Case Report

Frantz's Tumor an Unusual Pancreatic Neoplasm with Rare Presentation

Abstract

A solid pseudopapillary tumor of the pancreas (SPT) is alternatively called a 'Frantz tumor.' It was first named after its discoverer in the year 1959. This tumor has been previously designated as papillary epithelial neoplasm, low-grade papillary neoplasm, and solid and papillary neoplasm of the pancreas. It is a rare pancreatic neoplasm. It typically affects young people and has a female predilection. Its histogenesis is rather debatable. Acinar, endocrine, ductal, and progenitor cells have been postulated as a possible starting point of this tumor. It has a relatively favorable prognosis with low malignant potential. It accounts for less than 3% of all exocrine pancreatic neoplasms. Although most SPTs behave as benign tumors, up to 15% of the cases can show malignancy. We are presenting a rare case of a 19-year-old female who came with epigastric abdominal pain. Surgical excision was done, and a histopathology/Immunohistochemistry examination confirmed it as a Frantz tumor.

Keywords: *Frantz tumour, Pancreas, Papillary lesions, Solid lesions, Pseudo-papillary*

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Introduction

SPT is a very rare neoplasm of the pancreas. It typically affects young women in their third decade of life and has a relatively better prognosis. It has a low malignant potential. It is also known as the 'Frantz tumor,' described first by Dr. Frantz in the 1950s and later known as a "papillary tumor of the pancreas, benign or malignant." Its histogenesis is rather debatable. Acinar, endocrine, ductal, and progenitor cells have been postulated as a possible starting point of this tumor. It is a well-encapsulated tumor and accounts for less than 3% of all exocrine pancreatic neoplasms.[1, 2] It is a well-encapsulated mass, and almost 50 to 60% of the cases arise in the body or tail region of the pancreas.[3] It is usually asymptomatic or minimally symptomatic. Although most SPTs behave as benign tumors, up to 15% of the cases can show malignancy and manifest as metastases or vascular and perineural invasion.[4]

Case report

A 19-year-old female came with epigastric pain for the last 6 months duration. She had no previous history of hospitalization. Her vitals were within normal limits. A physical examination was done, which showed mild tenderness in the epigastric region. There were no palpable lesions, and bowel sounds were normal. She gave no history of vomiting/jaundice/loose stools/abdominal distension/trauma/fever/weight loss/loss of appetite/Diabetes/ hypertension/ tuberculosis/asthma. Routine laboratory (biochemical and hematological) tests were in the normal range. Abdominal Ultrasonography revealed a solid, round to-oval isoechoic lesion in the vicinity of the head of the pancreas. Computed Tomography (CT) scan abdomen revealed a well-defined heterogeneous lesion of size 31 x 29 x 26mm in the head of the pancreas. **(Figure 1a)** MR Imaging revealed (Axial T1WI, Axial T2WI) a well-defined, exophytic lesion with a regular smooth margin of similar size in the head of the pancreas, appearing hypo intense on T1WI and hyper-intense on T2WI. Imaging suggested a tumor with a mostly solid component of the head of the pancreas (**Figure 1b**).

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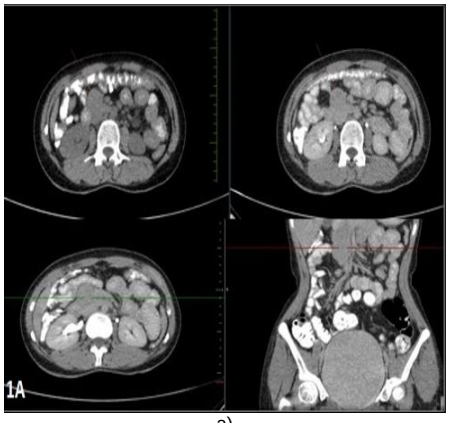

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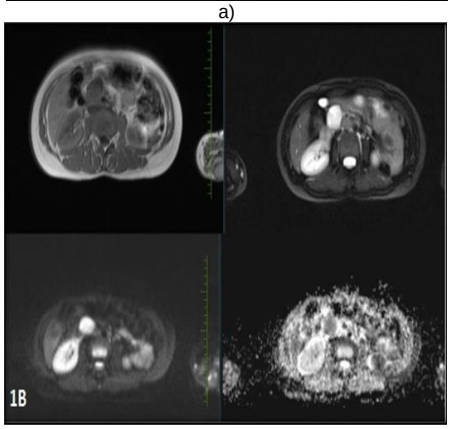

