Society of Medical Education & Research

2022, Volume 2, Issue 2, Page No: 20-23

Archive of International Journal of Cancer and Allied Science

Frantz's Tumor: A Rare Pancreatic Neoplasm with Distinctive Features

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Abstract

The solid pseudopapillary tumor of the pancreas (SPT), also known as a Frantz tumor, was first identified in 1959 by its namesake. This uncommon pancreatic tumor, previously known as papillary epithelial neoplasm, low-grade papillary neoplasm, or solid and papillary neoplasm, predominantly affects younger individuals, with a notable female predominance. Its cellular origin remains uncertain, with potential links to acinar, endocrine, ductal, or progenitor cells. Characterized by a favorable prognosis and low malignant potential, it constitutes under 3% of exocrine pancreatic tumors. While most SPTs are benign, malignancy occurs in up to 15% of cases. This report describes an unusual case of a 19-year-old female who presented with epigastric pain, was treated with surgical resection, and was confirmed as a Frantz tumor via histopathology and immunohistochemistry.

Keywords: Frantz tumor, Pancreas, Papillary lesions, Solid lesions, Pseudo-papillary

Introduction

SPT, a rare pancreatic neoplasm, primarily affects women in their 20s or 30s and is associated with a generally positive prognosis due to its low malignant potential. Initially documented by Dr. Frantz in the 1950s, it was termed a "papillary tumor of the pancreas, benign or malignant." The tumor's histogenesis is still under debate, with possible origins from acinar, endocrine, ductal, or progenitor cells. Typically well-encapsulated, it represents less than 3% of exocrine pancreatic neoplasms and often originates in the body or tail of the pancreas in 50–60% of cases [1, 2]. SPT is frequently asymptomatic or mildly symptomatic, but malignancy, including metastases or vascular/perineural invasion, can occur in up to 15% of cases [3, 4].

Access this article online

Website: https://smerpub.com/ **E-ISSN: 3108-4834**

Received: 23 June 2022; **Revised:** 08 October 2022; **Accepted:** 11 October 2022

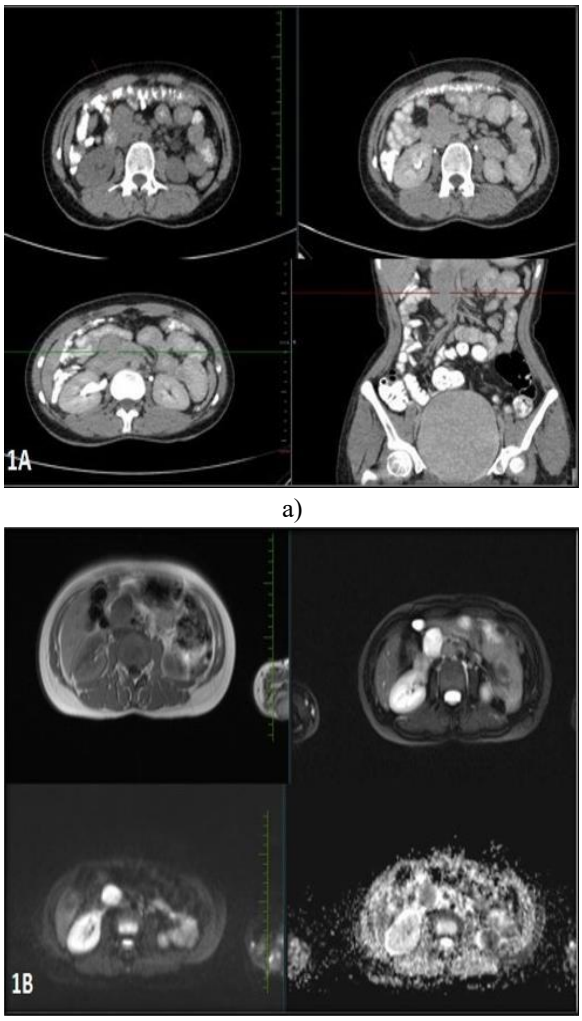
How to cite this article: Donati F, Cervelli R, Boraschi P. Frantz's Tumor: A Rare Pancreatic Neoplasm with Distinctive Features. Arch Int J Cancer Allied Sci. 2022;2(2):20-3. https://doi.org/10.51847/u0fGjt90j8

This report describes an unusual case of a 19-year-old female presenting with epigastric pain, treated with surgical resection and confirmed as a *Frantz tumor* via histopathology and immunohistochemistry.

Case Report

A 19-year-old female reported epigastric pain persisting for six months, with no prior hospitalizations. Vital signs were normal, and physical examination revealed mild epigastric tenderness without palpable masses or abnormal bowel sounds. She denied symptoms such as vomiting, jaundice, diarrhea, abdominal distension, trauma, fever, weight loss, or loss of appetite, and had no history of diabetes, hypertension, tuberculosis, or asthma. Routine laboratory tests (biochemical and hematological) showed normal results. Abdominal ultrasonography identified a solid, round-to-oval isoechoic lesion near the pancreatic head. A CT scan confirmed a well-defined, heterogeneous lesion measuring 31 x 29 x 26 mm in the pancreatic head (**Figure 1a**). MRI (Axial T1WI, T2WI) revealed a well-defined, exophytic lesion of similar size with smooth margins, hypointense on T1WI, and hyperintense on

T2WI, suggesting a predominantly solid tumor (**Figure 1b**).

