CASE REPORT

# Unveiling a Rare Pancreatic Tumor in Bangladesh: Solid PseudopapillaryTumor

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## Introduction

Solid Pseudopapillary Tumors are rare pancreatic neoplasms first identified as Hammoudi or Franz tumors, later classified by the WHO as SPTs in 1996. These tumors primarily affect young non-Caucasian women, with a male-to-female ratio of 1:11. Surgical excision remains the standard treatment. Solid Pseudopapillary Tumors (SPTs) within the pancreas are rare, accounting for less than 3% of pancreatic neoplasms. They predominantly occur in young women and are characterized by low malignant potential with high surgical cure rates. This report details the case of a 14-year-old girl in Bangladesh undergoing a successful Whipple's procedure for an SPT.

This case report aims to share our unique findings, which may contribute to understanding and knowledge of this rare medical condition and its treatment options.

## Case Report:

A 14-year-old girl presented at our institution with complaints of central and right upper abdominal pain for 2

months. She mentioned that her pain was constant, dull, aching, and moderate in severity with no radiation. She also complained of feelings of fullness and early satiety. There was no history of jaundice, haematemesis, melaena, loss of appetite, or weight loss.

On general examination, she was anemic but not icteric. On abdominal examination, she had mild tenderness in her epigastric region on deep palpation.

She underwent a Whipple procedure, and per operatively, a growth was found at the head of the pancreas with no evidence of liver metastasis, ascites, or peritoneal seedling.

Her post-operative course was uneventful.

The Histopathology report confirmed the diagnosis:

Sections of the pancreas show a neoplasm composed of uniform polygonal and oval cells arranged in a solid and papillary-like pattern. Confirming Pseudopapillary tumor of the pancreas

Margins: All the margins were clear

LN: 10 in number, all showed reactive hyperplasia. No Metastasis was seen.

No intra-abdominal lump was palpable, and there was no organomegaly. All other systemic examinations were regular.

Her lab investigations showed Hb of 8 g/dl. Other routine blood investigations included liver function tests and tumor markers( AFP, Ca-19-9, CEA).

**USG of W/A :**

Significant soft tissue mass in the right hypochondrium

with focal calcification, not separated from the liver and adherent to the head of the pancreas.

**CT scan of W/A:**

A well-defined soft tissue mixed density mass (6.7 cm 6.0cm) arising from the head of the pancreas compresses the duodenum.

Foci of calcifications are noted within the lesion. CBD was regular in Caliber.