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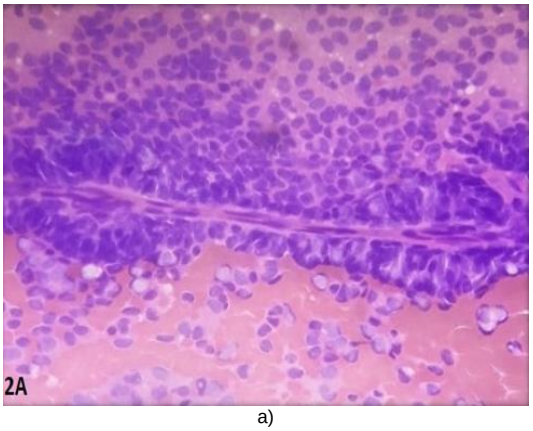
Figure 1. a) The image shows a well-defined heterogeneous lesion of size 31 x 29 x 26mm in the head of the pancreas. b) The image shows a well-defined, exophytic lesion with regular smooth margins in the head of the pancreas, appearing hypo intense on T1WI and hyper-intense on T2WI.

Endoscopic ultrasound-guided FNA revealed cellular smears showing multiple layers of monomorphic cuboidal tumor cells arranged in papillary fronds with central thin fibrovascular stroma. The tumor cells have granular eosinophilic cytoplasm, small round nuclei, fine chromatin, and occasional nuclear grooves. No significant mitotic activity **(Figure 2a)**.

The frozen section of enucleated tumor of the pancreas showed a neoplasm comprising small uniform tumor cells in multiple layers arranged around hyalinized blood vessels appearing as pseudo papillae **(Figure 2b)**.



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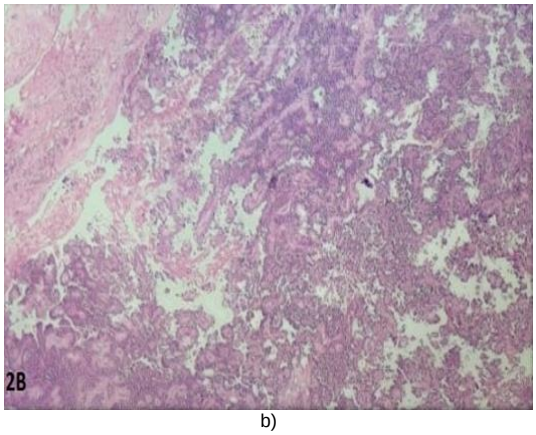

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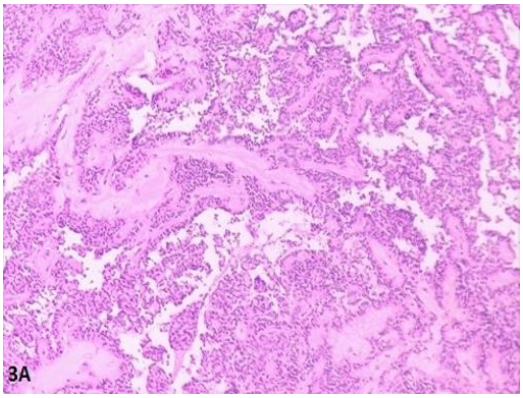
Figure 2. a) Photomicrograph shows cellular smears showing multiple layers of monomorphic cuboidal tumor cells arranged in papillary fronds with central thin fibrovascular stroma [H&E × 40]. b) Photomicrograph shows a neoplasm comprising of small uniform tumor cells in multiple layers arranged around hyalinized blood vessels appearing as pseudo papillae [H&E × 20]

On histopathology above findings were confirmed. The individual tumor cells showed mild to moderate pleomorphism, with nuclear grooves, dispersed chromatin, and inconspicuous nucleoli. Few areas of myxoid change were seen. There was no evidence of increased mitotic activity or necrosis **(Figures 3a-3c)**. Immunohistochemistry (IHC) was positive for vimentin and CD10 **(Figure 3d)**. The post-operative period was uneventful, and the patient is presently doing well.



