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Solid Pseudo-Papillary Neoplasm of Pancreas with Venous Invasion in A 9 Year Old Girl: A Case Report and Literature Review

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ABSTRACT

Solid Pseudo-papillary neoplasm of the pancreas (SPN) is a rare entity. It represents 0.2-2.7% of all pancreatic cancers. Predominantly occurs in young females in the second to third decades of life. The etiology of SPN involves mutations in the gene that encodes beta-catenin. SPNs are typically indolent tumors, usually confined to the pancreas. We report a case of SPN in a 9-year-old female presented with intermittent abdominal pain for four months. Imaging studies demonstrated a 2.8 cm mass in the tail of the pancreas. The patient underwent a distal pancreatectomy. Pathological evaluation was diagnostic for SPN in the tail of the pancreas. Our case is distinct because of the young age of the patient, peripancreatic soft tissue, perineural, and lymphovascular invasion. The tumor cells exhibited cytoplasmic and nuclear immunoreactivity for beta-catenin and progesterone receptors.

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Introduction

Solid Pseudo-papillary neoplasm of the pancreas (SPN), is a rare, benign, low grade malignant epithelial tumor of the exocrine pancreas [1]. This tumor was first described by Frantz et al., in 1959 and the World Health Organization (WHO) renamed it in1996 as SPN [2]. The mainstay of treatment of SPN is surgical resection. An aggressive surgical approach towards local and distant metastasis is justified due to excellent long-term prognosis and a high 5-year survival rate (95-100%). Pancreatic cancer ranks as the fourth most common cancer type in the western world and is responsible for 6% of all cancer-related deaths. Ductal adenocarcinoma is the predominant pathological subtype, which accounts for 90% of all pancreatic tumors. Here, we present a case of SPN in a 9-years old female who had a 2.8 cm mass in the tail of the pancreas.

Clinical history

A 9-year-old obese white girl had a 4-month history of intermittent abdominal pain. Family history is unknown. She presented to the hospital with epigastric pain for six weeks, not associated with fever, nausea, or vomiting. Laboratory results showed raised liver function tests (LFTs) and high amylase and lipase levels. CT scan of the abdomen and pelvis demonstrated a 2.8 cm mass of the tail of the pancreas. Distal pancreatectomy with the removal of body and tail was performed. One month after surgery, the patient presented with abdominal pain and fever. CT scan of the abdomen revealed showed a small fluid collection, encircling her Jackson-Pratt drainage (JP). Her symptoms resolved after removal of her JP and the patient was discharged and lost to follow up since then.

Results

The specimen consisted of an 8.5 x 4.0 x 2.5 cm pancreatic tissue, including body and tail. Cross-section showed a partially encapsulated 2 X1.5 X 1.2 cm mass with extensive necrosis. The mass was grossly confined to the distal body and tail of the pancreas. Margins were negative for tumor, and distance from the closest margin was 1.5 cm. Microscopically, the tumor extended to the peripancreatic soft tissue. The tumor was very cellular (figure 1a) and arranged in solid sheets and pseudo-papillae (figure 1b). Individual tumor cells were uniform, small, and polygonal with acidophilic cytoplasm and bland ovoid nuclei and few mitotic figures. Venous invasion was identified, highlighted by desmin immunostain (figure 2a and 2b). Perineural invasion was also present. By immunohistochemistry, the tumor cells were positive for beta-catenin, both nuclear and cytoplasmic (figure 1c) as well as for progesterone receptor (PR) (figure 1d); few cells were positive for synaptophysin, but the tumor was negative for chromogranin.

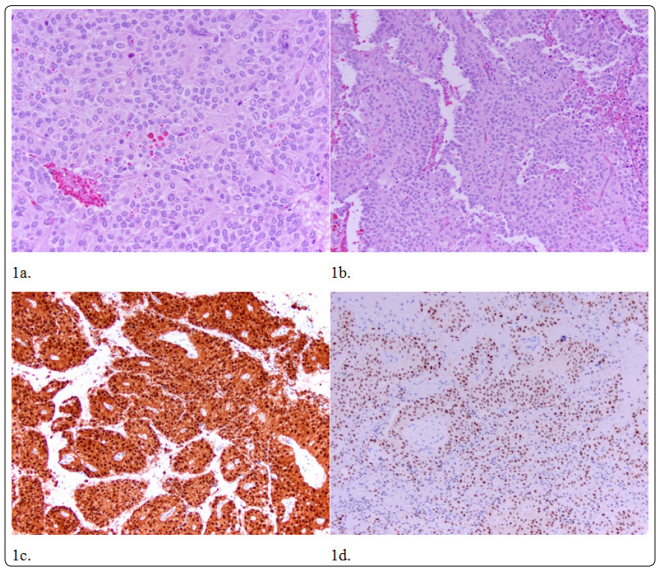

Figure 1: 1a & 1b shows the arrangement of solid sheets and pseudo papillae.
1c. Strong nuclear and cytoplasmic positivity of beta catenin immunostain.
1d. Positive progesterone receptor immunostain

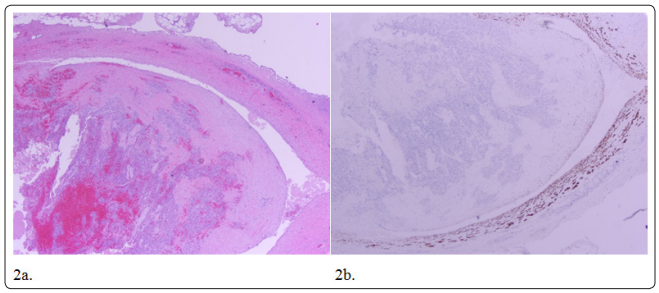

Figure 2: 2a.Tumor invading vein and 2b. Desmin immunostain.

