Solid pseudopapillary neoplasm of the pancreas in Sub-Saharan Africa: A case report and review of literature

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ABSTRACT

Solid pseudopapillary neoplasm of the pancreas is a rare indolent malignant tumor mostly found in young women in the second and third decades of life. Only about 3000 cases have been reported in English literature, with none in the West African region. Here, we report to the best of our knowledge, the first histologically confirmed case of solid pseudopapillary neoplasm of the pancreas in West Africa. A 29-year-old woman with a slow-growing left upper abdominal mass of 10 months. Abdominal ultrasound and computed tomography scans revealed a cystic-solid mass arising from the tail of the pancreas. She underwent surgical excision of the mass, with post-operative histopathology findings in keeping with solid pseudopapillary neoplasm of the pancreas. This case report highlights the importance of improved abdominal imaging, histopathology and the need for increased awareness as a differential diagnosis of an intra-abdominal tumor, especially in young women.

Key words: *Franz tumor, Hamoudi Tumor, Pancreatic Cystic tumor, Solid pseudopapillary Neoplasm, West Africa*

Solid and cystic pseudopapillary neoplasms of the pancreas are very uncommon tumors making up just about 2% of all exocrine pancreatic tumors [1]. These tumors were initially called Frantz or Hamoudi tumors, having been earlier described in 1959 among several synonyms (including papillary epithelial neoplasm of the pancreas, solid, and papillary tumors of the pancreas), until classification by the WHO in 1996 as solid pseudopapillary neoplasms [1]. It is considered an indolent malignant neoplasm, mostly found in women, usually in the second and third decades of life [1,2].

About 3000 cases have been reported worldwide [2], mostly in small series or case reports [3] with none reported from the West African region. Recognition and diagnosis are increasing due to the increasing interest [2], improved imaging with more efficient diagnostic armamentarium [1] and also the tumor's distinct histologic features [3]. Most series have reported overall favorable long-term outcomes following complete surgical resection, with just a few cases of recurrence recorded [1-3]. We report probably the first histologically confirmed case of solid pseudopapillary neoplasm of the pancreas in the West African region.

CASE REPORT

A 29-year-old housewife presented on referral to our surgical outpatient clinic with a history of the left upper abdominal swelling which has progressively increased in size over 10 months with an abdominal ultrasound finding of a thick-walled solid mass with internal echoes in the lesser sac. There was no associated pain, vomiting, or early satiety; and no history of gastrointestinal bleeding, jaundice, weight loss, or change in bowel habits.

On examination, the patient was well-preserved, afebrile, anicteric, acyanosed, and no generalized lymphadenopathy were present. The patient's blood pressure was 120/80 mmHg, pulse 90b/m and respiratory rate of 15 c/m. Abdominal examination revealed a 20 cm × 12 cm left upper abdominal mass which was firm, non-tender, and non-ulcerated with a well-defined edge.

Abdominal computed tomography ([CT] scan) showed a poorly enhancing hypodense cystic mass with debris arising from the tail and body of the pancreas. Other examination, as well as upper gastrointestinal endoscopy and routine hematological investigations, revealed normal findings: Hemoglobin 12 g/dl (10–14 g/dl), white blood cells 7 × 10^9/L (4–11 × 10^9/L), and platelets 200 × 10^9/L (150–400 × 10^9/L).

The patient underwent laparotomy with intra-operative findings of a huge cystic tumor with mural masses attached to the pancreatic tail in the lesser sac, measuring 12 cm × 12 cm × 11 cm. A complete excision of the tumor with part of the pancreatic tail was achieved (Fig. 1). No gross metastatic disease noted in the liver or peritoneum. The excised mass was received at the histopathology department of our institution, where sections were made, observing that it had a cystic and solid-to-friable cut surface.

Examination of the hematoxylin and eosin stained sections of paraffin-embedded tissues revealed a circumscribed cellular neoplasm composed of medium-sized to large cells with fairly uniform dark nuclei and variable amounts of predominantly eosinophilic vacuolated to clear cytoplasm; cells were disposed of in sheets, nests, and pseudopapillae with a variable amount of myxoid to hyalinized stroma especially perivascular. Furthermore, there were foam cells and cholesterol clefts mixed with foreign body-type giant cells and hyaline globules. Normal pancreatic lobules were noted in the pericapsular zones. These findings were consistent with a solid-cystic pseudopapillary neoplasm (Fig. 2 and 3).

