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Case Report

Giant Solid-Pseudopapillary Neoplasm of the Pancreas in a Child

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Abstract

Solid-pseudopapillary neoplasm of the pancreas (SPPNP) is a primary epithelial neoplasm of the exocrine pancreas with borderline malignant potential. The unusual features included the 'giant' size and the absence of metastases/ capsular or vascular invasion despite the large size and predominantly solid nature. At two years following complete excision, she is asymptomatic and the imaging shows no recurrence/metastases.

Keywords:Giant; solid pseudopapillary neoplasm; pancreas; child

INTRODUCTION

Solid-pseudopapillary neoplasm of the pancreas (SPPNP) is a distinctive, relatively rare primary epithelial neoplasm of the exocrine pancreas with borderline malignant potential [1-15]. Herein, certain atypical clinicopathological features of this neoplasm are presented.

CASE REPORT

A 14-year female presented with an abdominal mass and dull aching pain since 6 months. A large, firm, nontender fixed mass occupied the epigastric and umbilical quadrants. Abdominal CECT scan delineated a well circumscribed, encapsulated, mixed density, patchily enhancing, non-calcified mass replacing the pancreatic body and tail; there were no hepatic deposits/ ascites/ lymph node enlargement. The capsule enhanced at places but there was no vascular invasion (Figure 1). The baseline hematological and biochemical investigations were normal. At laparotomy, the mass was well encapsulated and had a smooth glistening surface. A distal pancreatectomy with gross total excision of the mass and a thin rim of the adjacent pancreas was performed.

The tumor measured 14x12x10 cms and weighed 1450 g (Figure 2). The cut surface was variegated; the solid portion was soft to firm, grayish white/yellow. The intervening cystic spaces were septated with hemorrhagic, granular and friable contents.

Histologically, the tumor was primarily solid with extensive small, poorly supported blood vessels (Figure 3A-3C). The solid component comprised of sheets, trabeculae and nests of uniform, polygonal epithelioid cells around a network of delicate blood vessels, surrounded by varying amounts of collagen. The tumor cells had eosinophilic cytoplasm and monomorphic round to oval nuclei. Considerable secondary degenerative changes (hemorrhage, necrosis) were observed. The remaining cells surrounded delicate hyalinized fibrovascular stalks forming pseudopapillae (Figure 3B and 3C). Neither lymph node involvement nor capsular invasion was seen. A thin rim of normal pancreas was identifiable at the mass –pancreas interface (Figure 3A). A final diagnosis of SPPNP was made.

At two years follow up, she is asymptomatic and the surveillance imaging (biannual chest X ray, CECT) is normal.

