveals a solid tumor, frequently voluminous, with a cystic part, as well as calcifications and necrotic areas in some patients.[32]

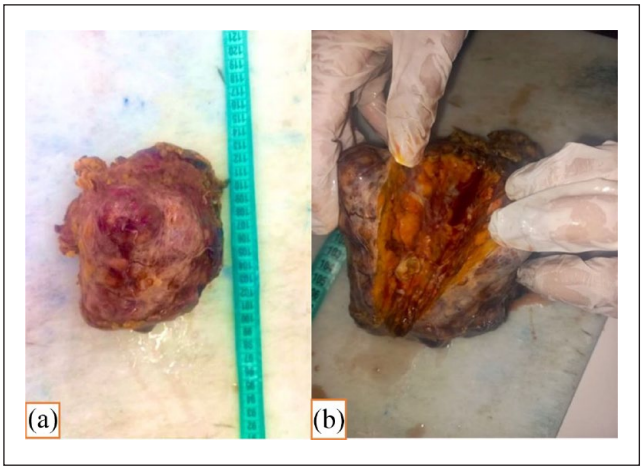


**Figure 6.** Macroscopic image of the surgical specimen. Macroscopic views showing: (a) A well-encapsulated mass (b) Variable amount of solid and cystic patterns with friable consistency in the middle.

In the case of SPEN, the tumor is a well-limited, heterogeneous mass of mixed density, with peripheral enhancement of solid contingents after injection of contrast, presenting a central necrotic-hemorrhagic component on CT scan. Peripheral calcifications can be found in one-third of cases.

The clinical similarities between these two entities as well as the similarities found on CT scan explain the possible confusion between the two diagnoses. There are several published cases of GIST mimicking SPEN[28,33] as well as SPEN mimicking GIST.[34,35]

In our patient's case, the diagnosis of GIST was initially made on the basis of clinical and CT data.

Ultrasound-guided percutaneous biopsy is an available and affordable tool that provides satisfactory results for sampling when the tumor is accessible and significant in size. However, the risk of tumor dissemination and bleeding is not negligible.[36]

EUS FNB is preferable to transparietal sampling. Its effectiveness is greater than 80%[29] in GISTs, and it has also been proven effective in the diagnosis of SPEN.[35] Despite these advantages, its availability is restricted and its cost may vary depending on the structure.

Histological results obtained by ultrasound-guided biopsies have sealed the diagnosis of SPEN in our case.

MRI is a useful complementary tool to CT, particularly for the evaluation of large tumors. In our case, it established the pancreatic origin of the tumor, adding further confirmation to the diagnosis of SPEN made by histology.

MRI is better than CT at distinguishing certain features,[37] such as hemorrhage and cystic degeneration.

In the case of SPEN, MRI reveals a well-delimited tumor with peripheral T1 and T2 hyposignal corresponding to the fibrous capsule. Hemorrhagic areas appear in T1 hypersignal and T2 hyposignal. Cystic areas present as a clear T2 hypersignal, and tissue areas as T1 hyposignal, and variable T2.

Surgery is the standard of SPEN treatment, and is generally curative in the case of localized disease, ranging from enucleation to partial or total pancreatectomy, depending on the topography and size of the tumor.[10,38,39]

For our patient, a caudal pancreatectomy was performed as she already had splenectomy. Spleen preservation should be attempted, if possible, as lymph node dissection is not required. Excision should be extended if there is invasion of surrounding organs, and any nodules of peritoneal carcinosis should be resected.[17,22] Resection of metastatic lesions should be attempted.

A study found that the prognosis for SPEN with resected liver metastases usually surpasses 5 years.[23] Tumor recurrence should be treated with surgical excision.[39] The use of radiotherapy[40] or chemotherapy[41] has rarely been reported, so it is difficult to judge the value of these treatments. Hormone therapy[42] is also reported to be not effective. The overall prognosis is good, with a 5 years survival rate of 95%.[43]

## Conclusion

In conclusion, we present here the case of a pseudo-papillary solid tumor of the pancreas initially mistaken as an exophytic gastric stromal tumor. Clinicians and radiologists need to be aware of the existing diagnostic pitfalls between these two entities because of their many similarities. It is essential that every large mass of heterogeneous morphology, with gastric or pancreatic relationships, be carefully assessed, in order to make the appropriate management decision.

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### Author contributions

A.A. contributed to Conceptualization, Methodology, Investigation, Writing—Original Draft; M.A.E. contributed to Validation, Investigation, Resources, Writing—Review & Editing, Supervision; M.J. contributed to Investigation, Resources, Writing—Original Draft; M.F.B. contributed to Investigation; M.N. contributed to Visualization, Investigation; S.M.B. contributed to Visualization, Investigation; S.B. contributed to Validation, Writing—Review & Editing, Supervision; F.N. contributed to Validation, Writing—Review & Editing, Supervision; T.A. contributed to Validation, Writing—Review & Editing, Supervision; M.T. contributed to Validation, Writing—Review & Editing, Supervision.

