Case Report

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Cytodiagnosis Of Solid Pseudopapillary Neoplasm: A Rare Pancreatic Tumour

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Abstract

Solid Pseudopapillary neoplasm (SPN) of the pancreas is a rare, low-grade malignant tumour that primarily affects young females. This case report presents a 30-year-old female diagnosed with SPN, detailing her clinical presentation, diagnostic workup and histopathological findings. The objective is to contribute valuable insights into the clinical management and prognosis of this unusual pancreatic neoplasm. The prognosis for SPN is generally favourable, with a low propensity for distant metastasis. However, long-term follow-up is crucial, given the potential for late recurrence. The patient presented with nonspecific abdominal pain and discomfort for several months. FNAC was done showing tumour cells arranged in papillary pattern suggestive of solid pseudopapillary neoplasm. Cytomorphological evaluation of smears was done for cellularity, cell type, nuclear details, and background and cytologic diagnosis were made which was later on confirmed by histopathology. Surgical exploration led to the successful resection of the tumour, and the patient's postoperative course was uneventful.

Keywords: - pseudopapillary neoplasm, Pancreas, Frantz's tumour, Cytomorphology.

INTRODUCTION

Solid Pseudopapillary neoplasm (SPN) of the pancreas is an exceedingly rare pancreatic tumour, accounting for approximately 1-2% of all pancreatic neoplasms. SPN predominantly affects young females.[1] The average age at diagnosis is around 30 years, as observed in the case of our 30-year-old female patient. While the exact etiology remains unclear, recent hypothesis is that this neoplasm may have been derived from genital-ridge-related cells attached to the pancreatic tissue during development.[2]

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Patients with SPN often present with vague abdominal symptoms, including pain and discomfort. These symptoms can be chronic, as seen in our patient, and are usually located in the epigastric or periumbilical region. Notably, SPN is often an incidental finding on imaging or during routine physical examinations, as it tends to grow slowly and may not cause alarming symptoms until it reaches a significant size.[3]

Imaging studies are essential for the evaluation and diagnosis of SPN. Computed tomography (CT) and magnetic resonance imaging (MRI) are the primary modalities used to assess the tumor.[4] SPN typically appears as a well-defined, mixed solid and cystic mass, with haemorrhagic or necrotic components. These findings are consistent with the heterogeneous nature of the tumour. Accurate imaging is critical for surgical planning and guiding biopsy procedures.[5]

Histologically, SPN is characterized by a distinctive appearance. The tumour typically consists of pseudopapillary structures with fibrovascular cores, enclosed by a fibrous capsule. This unique histopathological pattern is the basis for its name, "pseudopapillary." Immunohistochemistry plays a crucial role in confirming the diagnosis, with SPN commonly exhibiting positivity for beta-catenin and CD10.[6] Upon histopathological examination of the surgically resected specimen, SPN displays a characteristic appearance. The tumour comprises pseudopapillae, composed of small, uniform cells surrounding a central fibrovascular core. Additionally, the tumour is often encapsulated by a fibrous sheath. Immunohistochemically, SPN is marked by nuclear and cytoplasmic beta-catenin expression, along with positivity for CD10, further confirming the diagnosis.[7]

Surgical resection is the cornerstone of SPN management. In our patient's case, successful surgical exploration resulted in complete resection of the tumour, which is the primary curative approach. The extent of resection may vary, from enucleation to distal pancreatectomy or Whipple procedure, depending on the tumour's size, location, and involvement of adjacent structures. A minimally invasive approach may be considered when feasible, offering potential benefits in terms of recovery.[8]

The prognosis for SPN is generally favourable. The tumour exhibits a low propensity for distant metastasis, making complete surgical resection curative in most cases. Long-term survival rates are high, with a reported 5-year survival rate exceeding 95%. However, it is essential to note that SPN has the potential for late recurrence, underscoring the importance of vigilant, long-term follow-up. Adjuvant therapies such as chemotherapy or radiation are rarely indicated, given the tumour's indolent behaviour.[9]

CASE REPORT

A 30-year-old female presented with abdominal pain and a palpable, nontender, upper abdominal mass, abdominal discomfort, nausea, vomiting and early satiety. Computerised Tomography of abdomen showed pancreas with a well circumscribed, encapsulated, hypodense, heterogenous lesion with cystic degeneration. Endoscopic ultrasound guided transgastric FNAC was done which showed tumour cells arranged in papillary pattern suggestive of solid pseudopapillary neoplasm. Cytomorphological evaluation of smears was done for cellularity, cell type, nuclear details, and background and cytologic diagnosis were made which was later on confirmed by histopathology.

Cellular smear studied were moderate to hypercellular showing papillae formation, discrete cells and occasional pseudorosettes. The papillae formation showed delicate vascular cores and covered by two or several layers of cells. The individual tumour cells had round to oval eccentric nuclei with bland nuclear chromatin with nuclear grooves and convolution and moderate amount of pale pink cytoplasm. Background was hemorrhagic (Figure 1).