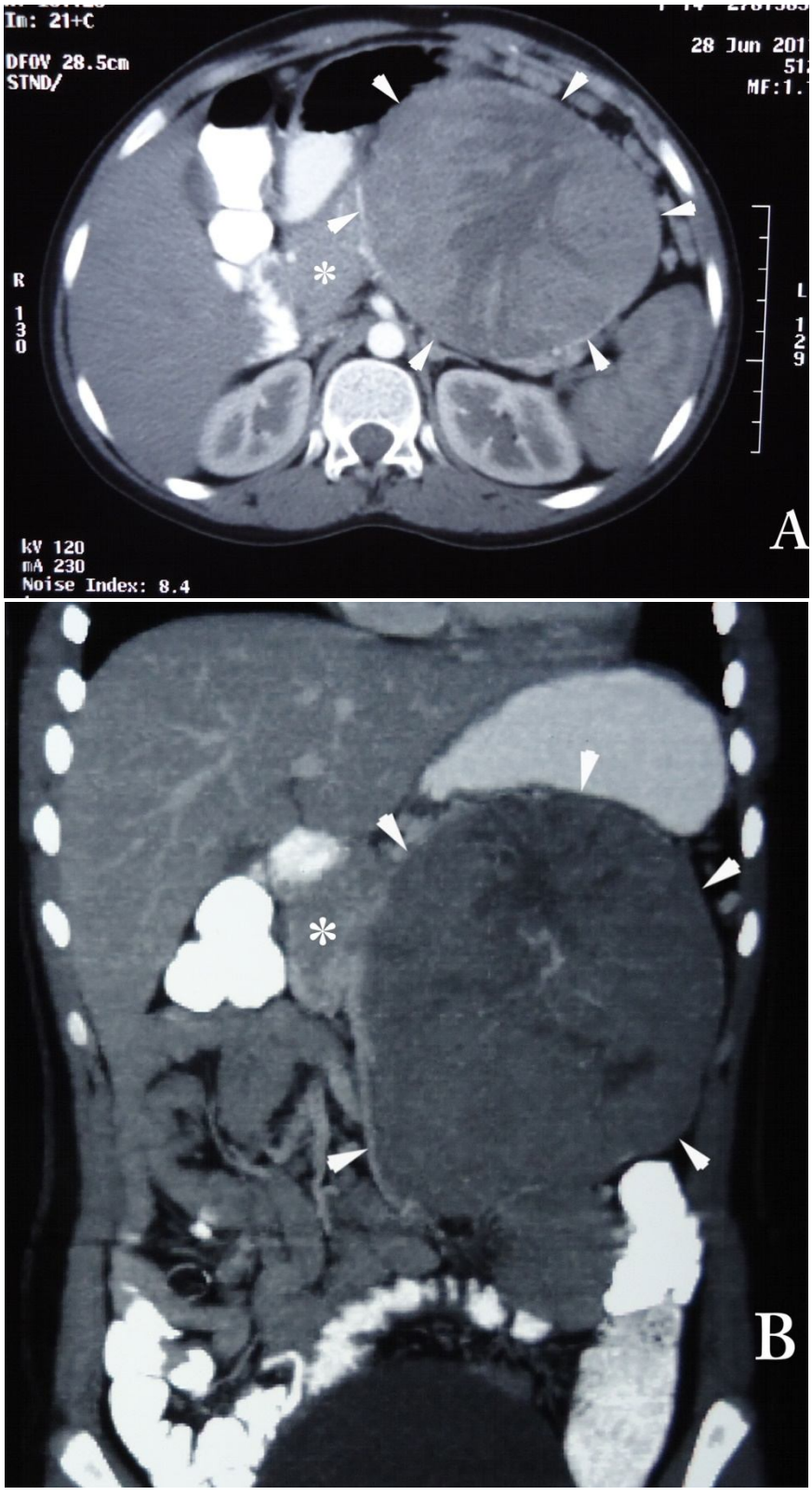

Figure 1: Abdominal CECT (A: Axial; B: Coronal) showing a well encapsulated, patchily enhancing mass replacing the pancreatic body and tail. Note the capsule enhancing at places (small arrows), the pancreatic head (*) and impending gastric outlet obstruction.

