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SOLID PSEUDOPAPILLARY NEOPLASM: A RARITY REVISITED

Radiology

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ABSTRACT

Solid pseudopapillary neoplasm (SPN) is a rare exocrine tumour of the pancreas that has an indolent course, though around 15% cases exhibit a definite malignant potential with distant metastasis. It is predominantly seen in young non-Caucasian women between second and third decades. It is imperative to accurately diagnose SPN so that appropriate management can be initiated. We report a case of a 20 year old Indian female who presented with epigastric pain and lump for a year. Laboratory data were unremarkable. The diagnosis was made on imaging and confirmed post-operatively on histopathology.

KEYWORDS

Solid pseudopapillary neoplasm, SPEN, cystic pancreatic tumour, SPT.

Introduction:

Solid pseudopapillary neoplasm (SPN) of the pancreas is an uncommon pancreatic tumour which has a low grade malignant potential and accounts for 0.9% to 2.7% of all pancreatic exocrine neoplasms [1]. It was first described by Frantz in 1959 [2]. In 2010, the World Health Organization (WHO) designated this tumor as solid-pseudopapillary neoplasm (SPN) and discouraged the use of synonyms namely solid-pseudopapillary tumour (SPT), solid-cystic tumour, papillary-cystic tumour, solid and papillary epithelial neoplasm (SPEN) and Frantz tumour [1]. Our case highlights the characteristic CT features of SPN along with the unusual enhancement characteristics.

Case Report:

We report a case of a 20 year old female who presented with chief complaints of abdominal lump and non-radiating epigastric pain for one year. On examination, a non tender, firm mass was noted in the epigastrium. Laboratory parameters, including liver function tests, serum amylase, lipase and tumour markers (CEA, CA-125, CA19-9 & AFP) were within normal limits.

A multiphasic contrast-enhanced computed tomography scan was performed with neutral oral and intravenous iodinated contrast. It showed a large, well-circumscribed mass measuring 9.1 × 8.6 x 7.1 cm, containing cystic and solid components and arising from the proximal and mid aspects of the body and neck of pancreas. On non-contrast CT (NCCT), multiple hyperdense areas (mean HU: 52) were noted, consistent with hemorrhagic foci (Figure 1). No calcification was seen. The mean Hounsefield attenuation (HU) value of the solid portion of mass and uninvolved pancreatic parenchyma, respectively, on various phases were as follows: 39 HU and 47 HU on NCCT, 97 HU and 71 HU on arterial phase, 159 HU and 126 HU on portal phase, 146 HU and 105 HU on venous phase while 98 HU and 84 HU on delayed phase (Figure 2). The solid enhancing areas were present in the periphery of the mass. A progressive fill-in of the enhancement was seen on subsequent phases (Figure 3). Main pancreatic duct was not dilated. Fat planes with adjacent structures were maintained without any evidence of invasion. The diagnosis of solid pseudopapillary neoplasm was rendered.

Central pancreatectomy was performed with resection of gastro-epiploic lymph nodes. Grossly, the tumour was a circumscribed globular mass measuring 11x10x6 cm. The cut section showed multiple hemorrhagic, necrotic and cystic areas. Microscopically, a well demarcated tumour was seen separated from the surrounding pancreatic parenchyma by a thick fibrous capsule. The tumour cells were arranged in solid sheets with pseudopapillae. The cells had moderate eosinophilic cytoplasm with hyperchromatic central to eccentric nuclei. Areas of foamy histiocyte collections, few eosinophilic globules, cystic degeneration and foci of necrosis were also noted (Figure 4). There was no stippled chromatin, lymphovascular or capsular invasion. The resected nodes showed reactive hyperplasia. On immunohistochemistry (IHC), the tumour cells showed positivity for beta catenin (nuclear), progesterone receptor (PR), NSE and CD56. Ki-67 labelling index was less than 1%. The cells were negative for chromogranin-A (Figure 5). These features were compatible with the diagnosis of SPN.