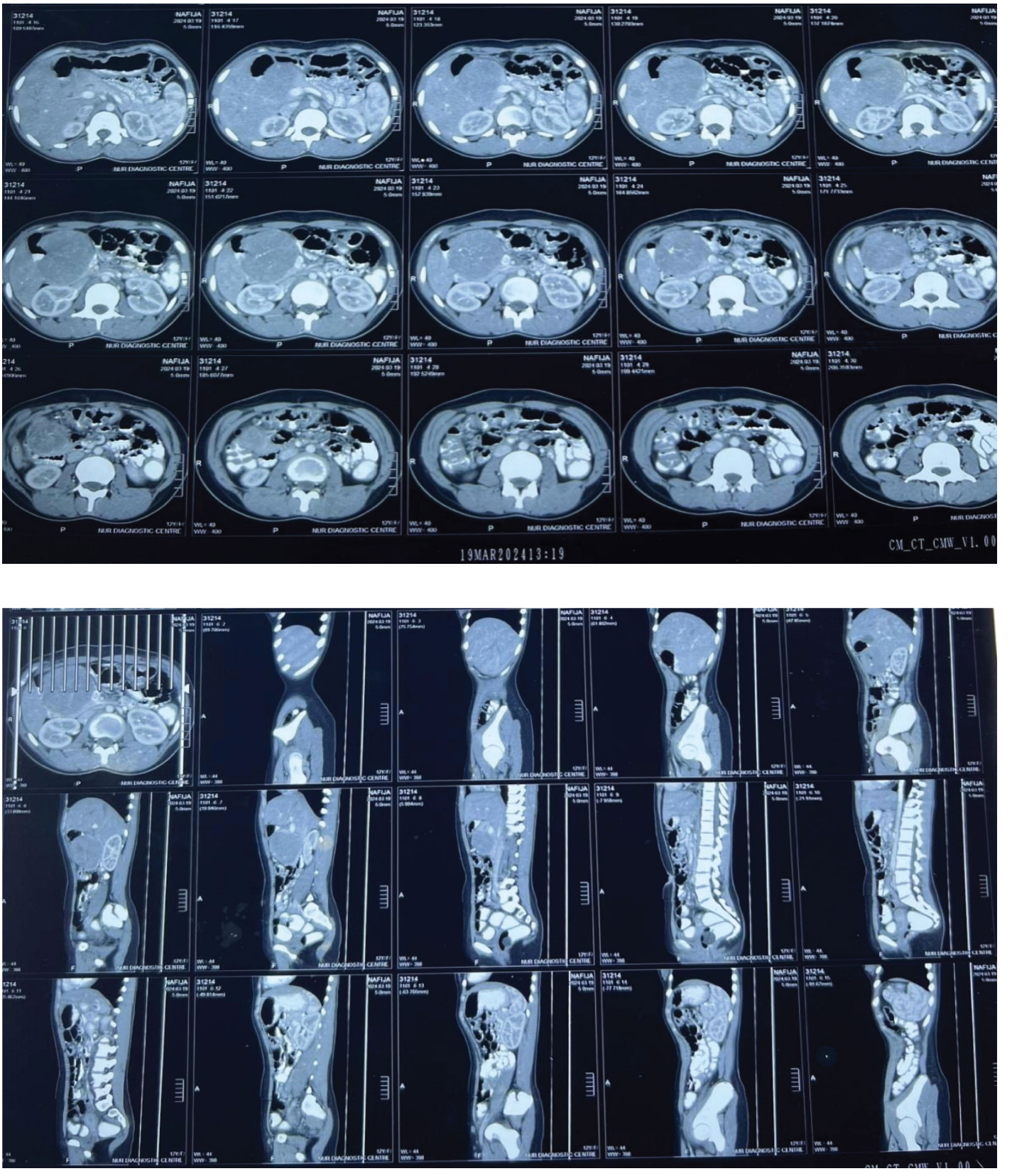


Figure-1:

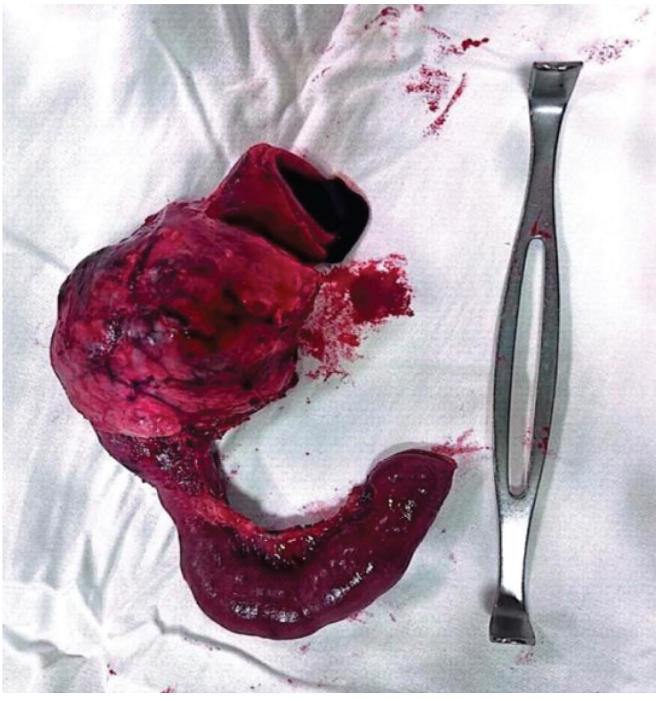

Figure-2:

## Discussion

SPTs are typically present in the pancreas but have also been observed in other areas like the mesentery and retroperitoneum. Although their metastatic potential is low, metastasis occurs in about 10% of cases, commonly affecting the liver and lymph nodes. Advances in imaging and immunohistochemical markers, such as anti-vimentin and Ki-67, play crucial roles in diagnosis. 90% of those affected happen to be young women, with an average age of 22 years, as seen in one study[4], and an average age of 28 years, as seen in another report[5]. The patient age group varies widely, ranging from two to eighty-five years.

In some studies, it has been postulated that the proximity of the primordial pancreatic cells to the ovarian ridge during the embryonic phase significantly contributes to the increased prevalence seen among female patients.

Patients may have no symptoms. Some may have unexpected symptoms, including bloating, discomfort, and stomach pain. Others may exhibit jaundice or vomiting as compression symptoms.

Ultrasound and CT scans are examples of imaging that can aid in diagnosis. The most effective examination is magnetic resonance imaging (MRI).

It is best to avoid preoperative biopsy under radiological or endoscopic supervision since it might result in problems including hemorrhage, pancreatic and biliary fistulas, as well as the possibility of tumor dispersion and the conversion of a tumor with a favorable prognosis into an aggressive tumor [9].

Immunohistochemical studies and pathological analysis are typically used to diagnose SPT positively, which is still challenging. Anti-vimentin antibody immunostaining, a marker for germline cells, yields positive results in almost 90% of cases, among other specific indicators that have been identified. In nearly 50% of patients, anti-alpha-1-antitrypsin and anti-neuron specific enolase (NSE) antibodies are positive[13]. An unfavorable outcome of SPT and the possibility of malignancy may be predicted by positive immunoreactivity for Ki-67[16].

Some differentials that should be kept in mind while diagnosing a case of SPT include pancreatic pseudocyst, calcified hemorrhagic pseudocyst, microcystic adenoma, pancreato-blastoma, non-functioning islet cell tumor, mucinous cystic neoplasm, etc.

Surgery is the only treatment option, with a 5-year survival rate of 95%[11] and an estimated 10-year survival of 93%[14]. Because of its propensity for degeneration, the tumor and its capsule are removed as part of the curative treatment for SPT. Depending on the location, this removal can be accomplished by enucleation, distal pancreatectomy, or pancreatoduodenectomy (the most typical operation undertaken).

Adjuvant therapy's function in treating SPT is unknown, although options for chemotherapy and radiotherapy have been investigated. Nevertheless, adjuvant treatment is not necessarily due to the tumor's high resectability rate and low propensity for malignancy.[7, 8]

## Conclusion:

Due to their varied clinical presentations, Diagnosing Solid Pseudopapillary Tumors requires a high index of suspicion. Early surgical intervention

provides excellent outcomes, reinforcing the need for awareness and timely management.

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