

# Solid and Papillary Neoplasm Located at the Pancreatic Tail: Report of Three Cases and Review of the Literature: Case Report

## Pankreas Kuyruğunda Lokalize Solid ve Papiller Neoplazm: Üç Olgu Bildirimi ve Literatürün Gözden Geçirilmesi

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**ABSTRACT** Solid and papillary neoplasm (SPN) is a rare, low-grade malignant tumor of the pancreas which predominantly occurs in women of second and third decades. Unlike other pancreatic malignancies, the prognosis with this tumor seems to be excellent. In this study, we presented 3 cases with SPN located at the pancreatic tail. Histopathologic diagnosis was performed for all patients and all cases were followed up for 1-6 years. No metastasis or recurrence was found during follow-ups. Every attempt should be made for complete surgical removal since cure rate is high, whereas chemotherapy and radiotherapy are of no benefit in this treatment.

**Key Words:** Pancreatic neoplasms; magnetic resonance imaging; tomography, spiral computed

**ÖZET** Solid ve papiller neoplazm (SPN) pankreasın nadir görülen, düşük dereceli malin bir tümörü olup, büyük ölçüde 2. ve 3. dekada olan genç kadınlarda görülmektedir. Diğer pankreatik malignitelerin aksine bu tümörün prognozunun çok iyi olduğu görülmektedir. Çalışmamızda pankreasın kuyruğunda lokalize olan 3 SPN olgusunu sunuyoruz. Tüm hastalar için histopatolojik tanı gerçekleştirildi ve hastalar 1 ile 6 yıl takip edildi. Hastaların takibinde metastaz veya nüks ile karşılaşmadı. Tedavide tam iyileşme oranı yüksek olduğu için, tümörün cerrahi olarak tümüyle çıkarılabilmesi için her türlü çaba gösterilmelidir, bunun yanında kemoterapi veya radyoterapinin tümörün tedavisinde yararı gösterilememiştir.

**Anahtar Kelimeler:** Pankreas neoplazmları; manyetik rezonans görüntüleme; tomografi, spiral bilgisayarlı

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Solid and papillary neoplasm (SPN) of the pancreas is a rare neoplasm that occurs predominantly in young women. This distinct lesion is different from cystadenoma, cystadenocarcinoma, microcystic adenoma and islet cell tumor and was first described by Frantz in 1959.<sup>1</sup> It is a rare, low-grade malignant neoplasm and deserves special note among pancreatic malignancies as it is frequently resectable and has a good long term survival rate after complete excision. Herein, we documented three patients with this neoplasm.

Between 1998 and 2002, 3 patients with SPN of the pancreas were admitted to our surgery clinic. Imaging studies included abdominal ultrasonography (USG), computed tomography (CT) and magnetic resonance imaging (MRI). Blood samples from each patient were analyzed for blood

glucose, amilase, transaminases, alkaline phosphatase and tumor markers (CA 19-9, CEA). Two patients underwent distal pancreatectomy and splenectomy. In the remaining patient, distal pancreatectomy was performed without splenectomy. Histopathologic examinations were performed in all patients. All cases were followed up every 3 months for the 1st year and every 6 months for the following years. During the follow-up visits, abdominal USG and CT were performed for every 6 months.

## CASE REPORTS

### CASE 1

A 16-year-old girl was admitted to the surgery clinic for left-upper abdominal pain. On physical examination, the left-upper abdomen was tender to palpation and otherwise she appeared healthy. Laboratory studies were within normal ranges.

On USG, a 6 x 5.5 cm hypoechoic mass lesion was found in the tail of the pancreas. The lesion showed posterior acoustic enhancement which was suggestive of the cystic nature of the lesion. On CT, a well-defined, round, hypodense mass lesion was located in the tail of the pancreas. Following contrast administration this cystic mass showed peripheral enhancement. The lesion was extending posteriorly to the anterior pararenal region and splenic hilum without invasion. The horizontal part of the duodenum was somewhat displaced medially. No liver or other organ metastases were detected. No abnormal lymph nodes were noted. T2 weighted MRI showed hyperintense, heterogenous tumor in the tail of the pancreas (Figure 1). Open surgical exploration revealed a mass with rubber consistency and with areas of cystic appearance in the tail of the pancreas, displacing the spleen and stomach. Distal pancreatectomy without splenectomy was planned, but after resection of the tail of the pancreas, splenic ischemia was observed and therefore splenectomy was performed as well. Cut surface of the tumor showed cystic and solid components but the cystic appearance predominated in the lesion. The large cystic areas were filled with hemorrhagic and necrotic material (Figure 2). Histopathologic examination revealed minimal nuclear pleomorphism,