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## Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

## Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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## References


1. Frantz V. *Atlas of tumor pathology, 7th section, 27–28th fascicles*. Washington: US Armed Forces Institute of Pathology, 1959.
2. Oertel JE, Mendelsohn G and Compagno J. Solid and papillary epithelial neoplasms of the pancreas. In: Humphrey GB, Grindey GB, Dehner LP, et al. (eds) *Pancreatic tumors in children. Cancer treatment and research*. Vol 8. Dordrecht: Springer, 1982.
3. Kuo TT, Su IJ and Chien CH. Solid and papillary neoplasm of the pancreas. Report of three cases from Taiwan. *Cancer* 1984; 54(7): 1469–1474.
4. Lima SO, Santana VR, Leao SC, et al. Solid-pseudopapillary tumor of pancreas in a young woman: a case report and literature review. *Rev Med Chile* 2012; 140: 1179–1184.
5. Mantese G. Gastrointestinal stromal tumor: epidemiology, diagnosis, and treatment. *Curr Opin Gastroenterol* 2019; 35(6): 555–559, https://journals.lww.com/co-gastroenterology/fulltext/2019/11000/gastrointestinal_stromal_tumor__epidemiology,.13.aspx
6. Canzonieri V, Berretta M, Buonadonna A, et al. Solid pseudopapillary tumour of the pancreas. *Lancet Oncol* 2003; 4(4): 255–256.
7. Crawford 2nd B. Solid and papillary epithelial neoplasm of the pancreas, diagnosis by cytology. *Southern Med J* 1998; 91(10): 973–977.
8. Yu PF, Hu ZH, Wang XB, et al. Solid pseudopapillary tumor of the pancreas: a review of 553 cases in Chinese literature. *World J Gastroenterol* 2010; 16(10): 1209.
9. A-Cienfuegos J, Lozano M, Rotellar F, et al. Solid pseudopapillary tumor of the pancreas (SPPT). Still an unsolved enigma. *Rev Esp Enferm Dig* 2010; 102(12): 722–728.
10. Choi SH, Kim SM, Oh JT, et al. Solid pseudopapillary tumor of the pancreas: a multicenter study of 23 pediatric cases. *J Pediatr Surg* 2006; 41(12): 1992–1995.
11. Choi JY, Kim MJ, Kim JH, et al. Solid pseudopapillary tumor of the pancreas: typical and atypical manifestations. *Am J Roentgenol* 2006; 187(2): W178–W186.
12. Denis MA, Frère A, Brixko C, et al. Tumeur pseudo-papillaire pancréatique révélée par la rupture de varices oeso-gastriques secondaire à une cirrhose biliaire par une compression cholédocienne. *Gastroentérol Clin Biol* 2005; 29(3): 291–293.
13. Podevin J, Triau S, Mirallié E, et al. Tumeurs pseudopapillaires et solides du pancréas: à propos de cinq cas et revue de la littérature. *Annales de Chirurgie,* 2003, 128(8): 543–548.
14. Mao C, Guvendi M, Domenico DR, et al. Papillary cystic and solid tumors of the pancreas: a pancreatic embryonic tumor? Studies of three cases and cumulative review of the world's literature. *Surgery* 1995; 118(5): 821–828.
15. Gonzalez-Campora R, JJ RM, JL VR, et al. Papillary cystic neoplasm of the pancreas with liver metastasis coexisting with thyroid papillary carcinoma. *Arch Pathol Laboratory Med* 1995; 119(3): 268–273.
16. Maffuz A, Silva JA and Torres-Vargas S. Preoperative gemcitabine for unresectable, solid pseudopapillary tumour of the pancreas. *Lancet Oncol* 2005; 6(3): 185–186.
17. Hasbi S, Menfaa M, Sakit F, et al. Tumeur pseudopapillaire et solide du pancréas: une cause rare de masse abdominale. *Pan Afric Med J* 2015; 22(1): 361.
18. Lam KY, Lo CY and Fan ST. Pancreatic solid-cystic-papillary tumor: clinicopathologic features in eight patients from Hong Kong and review of the literature. *World J Surg* 1999; 23: 1045–1050.
19. Wrba F. Solid and cystic tumor of the pancreas; a hormonal-dependent neoplasm? *Histopathology* 1988; 2: 338–340.
20. Nishihara K, Tsuneyoshi M, Ohshima A, et al. Papillary cystic tumor of the pancreas: is it a hormone-dependent neoplasm? *Pathol-Res Pract* 1993; 189(5): 521–526.
21. Zamboni G, Bonetti F, Scarpa A, et al. Expression of progesterone receptors in solid-cystic tumour of the pancreas: a clinicopathological and immunohistochemical study of ten cases. *Virchows Archiv A* 1993; 423: 425–431.
22. Lee W, Tzeng C, Chen R, et al. Papillary cystic tumors 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.
23. Papavramidis T and Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. *J Am Coll Surg* 2005; 200(6): 965–972.
24. Kim MJ, Jang SJ and Yu E. Loss of E-cadherin and cytoplasmic-nuclear expression of β-catenin are the most useful immunoprofiles in the diagnosis of solid-pseudopapillary neoplasm of the pancreas. *Hum Pathol* 2008; 39(2): 251–258.
25. Meshikhes A and Atassi R. Pancreatic pseudopapillary tumor in a male child. *JOP* 2004; 5(6): 505–511.
26. Vargas-Serrano B, Domínguez-Ferreras E and Chinchón-Espino D. Four cases of solid pseudopapillary tumors of pancreas: imaging findings and pathological correlations. *Eur J Radiol* 2006; 58(1): 132–139.
27. Sugito K, Kusafuka T, Hoshino M, et al. Application of radiofrequency ablation for giant solid pseudopapillary tumor of the pancreas. *Pediatr Int* 2010; 52(1): e29–e31.
28. Müller C, Beer A, Wrba F, et al. A gastrointestinal stromal tumour mimicking solid pseudopapillary neoplasia of the pancreas—A case report. *Eur Surg* 2018; 50: 68–72.
29. Casali P, Abecassis N, Bauer S, et al. Soft tissue and visceral sarcomas: ESMO–EURACAN clinical practice guidelines for diagnosis, treatment and follow-up. *Ann Oncol* 2018; 29: iv51–iv67.
30. Stanek M, Pisarska M, Budzyńska D, et al. Gastric gastrointestinal stromal tumors: clinical features and short-and long-term outcomes of laparoscopic resection. *Videosurg Other Miniinvas Techn* 2019; 14(2): 176–181.
31. Aghdassi A, Christoph A, Dombrowski F, et al. Gastrointestinal stromal tumors: clinical symptoms, location, metastasis formation, and associated malignancies in a single center retrospective study. *Digest Dis* 2018; 36(5): 337–345.

