CASE REPORT

Frantz's Tumor (Solid Pseudopapillary Tumor) of the Pancreas. A Case Report

Settar Bostanoglu, Emrah Otan, Saadet Akturan, Enver Okan Hamamci, Akin Bostanoglu, Aysun Gokce, Levent Albayrak

Numune Training and Research Hospital. Ankara, Turkey

ABSTRACT

Context A solid pseudopapillary tumor of the pancreas is a rare neoplasm which, for the most part, affects young women and has a relatively favorable prognosis with a low malignant potential. These tumors usually have unclear clinical features and may form very large masses before being diagnosed. **Case report** We report the case of a 29-year-old woman who underwent complete resection of the tumor using a distal pancreatectomy and splenectomy procedure. The patient is being followed-up and in good condition. A review of the relevant literature is also presented. **Conclusions** A solid pseudopapillary tumor of the pancreas is a rare condition with a low potential for malignancy and favorable prognosis; surgical resection is generally curative.

INTRODUCTION

A solid pseudopapillary tumor of the pancreas is an rare neoplasm accounting for less than 2% of exocrine pancreatic neoplasms [1, 2]. This rare entity was first described by the author Dr. Frantz in 1959, and was called "papillary tumor of the pancreas, benign or malignant" [1, 3]. Until it was defined by the World Health Organization (WHO) in 1996 as "solid pseudopapillary tumors" of the pancreas, this tumor was described by using various names including "solid cystic tumor", "papillary cystic tumor", "papillary epithelial neoplasia", "solid and papillary epithelial neoplasia", "papillary epithelial tumor", "Frantz's tumor", "solid and papillary tumor", solid-cystic-papillary epithelial neoplasm", "benign or malignant papillary tumor of the pancreas" and "adenocarcinoma of the pancreas in childhood" [4, 5]. These neoplasms with low malignant potential affect, for the most part, young women and tend to be treated with aggressive resection. After resection and follow-up, they generally have a relatively favorable prognosis [1]. We report our clinical experience with a solid pseudopapillary tumor and include a review of the current literature.

CASE REPORT

A previously healthy 29-year-old woman who presented with epigastric abdominal pain of 3 months duration was admitted with an abdominal mass. She had had no previous history of hospitalization. Physical examination revealed mild epigastric tenderness and a palpable mass about 10 cm. in diameter, located in the abdomen extending from the upper left quadrant to the upper right quadrant. Routine laboratory test results including tumor marker levels (carcinoembryogenic antigen, alpha-fetoprotein, CA 19-9) were within normal limits. Abdominal ultrasonography demonstrated a heterogeneous semisolid mass with cystic components adjacent to the left lobe of the liver. On abdominal CT, a solid mass, 11 cm in diameter, involving the pancreatic corpus and tail was observed (Figure 1). An abdominal MRI scan defined the mass as originating from the pancreatic corpus and tail, 10 cm. in diameter. Arterial and venous Doppler ultrasonography examination showed the mass to have an intimate relationship with the splenic vein. At laparotomy, exploration revealed an encapsulated mass of 15 cm in diameter in the tail and body of the pancreas involving the splenic artery and vein. There was no evidence of intra-abdominal metastasis. Distal pancreatectomy and splenectomy were performed. A solid pseudopapillary tumor was diagnosed on pathologic examination of the specimen (Figure 2). The postoperative period was complicated by a localized cyst at the operation site which was diagnosed by a CT scan performed 11 days after surgery. The cyst was 10 cm in diameter and percutaneous aspiration was carried out using ultrasonographic guidance. Diagnostic laboratory tests showed high levels of amylase which was interpreted to be proof of pancreatic origin. Abdominal CT carried out 1 and 3 months after the surgery showed the diameter of the cyst to have decreased (2 cm); the

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Correspondence Emrah Otan
Emek Mah. 8.Cad. No:217/11, Cankaya, 06510, Ankara, Turkey
Phone: +90-533.634.5856; Fax: +90-312.310.3460
E-mail: emrahotan@yahoo.de