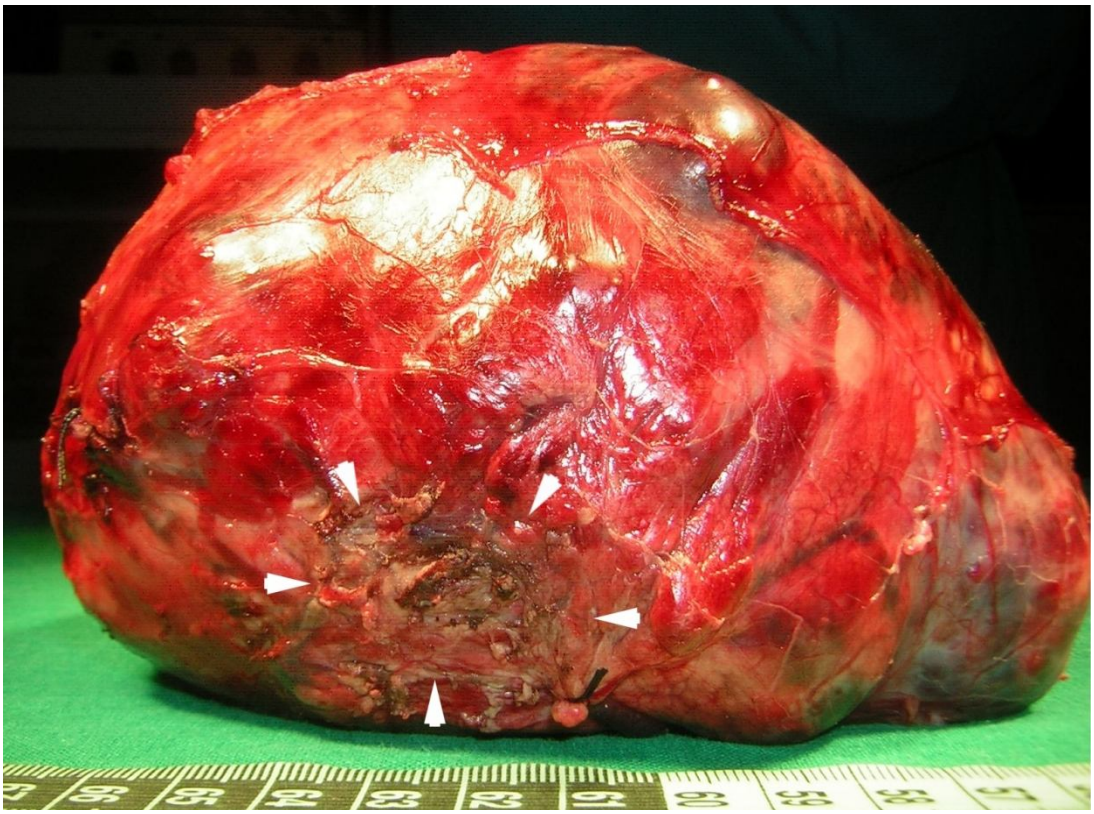

Figure 2: The well encapsulated tumor excised en-bloc. Note the wide surface of origin from the pancreas (small arrows) and the normal pancreatic tissue at the resection margin

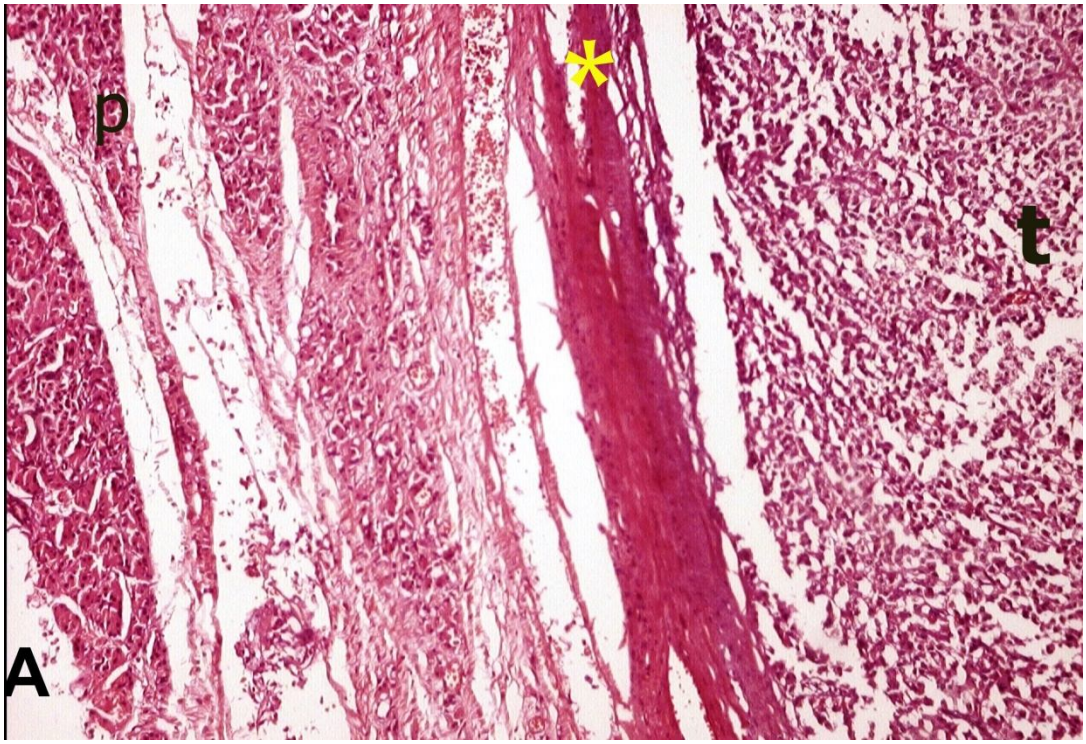

Figure 3 A: Microphotograph showing a capsulated neoplasm ([*]: capsule, t: tumor) with a portion of compressed pancreas (p).

