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Frantz Tumor in a 15 year old female: A Case report and Review Literature

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ABSTRACT
Solid pseudo papillary neoplasm of the pancreas (SPNs) is a rare entity, known for its indolence. The pathogenesis of these tumors still controversial. Classic imaging characteristics include a large size, mixed density with solid and cystic components, encapsulation and hemorrhage. SPNs can show several criteria of malignancy such as local invasion, metastases and recurrence. Radical surgery is usually curative. We present a case of a Frantz Tumor in a 15-year-old female, with typical imaging features.

Introduction

Solid pseudo papillary neoplasm of the pancreas (SPNs) is a rare and uncommon entity, accounting for only 0.2-2.7% of all pancreatic tumors [1, 2, 3]. It is known for its low malignant potential and its indolence, both important for its good prognosis and it is more common in females in second and third decades of life [3, 4]. Since Frantz described this tumor in 1959 as a "papillary tumor of the pancreas, benign or malignant," the number of reported cases has increased. Synonyms include solid and cystic tumor, solid and papillary epithelial neoplasm, papillary-cystic neoplasm, papillary cystic epithelial neoplasm, papillary-cystic tumor, and Franz tumor. In 1996, the World Health Organization renamed this tumor solid pseudo papillary tumor for the international histologic classification of tumor of the exocrine pancreas and reclassified them as solid pseudo papillary neoplasms (SPNs) in 2010 [2, 5, 6]. After the WHO reclassification in 2010, SPN is now classified as an epithelial tumor under the borderline subcategory and is pathologically characterized as a rare cystic pancreatic neoplasm (2).

Case report

A 15-year-old girl, in a good overall health, presented with a history of abdominal pain of 6-month duration. She was initially admitted to a peripheral hospital for incoercible vomiting. Her past medical and surgical history was unremarkable. Physical examination revealed normal, body temperature and stable hemodynamics. The abdomen was distended with a 5cm well defined, non-tender, non-pulsatile mass was palpable in the left hypochondrium.

All laboratory investigations including tumor markers were within normal limits.

Abdominal ultrasound (US) showed a well-defined lesion in the left hypochondrium with variable echogenicity predominantly cystic. Doppler ultrasonography showing no apparent tumoral vascularization. MRI scan demonstrated a 56x53x58mm encapsulated tumor in the body-tail of pancreas mainly cystic but with a peripheral solid component (figure 1).T2-weighted MR images showed an hypo intense rim (figure 1). This rim was also hypo intense on T1-weighted images, consistent with the presence of a tumoral fibrous pseudo capsule (figure 2). T1-weighted images also showed some internal areas of increased signal intensity compatible with hemorrhage (figure 2). Dynamic study of the tumor revealed peripheral gradual filling of contrast material with internal a non-enhancing cystic portion (figure 3).

Based on the previous imaging findings the diagnosis of solid pseudo papillary tumor was suggested and distal pancreatectomy was performed.

Pathology showed a neoplasm with extensive necrosis, with preserved tissue found in the tumor periphery under the fibrous capsule. The margin was clear

(R0).On immune-histochemical staining, the tumor cells were positive for vimentin, CD56 and CD10 and negative for chromogranin-A.

The diagnosis of solid pseudo papillary tumor was confirmed. There was no lymph node involvement.

Discussion

The cellular lineage of solid pseudo papillary neoplasm of the pancreas is unknown; general theories of histogenesis can be divided into three main groups: pancreatic duct cell origin, acinar cell origin, and primitive cell origin. Moreover, some assume that SPNs may originate from genital ridge related cells that are incorporated into the pancreas during organogenesis on the basis of some similarities between SPN and ovarian surface cells and the proximity between genital ridges and the pancreas anlage during early embryogenesis. Others have speculated that sex hormones may play a role in the pathogenesis or growth of SPNs because the tumors have a tendency to affect young women and the growth of an SPN in the pancreas seems to be enhanced by pregnancy. Patients with malignant transformation of SPN are often older at presentation and have a male predilection [2, 3, 7, 8].