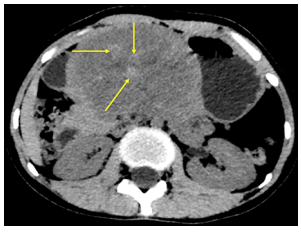

Figure 1: Axial non-contrast CT scan of the abdomen.

Multiple hyperdense areas (arrows) are seen within the tumour, consistent with haemorrhage.

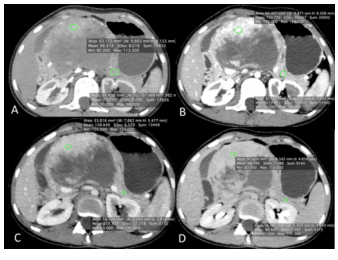

Figure 2: Axial contrast enhanced CT of the abdomen. (A) Arterial phase, (B) Portal phase, (C) Venous phase, (D) Delayed phase.

The images show enhancement of the solid areas more than the uninvolved pancreatic parenchyma. (Green circle: Region of interest).

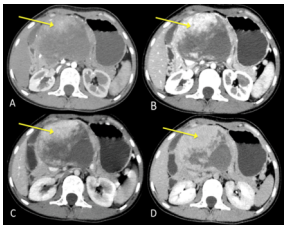

Figure 3: Axial contrast enhanced CT of the abdomen. (A) Arterial

phase, (B) Portal phase, (C) Venous phase, (D) Delayed phase.

The images show peripheral solid enhancing areas (arrows) with progressive fill-in of the contrast.

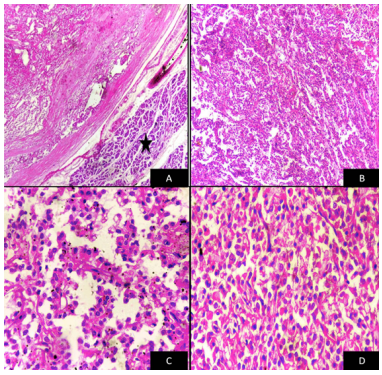

Figure 4: Microphotographs

(A) Well circumscribed tumour with adjacent normal pancreatic parenchyma (star), HE x 40

(B) Tumour cells composed in solid sheets and pseodopapillae, HE x 40

(C) and (D): Cells lining pseudopaillae and papillae, HE x 400

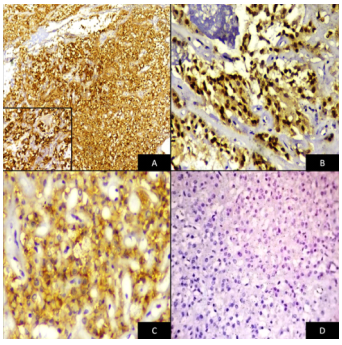

Figure 5: Immunohistochemistry

(A) Beta catenin showing diffuse positivity, IHC x100 and IHC x 400 (inset)
(B) Progesterone receptor showing nuclear positivity, IHC x 400
(C) CD56 membranous positivity, IHC x 200
(D) Chromogranin A showing negative staining, IHC x 100

Discussion:

SPN has a strong predilection for non-Caucasian women with a mean age of 28 years, which was concordant in our case. However, it can occur in men and children. The age group of affected men is slightly older than that of the female patients [3]. SPNs generally have a benign behaviour with favourable prognosis. Malignant transformation can occur in around 15% cases. Malignant SPN is more common in older patients, with a male preponderance [4]. The most common location of SPN within the pancreas is body and tail [5]. However, in the present case, the tumour was located at neck and the proximal & mid aspects of the body. Though the vast majority of SPNs arise in the pancreas, they can rarely develop outside the pancreas [6].

The clinical features of SPN are non-specific. The mass is usually non tender but obstructive or compressive symptoms may occur. The pancreatic enzymes and tumour markers are generally within normal limits.