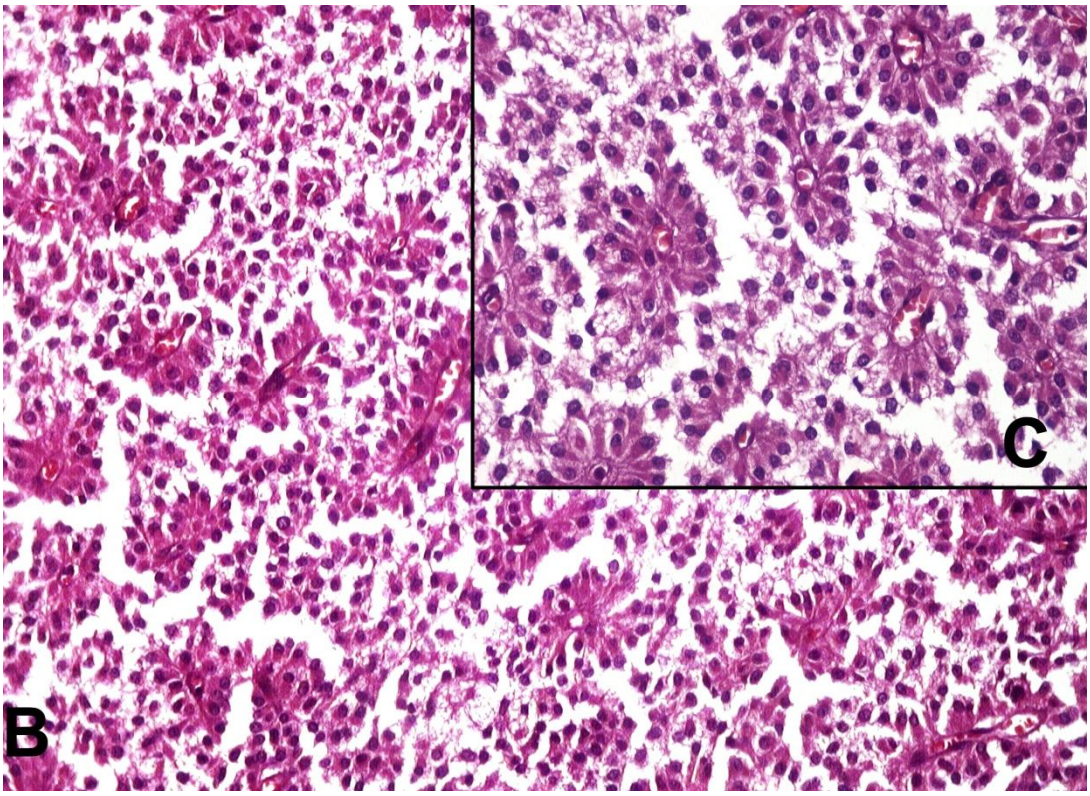

Figure 3 B and C (inset): Microphotograph showing pseudopapillary structures with tumor cells clinging to the delicate vessels and degenerated hypocellular areas away from the vasculature

DISCUSSION

Solid-pseudopapillary neoplasm of the pancreas (SPPNP) is uncommon primary pancreatic neoplasms accounting for 2-5% of pancreatic tumours. A progressive increase in incidence from 0.17-2.7% of exocrine pancreatic tumors to 6% is quoted [1]. In recent pediatric series, 60-65% of pancreatic neoplasms are SPPNP [2-4]. Initially described by Frantz and Hamoudi, it is thought to originate from the multipotent cells of the ductular epithelium or totipotential stem cell with endocrine/exocrine/dual differentiation [5-7]. The neoplasm is common in females (83-98%), especially adolescent girls and young women (mean age: 22 years). Most present with nonspecific symptoms (vague abdominal pain/discomfort, dyspepsia) making the diagnosis difficult. The abdominal mass is slow growing. Some manifest after trivial trauma or are incidentally detected at examination or imaging [1, 8]. Dramatic presentations like acute/recurrent pancreatitis and rupture/hemoperitoneum are rare [9].