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SPN has distinctive pathologic features. The smaller tumors are organized in solid sheets and nests of cells. However, large tumors contain a mixture of solid, cystic, and pseudo papillary patterns. Solid areas are contain of necrosis, foamy macrophages, cholesterol granulomas, and calcifications and pseudo papillae are supported by hyalinized fibro vascular stalks [2].

The most common localization of SPN is the tail of the pancreas, followed by the head and the body. Unusual presentations include multicentric tumors in the pancreas and extra pancreatic sites, such as the mesocolon, retroperitoneum, omentum, liver and duodenum, possibly representing synchronous tumor spread [9, 10].

The clinical presentation of SPN is nonspecific. Most of patients present with non-specific symptoms including abdominal discomfort, mild abdominal pain or palpable abdominal mass. Due to its slow growth, SPN often remains asymptomatic, until the tumor has enlarged considerably. Accordingly, many are detected incidentally on diagnostic imaging for unrelated diseases or after a blunt abdominal trauma [10]. Jaundice or obstruction of the main pancreatic ductare uncommon, and clinical biochemistry is uncharacteristic and there are no useful tumor markers in plasma [11, 7].

Immunohistochemically, SPNs are typically positive for vimentin, neuron-specific enolase (NSE), α-1-antitrypsin, CD10, CD56, progesterone receptors, and β-catenin and negative for chromogranin, epithelial membrane antigen, and cytokeratin (4). Most SPNs have an indolent clinical course, but some of these have malignant potentials, include vascular or perineural invasion, Ki-67 positive, significant cellular pleomorphism, nuclear atypia, increased mitotic activity and increased β-catenin [2, 9, 7, 12].

According to the WHO classification scheme, SPNs with clear criteria for malignancy (vascular and nerve sheath invasion or lymph node and liver metastasis) are designated as solid pseudo papillary carcinomas (SPCs). There have been attempts to identify imaging characteristics that aid in differentiating benign SPN from malignant SPC. Recent studies demonstrate that hepatic or peritoneal involvement, main pancreatic duct obstruction, infiltration of pancreatic parenchyma, vascular encasement, focal discontinuity of the capsule, large tumor size (> 6.0 cm) and pancreatic tail location may suggest malignancy of SPN [2, 13, 14].

The imaging features of solid pseudo papillary tumor reflect the pathologic findings of cystic and solid components, intratumoral hemorrhage, a fibrous capsule, and, less commonly, calcifications [15].

Solid pseudo papillary neoplasm can be detected by ultrasonography, computed tomography, magnetic resonance imaging, and positron emission tomography. Plain radiography (X-ray) does not have value but to show possible calcifications in the neoplasm. Computed tomography scan of the neoplasm demonstrates solid and cystic features with regions of hemorrhage and/or cystic degeneration. Calcifications and enhancing solid areas may be present at the periphery of the mass [1].

On ultrasound, solid pseudo papillary tumor presents as a well-circumscribed heterogeneous mass surrounded by a pseudo capsule of compressed pancreatic tissues and reactive fibrosis, sometimes with central cystic areas of necrosis. The capsule may be visualized as an echogenic or, less commonly, hypoechoic [15]. An unenhanced CT demonstrates a well-encapsulated lesion with varying solid and cystic components owing to hemorrhagic degeneration. Following contrast material administration, enhancing solid areas are typically noted peripherally, whereas cystic spaces are usually more centrally located [16].

MRI should be considered the best imaging technique due to the absence of radiation and its improved capacity for visualizing tumor components [15]. It is superior to CT in distinguishing certain tissue characteristics, such as hemorrhage, cystic degeneration or the presence of a capsule and may suggest correct diagnosis [10]. T1-weighted images show a surrounding hypo intense fibrous pseudo capsule and high signal intensity areas corresponding to internal hemorrhage, distinguishing features of solid pseudo papillary tumor. Similar dark rim is also seen on T2-weighted images corresponding to the pseudo capsule. The solid portions of the tumor are usually iso to hypo-intense to pancreas on T1-weighted images and slightly heterogeneously hyper intense on T2-weighted images [15, 11, 7].The most common enhancement pattern of solid pseudo papillary tumor consists of early, peripheral enhancement of the tumoral pseudo capsule during the arterial phase, compared with the normal pancreas and the lesion itself. In some cases there is a progressive but heterogeneous fill-in of the lesion during the portal venous and equilibrium phases, showing less enhancement that the adjacent normal pancreas consistent with a hypo vascular tumor [15, 17].