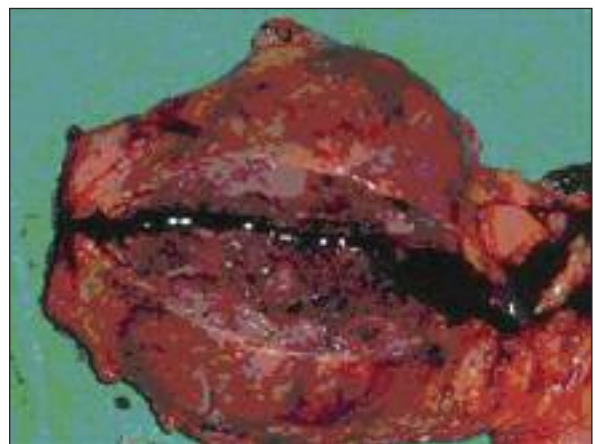
**FIGURE 1:** Fat-suppressed T2 weighted MR image of the same patient, showing heterogenous, hyperintense cystic mass in the same region.

but no mitotic activity (Figure 3). Immunostaining for vimentin and progesterone receptor were positive, Ki 67 was <1% (Table 1).

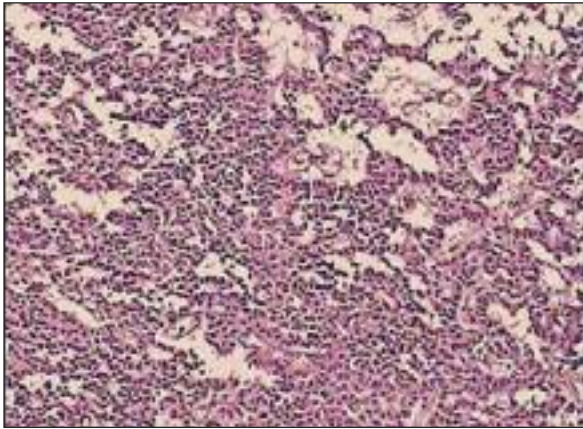
Postoperative course was uneventful. The patient was discharged on postoperative day seven. She was well during the 6 year follow-up except for some abdominal discomfort presumably due to gas distension which occurred occasionally. During this period no recurrence or distant metastasis was detected.

### CASE 2

A 73-year-old woman was admitted to the hospital with complained of abdominal distension and he-



**FIGURE 2:** The specimen of the patient. Cystic and solid areas seen on cross section of the tumor.



**FIGURE 3:** Areas of solid and pseudopapillary pattern. Fine eosinophilic granules are observed in the cytoplasm of some tumor cells. (HE, x100).

artburn. On physical examination a mobile left-upper abdominal mass was noted. Results of laboratory work-up were within normal ranges. Abdominal USG revealed a 89 x 66 mm well-demarcated, solid, hypoechoic mass located in the tail of the pancreas. CT showed a mass of 7 x 7 x 8 cm, located at the pancreatic tail which was initially thought to be a retroperitoneal mass (Figure 4). The lesion appeared had lobulated, well-defined tumor containing, hypodense areas.

During open surgical exploration a tumor with rubber consistency and with areas of cystic appearance was found in the tail of the pancreas displacing the spleen and stomach. Distal pancreatectomy without splenectomy was performed.

The cross section showed solid, papillary and cystic areas. Cystic lesions were filled with hemorrhagic or blood stained fluid. Pathological examination revealed SPN (Figure 5). Immunostaining was positive for progesterone only. Ki 67 was 1% (Table 1).

Postoperative course was uneventful and the patient was permitted to go home on postoperative day six. She has been well for 4 years. During the follow-up visits, neither recurrence nor metastasis was observed.

### CASE 3

A 68-year-old woman was admitted to the gynecology department for lower abdominal and back pain and was referred to surgery after abdominal USG examination which revealed a cystic tumor in the upper retroperitoneal region. Physical examination was normal, blood profile was within normal ranges except for blood glucose and amylase levels which were slightly higher than normal. CA 19.9 was moderately increased (58 u/mL).