The patient developed a seropurulent discharge from the edge of laparotomy wound on the sixth post-operative day, with an associated persistent fever of 38.8°C, nausea, vomiting, and leukocytosis of 14.24 × 10^9/L. The patient was successfully managed with appropriate wound care and antibiotics (intravenous ceftriaxone 1 g 12 hourly and intravenous metronidazole 500 mg 8 hourly all for 7 days). She was discharged on the twentieth post-operative day and has been symptom-free up to 2 months on follow-up.

DISCUSSION

Cystic neoplasms account for a small portion of all pancreatic tumors, of which solid pseudopapillary tumors (1–2%) are a subset; with serous cystadenomas (35%), mucinous cystadenomas (15%), and intraductal papillary mucinous neoplasms (10%) accounting for other cystic neoplasms [3]. Their origin is still being investigated [4], but they are considered slow-growing tumors and found mostly in the body and tail of the pancreas [1,2].

The neoplasm has a higher predilection to the female sex, with Law *et al*. reported a mean age of presentation of about 28 years and mean tumor size of 8.6 cm [1]. The largest series in Africa of 21 patients, reported a mean age of 34.2 years and tumor size of 7 cm [3]. The mean age was higher than in other series due to comparatively older ages of the male patients in that series. Bouassida *et al*. in Tunisia also reported a single case in a 50-year-old man [5]. The most frequent presenting complaint is abdominal pain, nausea, and vomiting [2]. However, a significant percentage of these tumors is an incidental finding on abdominal imaging or a palpable abdominal mass [6] similar to our patient, whose tumor was also slow growing for up to 10 months.

The most common method of pre-operative diagnosis for solid pseudopapillary tumors is an abdominal CT scan, which predominantly shows well-defined heterogeneous large cystic masses with solid components. There could also be areas of vascularization and calcification [2,3,6]. This has been useful in differentiating solid pseudopapillary tumors from common differentials such as pancreatic pseudocysts, non-functioning neuroendocrine tumors, and mucinous and serous cystadenomas, and carcinomas [3]. Abdominal ultrasonography also plays an important role in diagnosis; and mostly contributing to the incidental finding of these neoplasms, as a hypoechoic mass with mixed echogenicity [3,6], they may, however, appear more solid than cystic due to the presence of hemorrhagic and necrotic tissue [7].

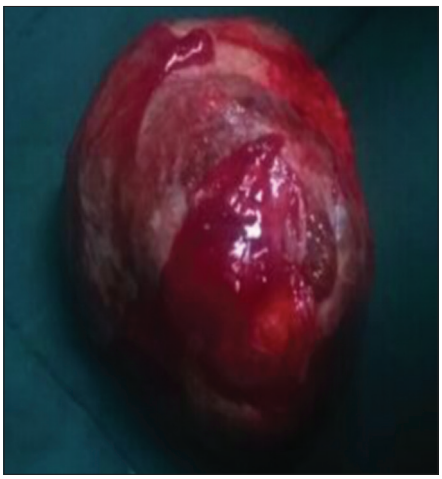

Figure 1: Completely excised tumor (original)

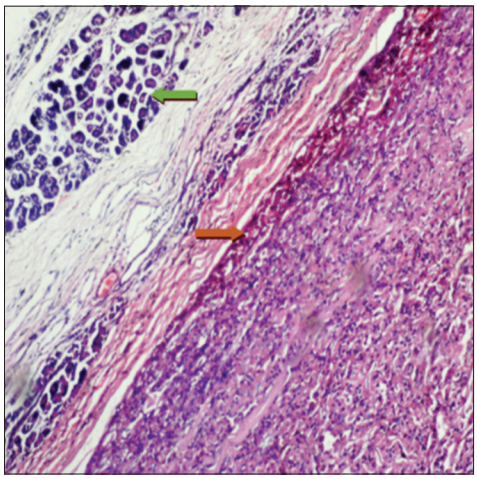

Figure 2: The section shows the circumscribed tumor (Orange arrow) and pancreatic tissue in the capsular area (green arrow) H and E × 40 (original)

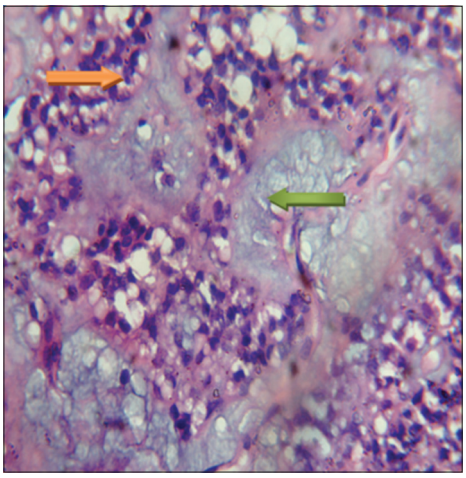

Figure 3: The section shows the cellular tumor with dark, bland nuclei (orange arrow), and myxohyaline areas (green arrow) H and E × 200 (Original)