32. Ginevra D, Gloria A, Diletta C, et al. Relationship between diagnostic imaging features and prognostic outcomes in gastrointestinal stromal tumors (GIST). *Acta Bio Medica: Atenei Parmensis* 2019; 90(Suppl 5): 9.
33. Kim HS, Sung JY, Park WS, et al. Gastrointestinal stromal tumors of the stomach with extensive calcification: report of two cases. *Intern Med* 2012; 51(18): 2555–2558.
34. Solanki P, Bose B, Mondal A, et al. Solid pseudopapillary neoplasm of the pancreas mimicking gastrointestinal stromal tumour. *BMJ Case Rep* 2023; 16(9): e254805.
35. Furuhashi S, Takamori H, Abe S, et al. Solid-pseudopapillary pancreatic tumor, mimicking submucosal tumor of the stomach: a case report. *World J Gastrointest Surg* 2011; 3(12): 201.
36. Pettinato G, Di Vizio D, Manivel JC, et al. Solid-pseudopapillary tumor of the pancreas: a neoplasm with distinct and highly characteristic cytological features. *Diagnos Cytopathol* 2002; 27(6): 325–334.
37. Cantisani V, Mortele KJ, Levy A, et al. MR imaging features of solid pseudopapillary tumor of the pancreas in adult and pediatric patients. *Am J Roentgenol* 2003; 181(2): 395–401.
38. Danon O, Mofredj A, Morsli F, et al. Tumeur papillaire solide du pancréas chez un enfant [Papillary solid tumor of the pancreas in a child: a case report], *Ann Med Interne* (Paris)2000, pp. 606–608.
39. Cheng-hong P, Dong-feng C, Guang-wen Z, et al. The solid-pseudopapillary tumor of pancreas: the clinical characteristics and surgical treatment. *J Surg Res* 2006; 131(2): 276–282.
40. Strauss JF, Hirsch VJ, Rubey CN, et al. Resection of a solid and papillary epithelial neoplasm of the pancreas following treatment with Cis-platinum and 5-fluorouracil: a case report. *Med Pediatr Oncol* 1993; 21(5): 365–367.
41. Fried P, Cooper J, Balthazar E, et al. A role for radiotherapy in the treatment of solid and papillary neoplasms of the pancreas. *Cancer* 1985; 56(12): 2783–2785.
42. Abid M, Salah KB, Guirat M, et al. Tumeurs pseudopapillaires et solides du pancréas: deux observations et revue de la littérature. *Rev Med Intern* 2009; 30(5): 440–442.
43. Moholkar S, Sebire NJ and Roebuck DJ. Solid-pseudopapillary neoplasm of the pancreas: radiological-pathological correlation. *Pediatr Radiol* 2005; 35: 819–822.