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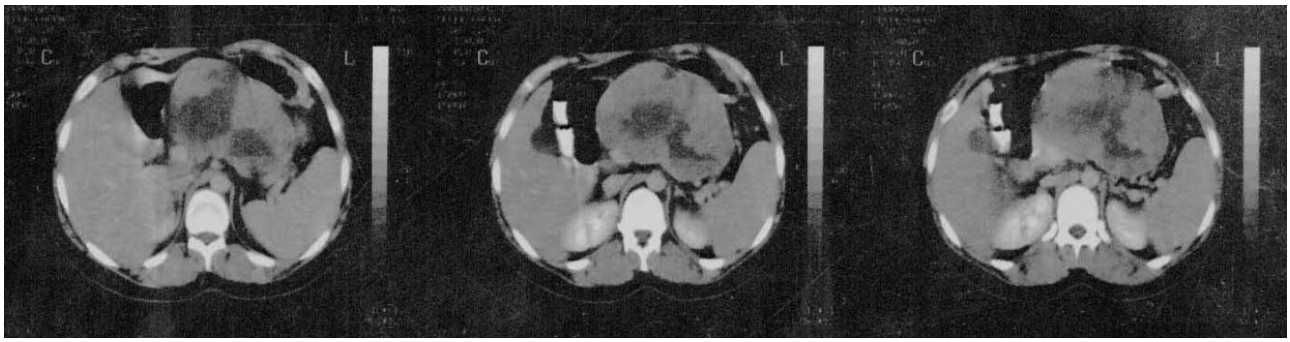

Figure 1. Computed tomographic scan. Encapsulated complex, solid and cystic mass, 11 cm in diameter, localized in corpus and tail of the pancreas.

physical examination findings and laboratory tests did not reveal any significant infectious process. The patient has been followed-up as an out-patient for 3 years and has remained healthy without any sign of local recurrence or distant metastasis.

DISCUSSION

Solid-pseudopapillary tumors are rare primary neoplasms of the pancreas with a low malignant potential which tend to occur primarily in young women [1, 2].

These neoplasms account for 5% of cystic pancreatic tumors and 1-2 % of exocrine pancreatic neoplasms [5]. There have been 718 cases reviewed in the English literature; female dominance has been found with a ratio of 10:1 and there is a mean age of 22 years [6].

The origin of solid pseudopapillary tumors still remains unclear. These neoplasms have been suggested to have a ductal epithelial, neuroendocrine, multipotent primordial cell, or even an extra-pancreatic genital ridge angle-related cell origin [5, 7].

The majority of the tumors are located in the pancreatic body and tail [5, 8, 9]. A combination of solid and cystic components with cellular degenerative changes alternating with pseudopapillary formation are frequently mentioned among the characteristic features observed with a light microscope which distinguish pseudopapillary tumors from other neoplasms of the pancreas [2, 10]. The solid portions of the tumor are composed of uniform and polygonal epithelioid cells with well-vascularized stroma and a discohesive arrangement [9, 10]. Specific findings for cellular differentiation in solid pseudopapillary tumor cells remain unclear and differential diagnosis requires immunohistochemical staining. Solid pseudopapillary tumor cells are usually absent when stains for endocrine markers are used (insulin, glucagon, somatostatin, serotonin, gastrin, vasoactive intestinal peptide, bombesine, chromogranin, lysosome and CD68) and are characteristically positive for vimentin, CD10, CD56, and alpha-1-antitrypsin [9, 10]. These cells may also reveal focal immunoreactivity for cytokeratin, neuron-specific enolase and synaptophysin, demonstrate abnormal nuclear localization of beta-catenin, present progesterone receptors and may express galectin-3, all of which are useful in differentiating them from endocrine pancreatic tumor cells [10, 11].