Discussion

Pancreatic neoplasms have been divided into two different categories, solid and cystic lesions [1]. Pancreatic cystic neoplasms are further subdivided according to their histological characteristics by the World Health Organization (WHO) in serous cystic tumor, mucinous cystic neoplasm, intraductal papillary mucinous neoplasm, and solid pseudo-papillary neoplasm.

SPN represents less than 10% of cystic neoplasms of the pancreas. According to the WHO classification, SPNs with clear criteria of malignancy (i.e., vascular and nerve sheath invasion, lymph node, and liver metastasis) are designated as solid pseudo-papillary carcinoma [1,2]. The name of this entity was proposed by Virginia Frantz in 1959. In 1970, Hamoudi et al. described the ultrastructure of the tumor [3]. Until its inclusion in the WHO classification of pancreatic tumors as solid pseudo-papillary tumor of the pancreas, this entity has been described by different names in literature such as "papillary epithelial neoplasm of pancreas", "solid and cystic tumor of the pancreas", "adenocarcinoma of the pancreas of the childhood" and "solid and papillary epithelial neoplasm". In the current WHO classification, SPN is defined as a low-grade malignant neoplasm of the exocrine pancreas [2].

It typically occurs in young females in their second to the third decade with a female to male ratio of 10:1. SPNs are almost always benign, although it has been reported that up to 20% of the cases can demonstrate a malignant potential. About 20-25% of the cases are seen in pediatric patients [1]. Very few cases in men and the elderly have also been reported. A retrospective study from 1992 to 2002 of 14 cases with SPN's of pancreas showed that all of them were female in the age group of 13-45 years [4]. The unique features of our case report are young age (9-year-old) with peripancreatic soft tissue, perineural, and venous invasion. Venous invasion, in this case, raises the possibility of distant metastasis.

SPN has been postulated to arise from primitive pancreatic cells (e.g., acinar cells, ductal epithelium, or endocrine cells) or from cell lines of the female genital bud. Beta-catenin mutations, alterations of the WNT pathway, and E-cadherin disorganization have been implicated in the development of SPN [5]. Cyclin D1, a downstream target of beta-catenin, is overexpressed in most cases [6,7]. APC/beta-catenin pathway and cyclin-D1 alterations are observed in over 90% of the cases [8,9]. The Ki-67 index has been suggested as a potential indicator of malignant potential [6,10]. The common expression of progesterone receptor and strong predilection for females suggests that it might be a hormone-dependent tumor [11].

The negativity for chromogranin A of the neoplastic cells of SPNs helps in reaching a diagnosis as well as is helpful in differentiating from other neoplasms such as neuroendocrine tumors [12].

Histologically, SPNs are usually well-circumscribed and encapsulated masses with degenerative cystic cavities and hemorrhages [13]. The tumor contains a mixture of solid, cystic, and pseudo-papillary patterns in various proportion [14]. The key histological hallmarks are solid and pseudo-papillary proliferation of monomorphous cells without increased mitosis or cytologic atypia. The diagnosis can be confirmed by immunohistochemical analysis [12].

Most patients present with non-specific symptoms, including abdominal discomfort, mild abdominal pain, or a palpable abdominal mass [15]. Due to its slow growth, SPN often remains asymptomatic until the tumor has enlarged considerably. Many are detected incidentally on diagnostic imaging for unrelated diseases or after abdominal trauma [16]. The signs and symptoms of SPNs are related to mass effect and consist mainly of abdominal pain and abdominal discomfort [15]. The most common localization of SPN is the tail of the pancreas, followed by head and body [1,17].

Routine laboratory parameters and tumor markers are of little help. Ultrasound and CT/MRI scans typically show a large, well-circumscribed, heterogeneous mass with varying solid and cystic components, generally demarcated by a peripheral capsule and occasional calcification. CT scan is the radiological diagnostic technique of choice. Prognosis is excellent, and surgical resection can result in complete cure [18,19]. SPNs are typically indolent tumors confined to the pancreas [17]. However, aggressive features such as ductal dilatation, extracapsular invasion into adjacent structures, including perineural and perivascular invasion, and nodal and distant metastasis have been reported. Though it shows low malignant potential, 10-15% of cases show aggressive behavior with metastatic involvement of the liver.

Conclusion

The possibility of SPN should be considered in the differential diagnosis of a pancreatic mass in pediatric patients. We recognized vascular invasion in this case which is a sign that tumor might metastasized. Malignant behavior is observed in only 10-15% of the cases and although not reported in pediatric patients before, it should be considered when evaluating the specimen.

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