Pseudo papillary solid tumor of the pancreas may have typical and atypical appearance. Typical as it was described above however misdiagnosing of SPT is common [18].

The variety of imaging techniques available may help differentiate SPT from other neoplasms [16]. The differential diagnosis of SPN is wide and includes in particular solid and cystic lesions such as serous microcystic adenoma, cystadenocarcinoma, mucinous cystic neoplasms, cystic neuroendocrine tumors, cystic acinar cell carcinoma, teratoma, pancreatoblastoma as well as a variety of congenital and acquired dysontogenetic, post-inflammatory and infectious cysts. However, the typical constellation of a pancreas-associated solid and cystic upper abdominal mass with or without calcifications in a young woman should always alert to the possibility of SPN [10].

The only curative treatment is radical surgery with free resection margins, and adjuvant oncologic therapy has no impact on survival. Due to the localized growth of STN, this is possible in most cases. Local tumor infiltration or metastatic disease is not a contraindication for operation since radical resection including all metastatic tissue may result in long-term survival and cure. The overall 5-year survival rate is more than 95.0% in large-scale reviews and the recurrence rate up to 6.6% [11].

Primary metastatic disease as well as recurrence have been treated according to different oncologic protocols including 5-FU, S-1, gemcitabine, sunitinib, and trans arterial embolization of liver metastases. However, the results are casuistic, and the general opinion is that oncologic treatment has a limited effect on SPN which calls for aggressive surgical resection including metastases and in case of recurrence [11, 19].

Conclusion

SPN is a rare tumor mostly seen in young women with unknown origin. SPN of the pancreas have an excellent prognosis. Classic imaging characteristics include large size, mixed solid and cystic nature, encapsulated appearance and

presence of hemorrhage. Surgery is curative in the vast majority of cases.

Conflict of interest

The authors do not declare any conflict of interest.

Author's contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

Figures

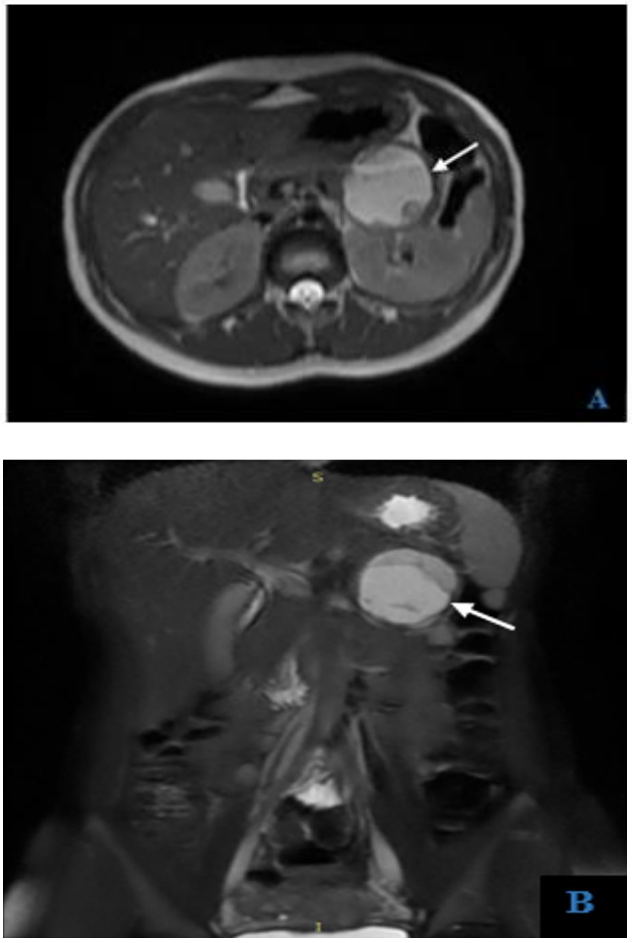

Figure 1. Axial (A) and coronal (B) single shot fast spin echo T2-weighted MR image showed an encapsulated tumor of a 56x53x58mm in the body-tail of pancreas mainly cystic but with a peripheral solid component. The lesion is limited by a continuous capsule (white arrow).