Abdominal CT showed a 5 x 4.5 cm tumor located in the tail of the pancreas. Before contrast administration, the tumor was isodense to the normal pancreatic tissue. But following contrast administration it showed heterogenous enhancement with areas without contrast probably due to cystic and/or necrotic changes. The lesion extended anteriorly to the peripancreatic fatty planes but neither vascular nor contiguous organ involvement was noted. Superior mesenteric artery and vein, splenic and portal veins were intact. There was no metastasis to distant parenchymal organs. No abnormal lymph nodes were observed throughout the abdomen. Fat-suppressed T2 weighted MRI showed heterogenous hyperintense lesion in the tail of the pancreas.

On surgical exploration, a mass was seen in the tail of the pancreas adjacent to the spleen and stomach. Distal pancreatectomy with splenectomy was carried out in a usual manner (Figure 6). Immunostaining for vimentin, cytokeratin, and progesterone receptor were positive (Table 1).

**TABLE 1:** Immunohistochemical staining of the patients.

	Vi	CK	PRG	EST	NSE	SYN	CHR-A	EMA	CEA	S-100	P53	Ki67
C1	+	-	+	-	-	-	-	-	-	-	-	<1%
C2	-	-	+	-	-	-	-	-	-	-	-	1%
C3	+	+	+	-	+	+	+	-	-	-	-	1%

CK: Cytokeratin, EMA: Epithelial Membrane Antigen, EST: Estrogen, CEA: Carcinoembryogenic antigen, SYN: Synoptophysin, PRG: Progesteron receptor, CHR-A: Chromogranin-A, Vi: Vimentin.



**FIGURE 4:** Axial CT scan of the patient reveals hypodense mass located at the tail of the pancreas.

Gross examination demonstrated predominantly solid tumor with areas of hemorrhage. Postoperative course was uneventful. She was discharged on postoperative day four. Twenty months following the operation she seemed well and had no complications related to the operation or the tumor itself.

## CONCLUSION

Papillary cystic tumor is a rare exocrine neoplasm of the pancreas and has specific clinicopathological features which differ from those of adenocarcinoma, cystadenocarcinoma and insulinoma. “Frantz’s tumor”, solid and cystic neoplasm”, “papillary-cystic neoplasm” “papillary cystic carcinoma” or “papillary and solid epithelial neoplasm of the pancreas” are the different names referred to this entity.<sup>2-5</sup> In contrast to more common forms of pancreatic neoplasm, this tumor appears to have favorable overall prognosis. In reported cases, patients were predominantly young women.<sup>6,7</sup> Zhou reported that solid and papillary neoplasm was probably the most common pancreatic tumor in the Asian pediatric population; however, two of our patients were over 65 years.<sup>8</sup>

Most of those patients complain from a large, slowly growing mass in the upper abdomen but they are otherwise healthy. Abdominal pain is the 2nd most frequent reason bringing these patients to the hospital. Our 2 patients had blunt abdominal

pain but the third patient’s complaint was only heartburn.

The USG appearance of the solid and papillary epithelial neoplasm is in the form of a well-encapsulated, cystic, and solid mass, but sometimes a pure solid-looking lesion with internal septations or calcifications can also be seen.<sup>9</sup>

CT shows a tumor with areas of high and low densities. Central cystic degeneration and occasional calcified margins may be found. Chavez et al reported that CT or USG guided aspiration biopsy or frozen section was useful for its pre-resection diagnosis.

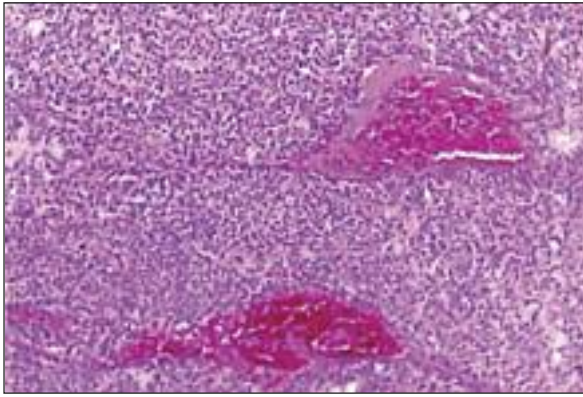
MRI in pancreatic imaging is generally preferred as problem solving method in cases that could not be solved or suspected in CT. Improvements of breath-hold and fat suppression techniques, and applicability in the decreasing peristalsis of the intestine brought wider use MRI that can provide 3D abdominal imaging with high soft tissue resolution.<sup>10</sup>

None of our patients underwent preoperative biopsy because we thought that the results of aspiration biopsy or frozen section would not change our decision of radical resection, and aspiration cytopathology would not reliably provide adequate information concerning differentiation of the lesion from other (especially high grade) cystic tumors, at least not in our institution.