Most of the patients present with unclear clinical features including abdominal discomfort, mild abdominal pain, increased abdominal girth, poor appetite and nausea which are related to tumor compression on the adjacent organs [2, 6]. When a palpable mass is presented, the average size of the tumor becomes quite remarkable (8-10 cm) [2, 7]. The majority of the small tumors are occasionally diagnosed during complementary imaging studies such as ultrasound or CT scan of the abdomen [8, 12]. Solid pseudopapillary tumors have not been associated with specific clinical laboratory test findings including serum tumor markers [8, 9].

The radiologic features of the tumor present similarities in most cases. Although CT scanning is the most frequent technique in diagnosing a solid pseudopapillary tumor, sonographic examination and magnetic resonance imaging (MRI) also define these hypervascular, well-encapsulated round mass with mixed cystic and solid components [8, 12]. Echo-endosonography may provide fine-needle puncture biopsy with an option of preoperative pathologic diagnosis of the tumor [12].

Despite the locally aggressive features, the tumor has a low-grade malignant potential and tends to have a favorable prognosis, even in the presence of metastatic disease [1, 7, 9]. Metastases with an incidence of 15%,

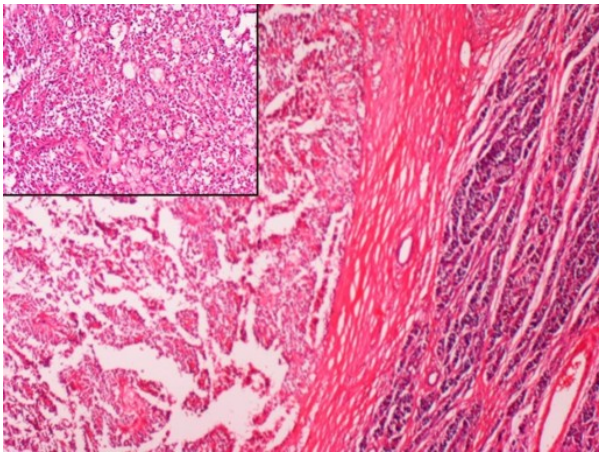

Figure 2. Tumor tissue presenting a notable encapsulation by the pancreatic parenchyma under low power (outer square, H&E, x4). Solid pseudopapillary tumor tissue presenting characteristic pseudopapillary architecture at a higher magnification (inner square, H&E, x100).

most of which are hepatic, and local recurrence have rarely been reported in the long term follow-up of these patients [1, 7, 8]. Most of the metastatic patients are male and these lesions are frequently found at the time of diagnosis [7, 8]. Lymph node involvement is a rare condition which has been reported in 4 cases to date [2, 10]. Overall 5-year survival is as high as 97% in patients undergoing surgical resection [2, 10]. A number of cases having a more aggressive but shorter clinical course leading to death have been reported in which the mean age of the patients is older and metastatic disease to the liver or peritoneum was diagnosed at the time of initial presentation [9, 10]. The mean survival rate of these patients was 90 months [10].

Surgery is the treatment of choice even in the case of distant hepatic metastasis or local recurrence which are not contraindications for surgical therapy [1, 6, 13, 14, 15]. There is limited experience regarding chemotherapy and radiotherapy with or without the presence of metastatic disease. Some experimental regimes have been used including 5-fluorouracil, doxorubicin, streptozocin, cisplatin, topotecan, iphosphamide, and etoposide without a significant clinical response [2, 12]. A favorable response to radiotherapy in locally advanced unresectable disease has been reported [1, 2].

In conclusion, a solid pseudopapillary tumor of the pancreas is a rare condition with a low potential for malignancy. These tumors usually affect women in the second or third decades of life. The prognosis is favorable even in the presence of distant metastasis. Although surgical resection is generally curative, a close follow-up is advised in order to diagnose a possible local recurrence or distant metastasis and choose the proper therapeutic option for the patient.

Conflict of interest The authors have no potential conflicts of interest

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