Magnetic resonance imaging (MRI) and endoscopic ultrasound have also been used [1-4]. There are inherent limitations of CT scan

when compared to an MRI, especially while evaluating specific tissue characteristics such as hemorrhage, the presence of a capsule, or cystic degeneration. Its role also has been emphasized in evaluating small lesions where they are often unencapsulated and do not show any hemorrhage. Magnetic resonant imaging is, therefore, superior to CT scan; however, its usefulness is limited by its cost and availability. Abdominal ultrasound though cheap and readily available is the least reliable modality among the three modalities [3-7]. The usefulness of positron emission tomography in the pre-operative diagnosis of these tumors is being investigated [2].

These features were seen in the imaging studies of our patient. The internal echoes and debris seen on imaging support the gross findings; which were a cystic tissue with some solid component, which had visibly distended vessels, with the cut surface showing a cyst with 460 ml of hemorrhagic material and areas of hard necrotic material. Pancreatic solid pseudopapillary neoplasms are distinct histologically [1,3]. Recently, endoscopic ultrasound-guided fine needle aspiration cytology has been an invaluable adjunct [1,3,4,6,8] in pre- and peri-operative tissue diagnosis [6], yielding a highly cellular aspirate, demonstrating numerous pseudorosettes and pseudopapillary branching tissue fronds in multiple layers with a slender fibrovascular core [4,6,8]. However, most cases are diagnosed with full tissue histology with the characteristic appearance similar to our patient as uniform, bland ovoid cellular architecture with eosinophilic cytoplasm, pseudopapillae with hyalinized cores, clusters of cholesterol clefts, foamy cells, and multinucleated giant cells [4,9]. These tissue slides, when subjected to immunohistochemical analysis, mostly test positive for Vimentin, CD-10, CD-56, Neuron-specific enolase, alpha-1-antitrypsin, and progesterone.

Surgical resection remains the standard care and only hope of a complete cure for solid-pseudopapillary neoplasms of the pancreas [1-4,6], with most series showing tumors with grossly clear margins and overall favorable resectability. Tumor invasion into adjacent tissue does not preclude complete resection. Several procedures have been offered on a case by case basis, with the most popular being a wide local resection as offered to our patient. However, other procedures done include pylorus-preserving pancreaticoduodenectomy for tumors involving the head or uncinate process, central pancreatectomy and re-implantation of the pancreatic remnant as a distal pancreaticojejunostomy or pancreaticogastrostomy for tumors involving neck and body of the pancreas, including portal vein resection in cases of tumor invasion [1-3].

Laparoscopic resection is a relatively new approach which has been gaining ground, with Torres *et al*. reported a small series of successfully carrying out laparoscopic pancreaticoduodenectomy for solid pseudopapillary tumors of the pancreas with a similar outcome as the open approach [10]. Our patient's tumor was only attached to the pancreatic tail, which was easy to mobilize and hence amenable to complete resection with a small margin of the pancreatic tail. Hemostasis was secured, and the pancreatic tail repaired with absorbable running sutures.

Post-operative complications were reported in 5% of all cases with the most common being pancreatic fistulas; other complications include stricture at hepaticojejunostomy after pancreaticoduodenectomy, bleeding, intra-abdominal fluid collections, and recurrence following metastasis [1-3]. Our patient developed a surgical site infection with a minimal intra-abdominal fluid collection. This was successfully controlled with improved wound care and intravenous Ceftriaxone for 1 week. Furthermore, a closed intra-peritoneal passive drain inserted at laparotomy which assisted in drainage and excluded the need for re-operation or image-guided drainage. Solid pseudopapillary neoplasms of the pancreas have excellent long-term prognosis following complete surgical resection, with survival rates reported as 95% [1] with a median follow-up of 81.5 months [2,3].

CONCLUSION

Solid pseudopapillary neoplasms are rare indolent malignant tumors which are mostly found in young women worldwide, with none histologically reported in the entire West African region. There is the need for improved awareness and enthusiasm for histology and subsequent immunohistochemistry of all pancreatic tumors for increased surveillance and diagnosis, as it is readily amenable to complete surgical excision and overall good prognostic outcome.

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