A partially calcified cystic lesion in the area of the pancreatic tail was incidentally detected when a 57-year-old woman underwent chest computed tomography (CT) for evaluation of chronic cough (Figure 1a). She had no history of pancreatitis, but reported severe abdominal trauma approximately 20 years earlier. A thrombosed pseudoaneurysm was therefore considered in the differential diagnosis. Endoscopic ultrasound (EUS) showed a calcified lesion 3.5 x 2.3 cm in size in the pancreatic tail, with an obvious hypoechogenic solid component as well as cystic areas (Figure 1b). Fine-needle aspiration (FNA) revealed cells arranged in distinct papillary-like structures. Immunohistochemistry was negative for synaptophysin and positive for low-molecular-weight cytokeratin and vimentin. The patient underwent distal pancreatectomy. Histological examination of the resected specimen confirmed a solid pseudopapillary tumor (SPT) (Figure 2).

SPTs of the pancreas, first described by Frantz in 1959 [1], are uncommon tumors of low malignant potential that most frequently affect young women and are often found incidentally. In a recent review [2] of 24 patients (20 women, mean age 39 years), the median tumor size was 8 cm – considerably larger than in the patient described here. A total of 22 patients underwent resection, four of whom had coexistent liver metastases. Resection was not possible in two patients due to involvement of mesenteric vessels. However, the unresectable patients had a long-term survival (up to 13 years), emphasizing the often indolent nature of this tumor. Expression of vimentin, α1-antitrypsin, neuron-specific enolase, and progesterone receptors is typical for SPTs [3]. Recent reports suggest that SPTs are genetically distinct from pancreatic ductal adenocarcinomas and typically harbor mutations in the APC/beta-catenin pathway [4,5].

Figure 1 Computed tomogram, showing a partially calcified mass in the area of the pancreatic tail. b Endoscopic ultrasonography using a linear-array echo endoscope reveals a solid hypoechogenic tumor with cystic areas (large arrow) and calcifications (arrowhead). Fine-needle aspiration is being carried out (the small arrow indicates the 22-gauge needle).

Figure 2 Histology showing a solid monomorphic proliferation of uniform cells often separated by fibrous stroma. Note the low mitotic rate. Similar cells surrounding fibrovascular stalks give the tumor a pseudopapillary architecture (hematoxylin-eosin, original magnification x 40).
EUS made it possible to delineate several typical SPT features, including the mixture of cystic and solid components in a well-demarcated tumor, as well as partial calcification. FNA provided the definitive diagnosis. This case underscores the value of EUS/FNA in the evaluation of patients with pancreatic lesions of unclear etiology.

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References

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