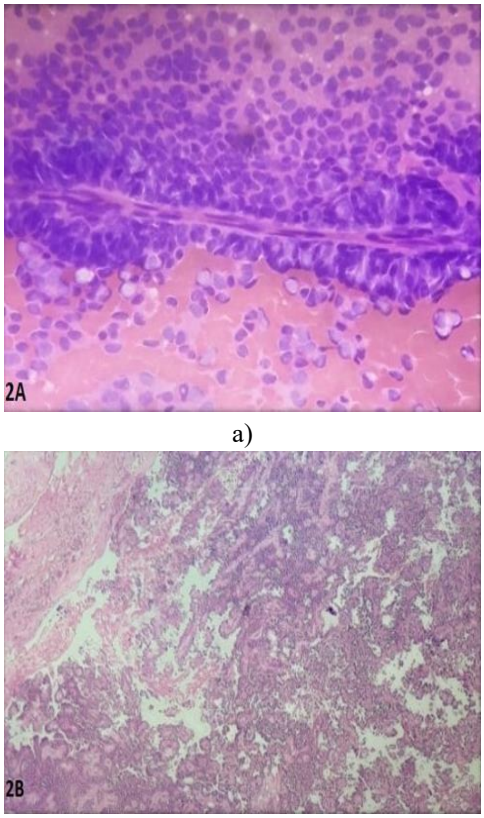


a)

b)

Figure 1. a) CT image displaying a heterogeneous, well-defined lesion (31 x 29 x 26 mm) in the pancreatic head, and b) MRI showing a well-defined, exophytic lesion with smooth margins in the pancreatic head, hypointense on T1WI and hyperintense on T2WI.

Histopathology confirmed these findings, revealing tumor cells with mild to moderate pleomorphism, nuclear grooves, dispersed chromatin, and inconspicuous nucleoli. Some areas showed myxoid change, but no increased mitotic activity or necrosis was observed (**Figure 2**). Immunohistochemistry was positive for vimentin and CD10 (**Figure 2**). The patient recovered uneventfully post-surgery and remains in good health.



a)

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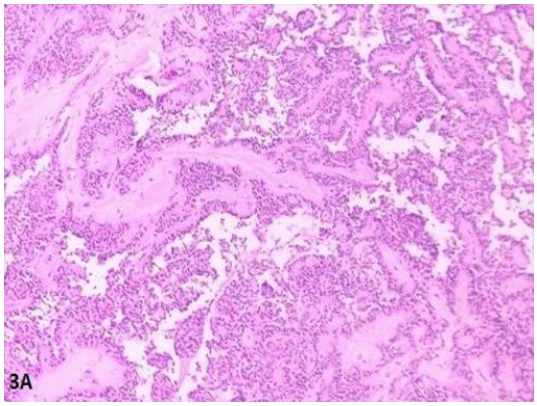
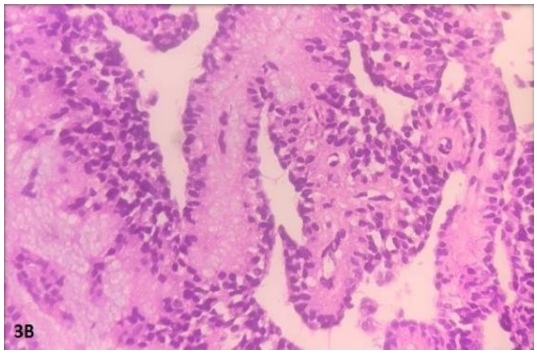
Figure 2. a) photomicrograph of cellular smears with monomorphic cuboidal tumor cells in papillary fronds around fibrovascular stroma [H&E × 40], and b) photomicrograph of a neoplasm with small, uniform tumor cells layered around hyalinized blood vessels, forming pseudopapillae [H&E × 20].

Histopathological examination confirmed the above findings. The tumor cells exhibited mild to moderate pleomorphism, characterized by nuclear grooves, dispersed chromatin, and inconspicuous nucleoli. Several areas with myxoid changes were observed. No significant mitotic activity or necrosis was noted (**Figures 3a-3c**). Immunohistochemistry (IHC) results were positive for vimentin and CD10 (**Figure 3d**). The postoperative course was smooth, and the patient is currently in good health.



a)



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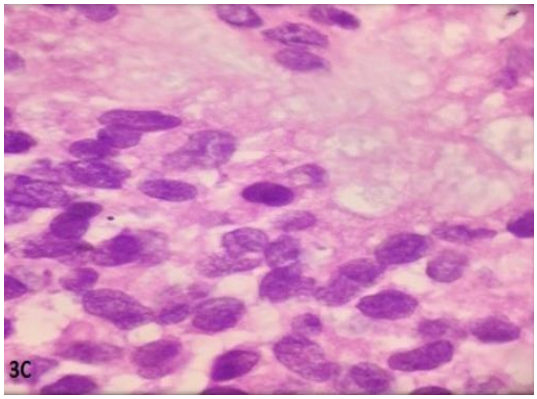

