CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 10 (2015) 118–120

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

A pancreatic pseudopapillary tumor enucleated curatively

Serdar Karakas[a], Abuzer Dirican[a,*], Vural Soyer[a], Süleyman Koç[a], Veysel Ersan[b], Mustafa Ates[b]

[a] Inonu University, Faculty of Medicine, Department of General Surgery, Malatya, Turkey
[b] Inonu University, School of Medicine, Department of General Surgery, Malatya, Turkey

ARTICLE INFO

Article history:
Received 9 February 2015
Received in revised form 11 March 2015
Accepted 23 March 2015
Available online 25 March 2015

Keywords:
Pancreas
Pseudopapillary
Enucleation

ABSTRACT

INTRODUCTION: 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.

1. Introduction

Pseudopapillary tumors (PPT) of the pancreas are very rare, comprising 0.3–2.7% of all pancreatic tumors [1–3]. The first case was described by Frantz in 1959, and thus PPT are referred to as Frantz tumors [4]. The well-capsulated ovoid tumor is most common in young women. Radiological images show heterogenic cystic, solid, and calcified components. Distant organ metastasis is rare. Only 10% of these tumors occur in males, although they may be more aggressive in males [5]. Still, they are usually benign. The most common symptoms of PPT are abdominal discomfort, abdominal pain, and jaundice in rare cases; often they are asymptomatic [6–7]. Complete resection is the main treatment option for these tumors, with a 95% 5-year survival rate [8]. Here, we present a completely resectable PPT in a young female who was treated curatively.

2. Case report

An 18-year-old female was referred to our department suffering from abdominal discomfort and upper quadrant abdominal pain. She had no known disease that could have caused these symptoms, nor a family history of illness. There was no tenderness or palpable mass on examination of the abdomen. Abdominal computed tomography (CT) revealed a 6- × 5-cm mass between the pancreatic head and right adrenal gland (Fig. 1). No metastasis to adjacent organs was seen. Biochemical and tumor markers, including bilirubin, amylase, and lipase, were normal. The mass was removed successfully without complications. There was no invasion of the pancreas or duodenum, although it was adherent to the duodenum and pancreatic head (Fig. 2). The patient was discharged 2 days postoperatively with no complications. There were no complications or recurrence at the 3-month follow-up, and no chemo- or radiotherapy was administered. The histological assessment was a solid PPT of the pancreas with intact surgical borders.

3. Discussion

PPT are very rare, comprising approximately 5% of cystic pancreatic tumors and ~1% of exocrine pancreatic neoplasms [9]. PPTs

* Corresponding author at: Inonu University, Faculty of Medicine, Department of General Surgery, 44315 Malatya, Turkey. Tel.: +90 533 4233462.
E-mail address: abuzerdirican@hotmail.com (A. Dirican).

http://dx.doi.org/10.1016/j.ijscr.2015.03.040

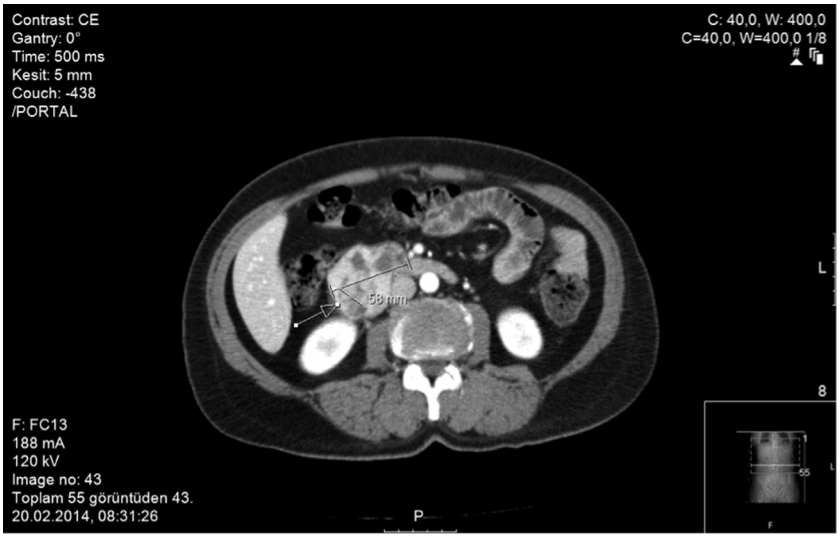

Fig. 1. 6 × 5 cm mass between pancreatic head, and right surrena.

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.

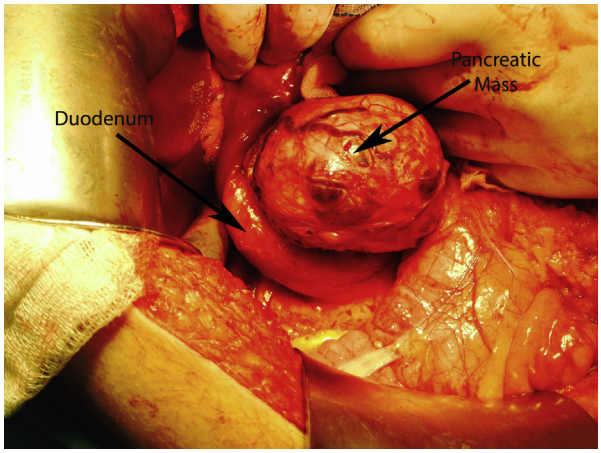

Fig. 2. Mass between duodenum and pancreatic head.

