

Management Solid Pseudopapillary Neoplasm of the Pancreas

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Abstract

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare tumor with low malignant potential. SPN accounts for approximately 1% to 2% of all pancreatic neoplasms. Most of the patients are young women. It is generally located in the body and tail of the pancreas. Most of the cases are asymptomatic and are usually diagnosed during incidental examinations. Neoplasms have a low malignant potential and the reported incidence of malignant transformation is approximately 15%. Metastasis or local invasion are the most important criteria for malignant transformation. Radical resection of the tumor provides long-term survival. There is no clear recommendation in the literature regarding the resection of the primary neoplasm and the management of its metastasis, if any. The aim of this study is to evaluate the diagnostic approach and treatment options of pancreatic SPN disease.

Case Presentation

56-year-old male patient presented with complaints of abdominal pain and nausea. On examination, there was mild tenderness in the epigastric region. Laboratory tests revealed increased urea and creatinine. MRI, a well-circumscribed nodular lesion enhancing late period and thought to show mild diffusion restriction, located just to the left of the midline at the level of the pancreatic corpus and reached a diameter of approximately 20 mm, PET CT was planned for the patient. On PET CT (Figure 1B, C), a nodular hypodense lesion, approximately 2 cm in size, showing increased FDG activity (SUVmax: 11.01) at the level of the pancreatic corpus was detected. Surgical decision was made for the lesion in the pancreas. Tumor markers of the patient were, Ca 19.9: 24.7 U / mL (N: 0-35 U / ml), CEA: 7.09 ng / ml (N: 0-7 ng / ml).

On exploration, the lesion in the pancreatic body was palpated and confirmed by peroperative USG (Figure 1D). It was excised with the enucleation technique (Figure 2). Pathology result was reported as 2.2x2.7x2 cm solid pseudopapillary neoplasia (Figure 3). The patient was discharged on the 4th postoperative day without any complications.

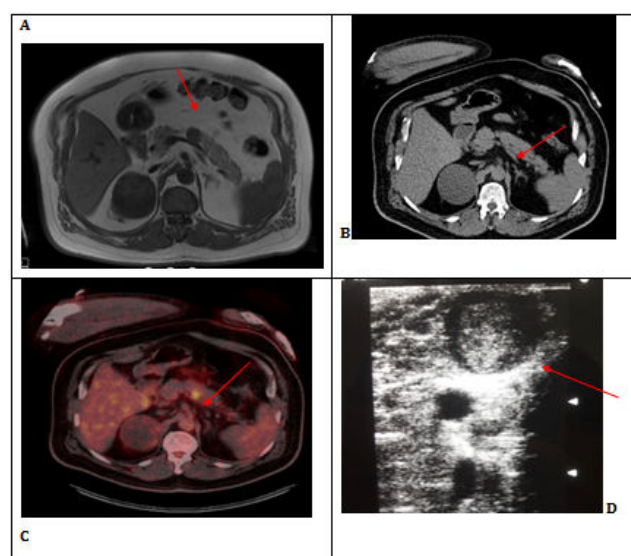


Figure 1: A:Dynamic MR: T1W hypointense, T2W heterogeneous-slightly hyperintense nodular lesion.

B: CT imaging

C: PETCT, increased FDG activity (SUVmax: 11.01).

D: well-circumscribed 35x28 mm lesion was detected on peroperative USG.



Figure 2: Macroscopic view of the lesion.

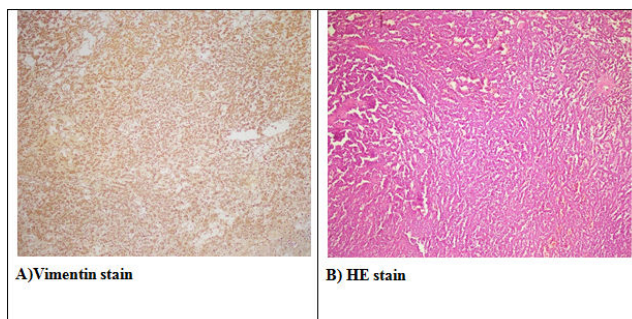


Figure 3: Microscopic view of the lesion: A) Vimentin staining, B) The appearance of pseudopapilla in HE staining. Also synaptophysin, progesterone receptor, β -catenin, CD 56 positive, The KI 67 index was found to be less than 1%.

Discussion

Solid pseudopapillary neoplasm (SPN) of the pancreas was first reported by Frantz in 1959. Since the first definition of SPN in the literature, according to its varying components, "papillary cystic neoplasm", "solid and papillary epithelial neoplasm", "papillary and solid neoplasm", "papillary and cystic tumor", "solid and cystic papillary epithelial neoplasm" and Frantz. Various names have been used such as tumor. The world health organization recently named this tumor as solid pseudopapillary tumor of the pancreas in 1996 [1, 2].