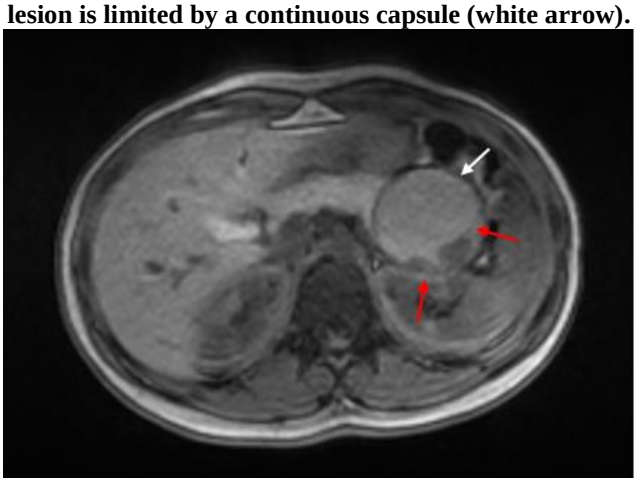

Figure 2. Axial unenhanced fat-suppressed T1- weighted MR image the fibrous pseudo capsule is also hypo intense (white arrow) and there is an internal peripheral high signal intensity rim (red arrow) a finding consistent with hemorrhage.

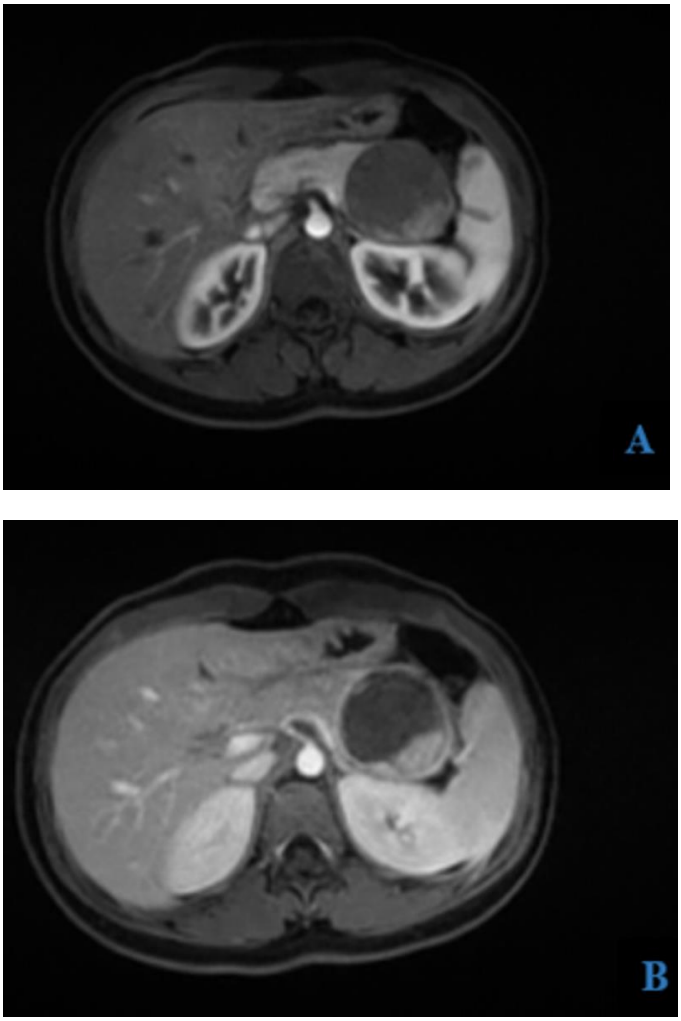

Figure 3. On the dynamic MRI, the mass shows heterogeneous peripheral enhancement in the arterial phase (A), and heterogeneous peripheral gradual enhancement with internal non-enhancing cystic portion in the delayed.

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