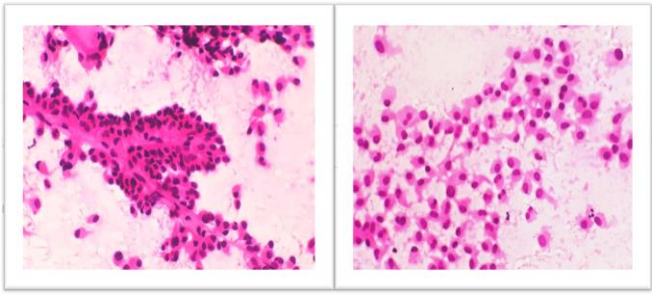

Figure 1: EUS guided FNAC showing papillae formation with vascular cores. Individual cells showing plasmacytoid appearance with convoluted nuclei and nuclear groves.

A subsequent Biopsy (H & E stain) showed show sheets and cords of cells having round to ovoid uniform nuclei, convoluted and grooved having fine chromatin and inconspicuous nucleoli with scant eosinophilic cytoplasm, arranged around delicate fibrovascular septa. There were marked degenerative changes including microcysts, haemorrhage, aggregates of foamy cells and cholesterol granulomas forming a pseudopapillary pattern and pseudorosettes

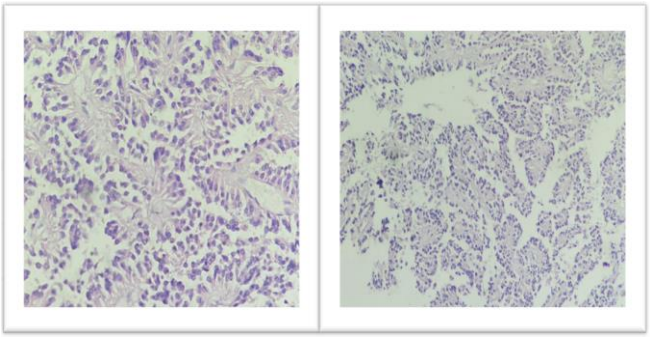

Figure 2: Biopsy of pancreatic mass showing sheets and cords of cells with pseudopapillae around fibrovascular stalks and rosette-like structures [HE]

DISCUSSION

Solid Pseudopapillary neoplasm (SPN) of the pancreas is an uncommon tumour, predominantly affecting young females, as exemplified by the case of our 30-year-old female patient. This discussion aims to provide insights into the clinical, cytological, and histopathological aspects of SPN, emphasizing the importance of accurate diagnosis and multidisciplinary management. Our patient's clinical presentation was consistent with the typical features of SPN. Abdominal pain, abdominal mass, and nonspecific gastrointestinal symptoms such as nausea, vomiting, and early satiety are frequently reported. These symptoms often result from the mass effect of the tumour as it enlarges within the pancreas. The palpable, nontender upper abdominal mass noted in our case further underscores the significance of clinical examination in early detection.[10]

Endoscopic ultrasound-guided fine-needle aspiration cytology (EUS-FNAC) played a pivotal role in establishing the preliminary diagnosis of SPN in our patient. The cytological evaluation revealed characteristic features of SPN, with tumour cells arranged in papillary patterns. These cells exhibited a distinctive cytologic profile, including eccentric round to oval nuclei, bland nuclear chromatin with nuclear grooves and convolution, and moderate pale pink cytoplasm. The presence of delicate vascular cores within the papillae and the formation of pseudorosettes were also noted.[11]

In our case cytological findings were consistent with previous reports, highlighting the utility of EUS-FNAC as a valuable diagnostic tool for SPN. Accurate cytological diagnosis can aid in early treatment planning and surgical intervention, contributing to optimal patient outcomes. Histopathological examination of the subsequent biopsy specimen provided definitive confirmation of SPN. The tissue sections demonstrated characteristic features, such as sheets and cords of cells arranged around delicate fibrovascular septa. The tumour cells displayed round to ovoid nuclei, convoluted and grooved with fine chromatin and inconspicuous nucleoli. These findings align with the known histological profile of SPN.[12]

Additionally, the biopsy revealed marked degenerative changes, including microcysts, haemorrhage, foamy cell aggregates, and cholesterol granulomas, forming a pseudopapillary pattern and pseudorosettes. These degenerative changes are hallmark features of SPN and further substantiated the diagnosis.[13]

The successful management of SPN necessitates a multidisciplinary approach. Surgical resection remains the cornerstone of treatment, and the choice of surgical procedure depends on factors such as tumour size, location, and involvement of adjacent structures. Our case underscores the importance of complete resection, which is often curative, given the tumour's low propensity for distant metastasis.[14]

The prognosis for SPN is generally favourable, with a reported 5-year survival rate exceeding 95%. However, long-term follow-up is crucial, as late recurrences have been documented. The rarity of SPN warrants ongoing research to better understand its molecular basis and refine treatment strategies.[15]

CONCLUSION

Solid pseudopapillary tumours of the pancreas are rare tumours. A high index of suspicion along with knowledge about cyto-histopathological features of this tumour is essential for early diagnosis and further management as early diagnosis is found to be associated with an excellent outcome.

Conflict of interest

None

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