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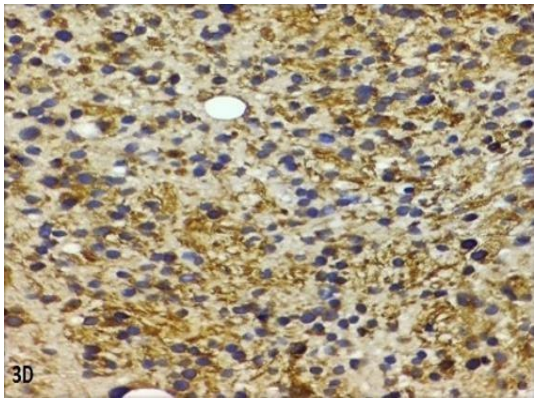

d)

Figure 3. a) photomicrograph of tumor cells in papillary formations [H&E × 20], b) photomicrograph showing mild to moderate pleomorphism in papillary tumor cells [H&E × 40], c) photomicrograph of tumor cells with hyperchromatic, pleomorphic nuclei and nuclear grooving [H&E × 100], d) photomicrograph showing vimentin positivity [H&E × 40].

Results and Discussion

SPT, a rare pancreatic tumor named *Frantz tumor* after its discovery by Dr. Frantz in 1959, has been historically labeled as papillary epithelial neoplasm, low-grade papillary neoplasm, or solid and papillary neoplasm. It primarily affects young women in their second or third decade and occasionally children. Its female predominance may be linked to an extrapancreatic origin, possibly genital ridge cells, due to the proximity of genital ridges to the pancreatic anlage during embryogenesis [5]. Mutations, particularly those affecting the Wnt signaling pathway and leading to nuclear expression of beta-catenin and vimentin, are noted in up to 90% of cases [6, 7].

Often detected incidentally on imaging [5], SPT appears as a well-demarcated, solitary pancreatic lesion. Symptoms, when present, include vague abdominal pain or, in larger tumors, a palpable mass. Rarely, it may rupture following blunt trauma or metastasize to the liver in 15% of cases, with local recurrence being uncommon [1, 5]. Microscopically, tumor cells form loose sheets with prominent stromal blood vessels, creating a pseudopapillary pattern. Solid areas may contain foamy histiocytes, cholesterol granulomas, calcification, or myxoid change [8]. Immunohistochemistry consistently shows positivity for alpha-1-antitrypsin, beta-catenin, CD56, CD10, and vimentin. Radiologically, SPTs are hypervascular, well-encapsulated, and predominantly solid with occasional cystic components. Endosonography facilitates preoperative biopsy, aiding diagnosis. Surgical resection is the primary treatment, offering a good prognosis. Limited data exist on chemotherapy or radiation, though radiation may benefit unresectable, locally advanced cases [1].

Conclusion

SPT of the pancreas, a rare entity predominantly seen in young women, exhibits low malignant potential and a

favorable prognosis following surgical resection. Regular follow-up is recommended to monitor for rare instances of local recurrence or distant metastasis.

Acknowledgments: None

Conflict of Interest: None

Financial Support: None

Ethics Statement: None

References

1. Zinner MJ, Shurbaji MS, Cameron JL. Solid and papillary epithelial neoplasms of the pancreas. Surgery. 1990;108(3):475-80.
2. Martin RC, Klimstra DS, Brennan MF, Conlon KC. Solid- pseudopapillary tumor of the pancreas: a surgical enigma? Ann Surg Oncol. 2002;9(1):35-40.
3. Torres OJM, Rezende MB, Waechter FL, Neiva RF, Moraes-Junior JMA, Torres CCS, et al. Pancreatoduodenectomy for solid pseudopapillary tumor of the pancreas: a multi-institution study. Arq Bras Cir Dig. 2019;32(2):e1442.
4. Zuriarrain A, Nir I, Bocklage T, Rajput A. Pseudopapillary tumor of the pancreas in a 17-year-old girl. J Clin Oncol. 2011;29(14):e395-6.
5. Mirminachi B, Farrokhzad S, Sharifi AH, Nikfam S, Nikmanesh A, Malekzadeh R, et al. Solid Pseudopapillary Neoplasm of Pancreas; A Case Series and Review Literature. Middle East J Dig Dis. 2016;8(2):102-8.
6. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. J Am Coll Surg. 2005;200(6):965-72.
7. Gurzu S, Bara T, Sincu M, Gabos S, Vlad DM, Bara T Jr, et al. Solid pseudopapillary neoplasm of pancreas: Two case reports. Medicine (Baltimore). 2019;98(29):e16455.
8. Alomari SA, Alfaqihi AS, Alzahrani KA, Alzahrani KH, Alzahrani KM, Alzahrani KA, et al. An Overview on the Role of Surgery in Resectable Pancreatic Tumors, Literature Review. Pharmacophore. 2021;12(4):86-91. doi:10.51847/RxWepx87LV