The diversity of immunostaining emphasizes the tumor cell phenotype expressing epithelial, mesenchymal, and endocrine lines. These tumors tend to develop in women and the presence of progesterone receptors which were positive in all of our patients, may indicate a possible genital link.<sup>11,12</sup> This idea might be substantiated by closeness of genital ridges to pancreatic anlage during embryogenesis.<sup>13</sup>

Based on high rates of positivity for markers of various pancreatic cells (neuron specific enolase, alfa 1-antitrypsin, vimentin, antichymotrypsin) it has been suggested that the tumor originates from pancreatic pluripotent stem cells.<sup>14-16</sup> Younger patients tend to present with slightly smaller tumors and this may suggest that these tumors arise in





**FIGURE 5:** Tumor tissue containing papillary structures and hemorrhagic regions. (HE, x100).



**FIGURE 6:** Postoperative specimen of the patient shows distal pancreatotomy and splenectomy.

childhood and grow slowly over time.

Clearly malignant lesions such as pseudopapillary carcinoma or borderline tumors which show subtle or limited morphological malignant characteristics may be found.<sup>17</sup>

Local recurrence after incomplete resection represents the malignant biological nature of SPN and metastases are found in relatively older patients.<sup>18</sup>

Cure is possible with only surgical resection. Conservative resection provided safe margins is the treatment of choice if feasible. In most cases distal pancreatectomy would be sufficient. Total pancreatectomy with or without splenectomy would be warranted in some cases. In one case we performed distal pancreatectomy without splenectomy. The role of splenectomy in the prevention of local recurrence and impact on the cure rate of the operation is not known in this particular tumor. The presence of metastasis does not exclude surgical resection because of the clinical benefits associated with tumor resection.<sup>19</sup> Tumor related death is uncommon.<sup>20</sup> The relatively high rate of recurrence among patients with locally excised tumors and the low operative morbidity and mortality rates after pancreatic resection strongly support the concept of radical, curative tumor resection, regardless of the patient age.<sup>21,22</sup>

Chemotherapy and radiotherapy have both proved ineffective despite the report of a single

case by Fried et al who concluded that after radiotherapy if the patient required surgery, it would be far less radical than the operation that the patient would have needed initially.<sup>23</sup> However, it remains to be shown in a number of patients whether the lesion will eventually prove to be controlled permanently by radiotherapy. We did not use radio or chemotherapy in our patients, and no recurrence or metastasis was found between 1-6 years.

In the literature, Aydiner F et al reported three cases of SPN located in the distal pancreas.<sup>24</sup> Distal pancreatectomy with splenectomy was performed in 2 of these cases because of adherence to the spleen. Only distal pancreatectomy was performed in the 3rd case. All patients survived with no recurrence or distant metastasis. They suggested that the histomorphologic features of this tumor were rather characteristic, and differential diagnosis via immuno-histochemistry is not helpful.

Zeytinlu M et al reported four consecutive patients with SPN of the pancreas.<sup>25</sup> In one case, splenectomy was performed with distal pancreatectomy because of the adherence to the spleen. For the remaining three patients, distal pancreatectomy and enucleation were performed. All patients survived with no recurrence or metastasis.

By virtue of its biological behavior, papillary cystic neoplasm of the pancreas is a highly curable tumor, unlike the more common adenocarcinoma.

Once the diagnosis is established, the definitive resection will be curative and will provide excellent survival in most of the cases.

Since the presentation of the disease is not specific despite supportive modern diagnostic imaging, the preoperative diagnosis may still be very diffi-

cult. This unusual tumor should be considered in the differential diagnosis of large abdominal masses, especially in young females. Surgeons should be aware of this uncommon malignant lesion with excellent prognosis, complete curative excision of which is possible.

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