Pseudopapillary tumors (PPT) of the pancreas are very rare, comprising 0.3–2.7% of all pancreatic tumors, and they occur mostly in young women. Generally, they are benign, but in rare cases they can enlarge, invade adjacent organs, and metastasize distantly. Radiological assessments and biochemical markers are important for diagnosing tumor characteristics. The main treatment is tumor resection.

PRESENTATION OF CASE: An 18-year-old female was referred to our department suffering from abdominal discomfort and upper quadrant abdominal pain. Abdominal computed tomography (CT) revealed a 6–5 cm mass between the pancreatic head and right adrenal gland (Fig. 1). The histological assessment was a solid PPT of the pancreas with intact surgical borders.

DISCUSSION: PPT are very rare, comprising approximately 5% of cystic pancreatic tumors and ~1% of exocrine pancreatic neoplasms and present mainly during the second and third decades of life. PPTs are usually indolent tumors. As such, they tend to produce vague nonspecific symptoms or may be detected incidentally on imaging. Complete surgical resection (R0) is the most effective therapy for PPT. CONCLUSION: Although PPT is a very rare, benign tumor, it has the potential to metastasize to adjacent and distant organs. Consequently, they should be detected early, so that they can be treated surgically before malignant conversion.

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are usually indolent tumors. As such, they tend to produce vague nonspecific symptoms or may be detected incidentally on imaging. As these lesions enlarge, they may then cause symptoms from mass effect, such as vomiting and early satiety due to gastric outlet obstruction. They present mainly during the second and third decades of life [10]. The origin of solid PPT remains unclear. The tumor cells moderately express the progesterone receptor, and this tumor predominantly develops in women, suggesting an association between female sex hormones and tumorigenesis [11–14].

The preoperative diagnosis is based primarily on radiological assessment that includes abdominal CT and magnetic resonance imaging (MRI). The PPT appears as an encapsulated, well-defined mass with central areas of calcification, necrosis, hemorrhage and cystic degeneration [15]. Histological confirmation is not necessary, although in unresectable cases, fine needle biopsy may be performed with 62–70% preoperative accuracy [6,7,16].

Abdominal discomfort and pain are the most common symptoms [1,17]. Our patient presented with abdominal discomfort. There are no pathognomonic features on blood investigations and tumor markers are usually unremarkable.

PPT often develop in the pancreatic tail or head [2,18]. Most metastases of these tumors are to the liver, lymph nodes, and peritoneum [2,17,18]. In our case, the tumor was separate from the pancreas and duodenum, with no adjacent organ metastasis.

Histologically, the tumor is a heterogeneous mass that contains hemorrhagic, cystic, necrotic, and calcific components, and is encapsulated with a sharp margin [2,11,19]. Since it is usually a benign tumor of low malignant potential, the prognosis after surgical resection is excellent. Complete surgical resection (R0) is the most effective therapy for PPT [20]. Pancreatectoduodenectomy, distal pancreatectomy (with or without splenectomy), middle pancreatectomy, or enucleation can be performed based on the location, size, angioinvasion, and adjacent organ compromise. A classic or pylorus-preserving pancreatectoduodenectomy is indicated in cases with tumors located in the pancreatic head or uncinate process. Given the excellent prognosis, in patients with PPTs which involve the superior mesenteric vein or/and portal vein, vein resection and reconstruction should be considered. Distal pancreatectomy with or without splenectomy can be performed for tumors located in the pancreatic body or tail. For patients with tumors located in the neck or body of the pancreas, without vessel involvement, we prefer to perform middle pancreatectomy with distal pancreateojunostomy, preserving the rim of the head, the uncinate process, and the tail portion. If the tumor is unresectable, radiotherapy is a treatment option [3].

In conclusion, although PPT is a very rare, benign tumor, it has the potential to metastasize to adjacent and distant organs. Consequently, they should be detected early, so that they can be treated surgically before malignant conversion. These tumors can be resected easily because of their sharp margins. As in our case, resection might be a curative treatment.

Conflict of interest

There is no conflict of interests.

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Author contribution

Serdar Karakas: writing.
Abuzer Dirican: data analysis.
Vural Soyer: study design.
Suleyman Koc: data collections.
Veysel Ersan: grammar checks.
Mustafa Ates: data collections.
Consent

Instant confirmation forms is taken from the patient at the first application.

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