Surgical resection is the mainstay of treatment. There is no significant role of radiotherapy or chemotherapy in treatment of SPN at present [7].

Histologically, SPN is characterized by pseudopapillae and solid sheets. Other features include nuclear grooving and foamy macrophages. SPNs show a demarcation from the surrounding pancreas by a fibrous capsule. The present case showed similar findings, leading to the diagnosis of SPN, which was confirmed by IHC. Cases with capsular lymphovascular or nerve sheath invasion and presence of synchronous metastasis have an aggressive course[4,8]. The microscopic differential diagnosis includes pancreatic neuroendocrine tumours which may be distinguished on immunohistochemical (IHC) examination, most notably, by Beta catenin[9] (Table 1).

Table 1: IHC markers in the differential diagnosis of SPEN (Solid pseudopapillary neoplasm) and pancreatic NET (Neuroendocrine tumour).

	SPEN	NET	Present case
Beta Catenin	+	-	+
Progesterone receptor	+	-/+	+
Chromogranin-A	-	+	-
Synaptophysin	+	+	NA
NSE	+	+	+
CD 56	+	+	+

SPN, on imaging, is a well-demarcated neoplasm with a mean diameter of 9 cm. The appearance ranges from predominantly cystic to solid, reflecting the degree of hemorrhagic degeneration and necrosis. Smaller tumours tend to be more solid and less sharply circumscribed [10]. CECT shows enhancement of the thick capsule with early peripheral heterogeneous enhancement in the arterial phase and progressive filling-in of the tumour in portal, venous and delayed phases, as was seen in our case. The enhancement in all phases is typically less than that of the surrounding uninvolved pancreatic parenchyma [11]. Contrary to this, our case showed increased enhancement of the solid portion as compared to the uninvolved pancreatic parenchyma on all phases. Typically, the solid enhancing areas are located in the periphery of the tumour and the cystic/necrotic areas occupy the central part. Areas of haemorrhage, fluid-fluid or fluid-debris levels, presence of a capsule are important clues to the diagnosis of SPN. Calcifications in the periphery are noted in 30% cases on CT [12].

The MRI features of SPN mirrors those seen on CT, although enhancement of solid components and fibrous pseudocapsule are better seen. The mass appears heterogeneously hyperintense on T2WI. Areas of high signal on T1WI and low signal on T2WI can help determine the presence of hemorrhagic products which helps in limiting the differential diagnosis. The presence of fluid-fluid or fluid-debris levels are quite suggestive of SPN. The fibrous capsule appears as hypointense on T2WI. On post gadolinium T1WI, early peripheral heterogeneous enhancement of the solid components is seen with progressive fill-in of the contrast material [10].

Metastasis occurs in about 15% of all cases of SPNs, the most common sites being liver, regional lymph nodes, omentum, mesentery and peritoneum. Morphology of liver metastasis is similar to the primary tumour. Malignant SPNs may also cause invasion of adjacent structures including blood vessels, stomach, duodenum and spleen [7].

CONCLUSION:

SPNs are uncommon neoplasms of the pancreas which require prompt diagnosis, as they have a low grade malignant potential. Histological features and immunohistochemical markers are necessary for definitive diagnosis. Surgical excision is the treatment of choice. It has an excellent prognosis when completely resected. The typical features of SPN on CT include a large, well-circumscribed & encapsulated tumour containing areas of hemorrhage, peripheral curvilinear calcification and heterogeneous peripheral solid enhancing areas with progressive fill-in of the contrast. Our case highlights the enhancement of solid areas more than the adjacent pancreatic parenchyma on all phases of CECT and the uncommon location of SPN in neck & proximal body. Knowledge of characteristic CT features of SPN as described herein, can help make an accurate preoperative diagnosis and thus, aid in complete resection of the tumour which is usually curative and even recurrences can be treated with a re-surgery.

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