Solid pseudopapillary neoplasms comprise approximately 1% to 2% of all pancreatic neoplasms and 6% of exocrine pancreatic tumors [3]. 90% of the patients are young women in the 2nd and

3rd decades [4]. The origin of solid pseudopapillary tumors is uncertain. SPN can originate from multidirectional primordial cells, ductal cells, acinar cells, endocrine cells or extrapancreatic cells [5].

Microscopically, the tumor consists of solid areas, pseudopapillary structures and cystic cavities. Mitosis is rarely seen in tumor cells. Tumor markers; β -catenin, cyclin D1 and D3 are vimentin, antitrypsin, NSE and progesterone receptors. These immunohistochemical markers do not correlate with malignancy potential. In our patient; The β -catenin, vimentin, antitrypsin, and progesterone receptors were found to be positive and the KI 67 index was less than 1%.

The differential diagnosis includes mucinous cystic neoplasm, nonfunctional islet cell tumor, microcystic adenoma, pancreatic adenocarcinoma, pancreaticoblastoma, cystic degeneration of solid tumor and calcified hemorrhagic pseudocyst [5]. Neoplasms have a low malignant potential and the reported incidence of malignant transformation is around 15% [6]. The most accepted criteria for malignant transformation are metastasis or local invasion. Tumor diameter over 5 cm is also significant for malignant potential. In our patient, the tumor was 2 cm in diameter and there was no local invasion or metastasis.

Radical resection of malignant SPN provides long-term survival. Post-surgery recurrence rate is very low and it has been reported as 3-7% [7]. Local invasion, metastasis and recurrence are not contraindications for surgery.

Contrast-enhanced CT scanning is the most commonly used imaging method in the diagnosis of these neoplasms. They are seen as large-sized solid masses with a typical appearance on CT, which can be clearly distinguished from the adjacent pancreas, with peripheral solid, central necrosis and areas of cystic degeneration in varying degrees due to intra-tumor bleeding [8, 9]. In magnetic resonance imaging (MRI), the hemorrhagic component of the lesions can be visualized much better. The lesion is larger than 5 cm, the cyst wall is thicker than 3 mm, the detection of intramural nodules, solid-cystic bleeding and necrosis areas are the findings that increase the malignancy potential [10]. In our patient, the size of the lesion was 2x2 cm in size, and there were no findings that increased the potential for other malignancies.

The reported SUV max values of primary SPN ranged from 2.6 to 16.2. Nakagohri et al. reported that four of 5 SPN tumors with intense FDG uptake had signs of malignant potential, such as microscopic or venous tumor invasion [11, 12]. In our patient, PET CT was reported as a nodular hypodense lesion with a size of 2 cm showing increased FDG activity (SUV max: 11.01).

In a study by Castro et al. a definite diagnosis of SPN was reached with percutaneous FNAB in two out of three patients. In the study of Bardales et al. this rate is one sixth [13]. Therefore, fine needle aspiration cytology may be helpful in making the diagnosis, but is not usually necessary. 5-year survival is reported to be between 95-98%.

Complete surgical excision is curative in more than 95% of patients with SPN limited to the pancreas. In locally advanced cases, resection of the involved organs with a 1 cm intact

surgical margin is sufficient. Some studies suggest more limited surgery in the form of enucleation rather than radical surgery in small lesions. For larger lesions, the type of surgical resection depends on their location. Lesions on the head of the pancreas are treated with a pancreatoduodenectomy (Whipple procedure), while those on the trunk and tail are best treated with a distal pancreatectomy.

The incidence of lymph node metastasis is extremely rare. In the study by Mao et al in which 292 SPNs were reviewed, 43 of the cases (15%) were reported as malignant, and lymph nodes were involved in only five of them (2%) [14]. A formal lymph node dissection was not planned in our patient.

In our patient, enucleation technique was used to avoid complications of radical surgery and to prevent long-term hospitalization. In a series of 3 cases we previously operated, patients who underwent laparoscopic distal pancreatectomy, distal subtotal pancreatectomy and whipple procedure, had a hospital stay of 8, 9, 19 days, respectively. This patient was discharged on the 4th day without any complications.

Conclusion

PPPD (pylorus preserving pancreatoduodenectomy), whipple procedure and distal pancreatectomy in the tail are the most commonly used surgical methods in lesions compatible with SPN in the preoperative evaluation of the pancreas. In lesions compatible with small sized SPN in the pancreatic body in selected patients in experienced centers; corpus resection or enucleation can be performed with low complications and high surgical success rates. In this way, the number of days of hospitalization and treatment costs can be reduced.

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