At Contrast Enhanced Computerized Tomography (CECT), the tumor is usually large, well circumscribed and heterogeneous. It shows patchy capsular and solid area enhancement, the latter is typically hypoattenuating [10]. Satellite calcification is occasional. A detailed multidetector row computerized tomography (MDCT) accurately delineates vascular/capsular invasion and contiguous spread. In tumors > 6 cm, these are significantly increased [11]. Interestingly, this giant tumor had none. Besides typical features on T1-images, MRI demonstrates capsular integrity and tumoral hemorrhage better [10].

Grossly, SPPNP may arise anywhere in the pancreas. They are frequently exophytic and the site of origin may be obscure [8]. The mass may also replace the entire pancreas. SPPNP are usually circumscribed but may be partially encapsulated. Adult series cite an average diameter of 9–10 cm and pediatric series quote a median tumor diameter of 5 cm (3.5-12 cm) [4]. The larger neoplasms show patchy to complete degenerative cystic changes. The solid areas are friable and hemorrhagic [8]. This tumor had ‘giant’ dimensions; yet was predominantly solid. Sometimes, the mass is radiologically demarcated but microscopically invasive [10]. Although intraoperative evaluation for diagnosis and negative pancreatic resection margins are optimal,

gross total excision and subsequent histological confirmation are mandatory [1].

The microscopic appearance is characteristic with solid sheets of uniform polygonal cells separated into nests by abundant capillary sized vessels. There is no true lumen formation. The cells are relatively discohesive; in areas away from the vessels, they drop away leaving a ragged cuff of neoplastic cells clinging to the blood vessels (pseudopapillae). The cytoplasm is usually eosinophilic and the nuclei uniformly round to oval. The nuclei may be oriented away from the vessels, resulting in a zone of cytoplasm separating the capillaries from the nuclei. Mitoses are very rare. Despite the well-circumscribed gross appearance, the neoplastic cells often invade the contiguous pancreatic parenchyma [12, 13]. This tumor did not show any such invasion.

The differential diagnosis includes other solid, cellular pancreatic neoplasms (Endocrine neoplasms, acinar cell carcinoma, pancreatoblastoma). True luminal spaces suggest one of these rather than SPPNP. In difficult cases, staining for trypsin, chymotrypsin and chromogranin will usually help to confirm one of the alternative diagnoses; if these markers are negative, positive staining for vimentin, CD10, CD56, and alpha-1-antitrypsin confirms the diagnosis [12].

SPPNP is extremely indolent. Metastases may be seen in 10-15% with common sites being the liver, mesentery, omentum and peritoneum [8, 13] Lymph node metastases are uncommon [4, 7]. The tumor can also invade the adjoining vessels. There is no consensus on the diagnostic criteria for malignancy; clinical factors (age, sex, and symptoms), tumor size and location seem unrelated [10]. Yet, some consider tumor metastasis at operation, invasion of adjacent organ, large tumor size, young patient age, tumor rupture, and inadequate resection to increase the risk of recurrence [14].

Complete surgical extirpation with clear margins is the mainstay of treatment. A distal / subtotal pancreatectomy with/ without splenectomy usually suffices in the majority. Rarely a small (<3 cm), fully encapsulated tumor may be amenable to local resection. Other procedures that have been performed include pancreaticoduodenectomy and resection of isolated metastases [1]. Complete surgical resection results in a cure in the majority. The overall 5 year survival is 95% [15].

In conclusion, it is essential to be aware of this distinctive and rare neoplasm with a low grade malignant potential. A high index of clinical suspicion is necessary to suspect and diagnose SPPNP, particularly in a young female presenting with nonspecific symptoms and an indolent pancreatic mass. The features on imaging suggest the diagnosis and histopathology is confirmative. Complete surgical excision is the standard of care and should be attempted even with localized resectable metastases. 'Giant' predominantly solid SPPNP, like the currently described case, can be bereft of metastases/capsular or vascular invasion, and be associated with the usual favorable prognosis.

CONFLICTS OF INTEREST

Authors declare that there are no conflicts of interest.

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