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]. Pancreatoduodenectomy, 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 pancreatoduodenectomy 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 pancreatojejunostomy, 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.

Funding

There is no sources of funding for our research.

Author contribution

Serdar Karakaş: 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.

References

[1] S.G. Tipton, T.C. Smyrk, M.G. Sarr, G.B. Thompson, Malignant potential of solid pseudopapillary neoplasm of the pancreas, Br. J. Surg. 93 (6) (2006) 733–737.
[2] C.M. Kang, K.S. Kim, J.S. Choi, H. Kim, W.J. Lee, B.R. Kim, Solid pseudopapillary tumor of the pancreas suggesting malignant potential, Pancreas 32 (3) (2006) 276–280.
[3] J.A. Zauls, A.E. Dragun, A.K. Sharma, Intensity-modulated radiation therapy for unresectable solid pseudopapillary tumor of the pancreas, Am. J. Clin. Oncol. 29 (6) (2006) 639–640.
[4] V.K. Frantz, Papillary tumors of the pancreas: benign or malignant tumors of the pancreas, in: U.K. Frantz (Ed.), Atlas of Tumor Pathology, 1st Series, Fascicles 27 and 28, Armed Forces Institute of Pathology, Washington, DC, 1959, pp. 32–33.
[5] C. Sperti, M. Berselli, C. Pasquali, D. Pastorelli, S. Pedrazzoli, Aggressive behavior of solid-pseudopapillary tumor of the pancreas in adults: a case report and review of the literature, World J. Gastroenterol. 14 (2008) 960–965 [PMID: 18240360].
[6] B.E. Crawford Jr., Solid and papillary epithelial neoplasm of the pancreas, diagnosis by cytology, South. Med. J. 91 (10) (1998) 973–977.
[7] S. Reddy, J.L. Cameron, J. Scudiereetal, Surgical management of solid-pseudopapillary neoplasms of the pancreas (Franz or Hamoudi tumors): a large single-institutional series, J. Am. Coll. Surgeons 208 (2009) 950–957.
[8] S. Potrc, R. Kavalar, M. Horvat, E.M. Gadzijev, Urgent whipple resection for solid pseudopapillary tumor of the pancreas, J. Hepatobiliary Pancreatic Surg. 10 (2003) 386–389, http://dx.doi.org/10.1007/s00534-002-0836-1 [PMID: 14598141].
[9] D.S. Klimstra, B.M. Wenig, C.S. Heffess, Solid pseudopapillary tumor of the pancreas: a typically cystic carcinoma of low malignant potential, Semin. Diagn. Pathol. 17 (2000) 66–80.
[10] P.F. Yu, Z.H. Hu, X.B. Wang, J.M. Guo, X.D. Cheng, Y.L. Zhang, Q. Xu, Solid pseudopapillary tumor of the pancreas: a review of 553 cases in Chinese literature, World J. Gastroenterol. 16 (2010) 1209–1214.
[11] M. Kosmahl, L.S. Seada, U. Janig, D. Harms, G. Kloppel, Solid-pseudopapillary tumor of the pancreas: its origin revisited, Virchows Arch. 436 (2000) 473–480.
[12] T. Papavramidis, S. Papavramidis, Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature, J. Am. Coll. Surgeons 200 (2005) 965–972.
[13] S.M. de Castro, D. Singhal, D.C. Aronson, et al., Management of solid-pseudopapillary neoplasms of the pancreas: a comparison with standard pancreatic neoplasms, World J. Surg. 31 (2007) 1130–1135.
[14] G. Pettinato, D. di Vizio, J.C. Manivel, S.E. Pambuccian, P. Somma, L. Insabato, Solid-pseudopapillary tumor of the pancreas: a neoplasm with distinct and highly characteristic cytological features, Diagn. Cytopathol. 27 (2002) 325–334.
[15] E.L. Seung, J.Y. Jang, W.H. Dae, K.W. Park, S.W. Kim, Clinical features and outcome of solid pseudopapillary neoplasm: differences between adults and children, Arch. Surg. 143 (2008) 1218–1221.
[16] N. Jhala, G.P. Siegal, D. Jhala, Large, clear cytoplasmic vacuolation: an under-recognized cytologic clue to distinguish solid pseudopapillary neoplasms of the pancreas from pancreatic endocrine neoplasms on fine-needle aspiration, Cancer 114 (2008) 249–254.
[17] H.E. Seo, M.K. Lee, Y.D. Lee, S.W. Jeon, C.M. Cho, W.Y. Tak, et al., Solid-pseudopapillary tumor of the pancreas, J. Clin. Gastroenterol. 40 (10) (2006) 919–922.
[18] D. Santini, F. Poli, S. Lega, Solid-papillary tumors of the pancreas: histopathology, JOP 7 (1) (2006) 131–136.
[19] K.K. Roggin, J. Chennat, A. Oto, A. Noffsinger, A. Briggs, J.B. Matthews, Pancreatic cystic neoplasm, Curr. Prob. Surg. 47 (6) (2010) 459–510.
[20] S. Ji, J. Xu, B. Zhang, Y. Xu, C. Liu, J. Long, Q. Ni, X. Yu, Management of a malignant case of solid pseudopapillary tumor of pancreas: a case report and literature review, Pancreas 41 (2012) 1336–1340.