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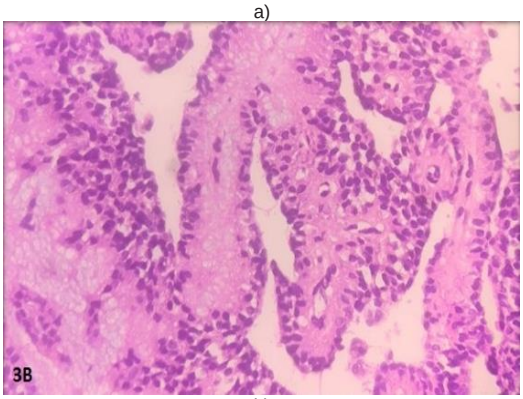

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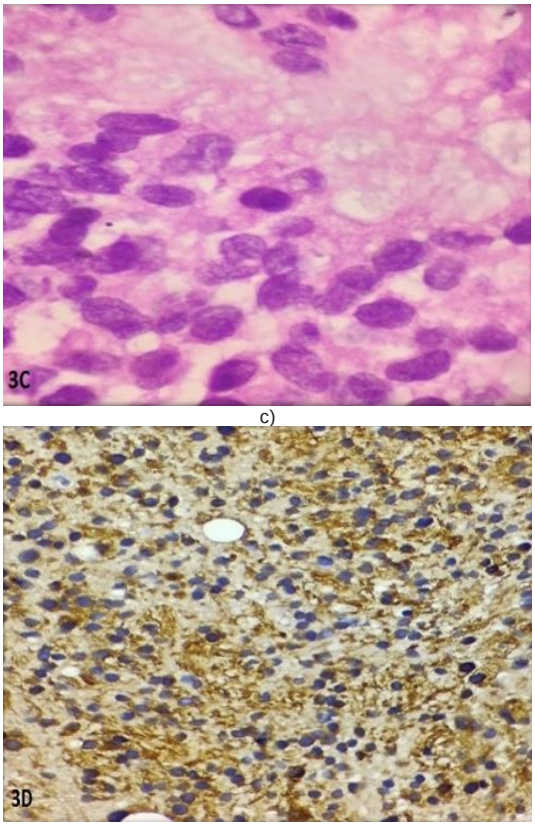

c)

d)

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

Results and Discussion

SPT lesion of the pancreas is a very rare tumor. First described by Dr. Frantz in 1959, hence named as Frantz tumor. This tumor has been previously designated as papillary epithelial neoplasm, low-grade papillary neoplasm, solid and papillary neoplasm, etc. It mostly occurs in comparatively younger women in the second or third decade. It is also occasionally diagnosed among children. Similarities between SPT and ovarian surface cells usually show an extra pancreatic origin of cells mainly from the genital ridge cells, thereby explaining female preponderance. It is also known that during the embryonic stage, the genital ridges are in the vicinity of pancreatic anlage.[5] Other studies consider the development of SPNs to various mutations. The nuclear expression of b-catenin and vimentin secondary to this mutation and its interference with the Wnt signaling pathway is among the most studied ones. It is reported to be present in up to 90% of these tumors.[6, 7]

It is usually an incidental finding on radiologic examination.[8] Typically, a tumor presents as a well-demarcated, solitary lesion arising from the pancreas. Symptoms are nonspecific, such as abdominal discomfort or pain and sometimes a palpable abdominal mass (in large SPTs). In some instances, it even presents as a rupture, usually seen following blunt abdominal trauma.[5] Metastases are also reported (15% of cases), which can go to the liver. Although quite rare, local recurrence is also seen.[1]

On microscopy, the tumor cells are organized in loose sheets with prominent blood vessels in the stroma, imparting the pseudopapillary appearance. Solid areas may be seen containing foamy histiocytes, cholesterol granulomas, calcification, and myxoid change.[9] On IHC, these tumor cells are invariably positive for alpha-1- antitrypsin, beta-catenin, CD56, CD10, and Vimentin. Radiologically, these tumors appear as hypervascular, well-encapsulated, round lesions with solid and occasionally focal cystic components. Endosonography may provide us with biopsy material with the possibility of pre-operative pathologic diagnosis. Surgically complete resection remains the only treatment with a better prognosis. There is limited literature regarding the use of chemo-radiation for this tumor. A significantly favorable outcome to radiation therapy, especially in locally advanced cases which are unresectable, has been reported.[1]

Conclusion

SPT of the pancreas is extremely rare, seen predominantly in the young female population with a low potential for malignancy, and usually has a favorable long-term prognosis after surgical resection. However, a close follow-up is strongly advised to check for possible local recurrence or distant metastasis.

Acknowledgments
None.

Conflict of interest
None.

Financial support
None.

Ethics statement
Prior permission was taken from the Institutional Ethics Committee (DPU-IEC) to conduct this study.

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