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Case report

Solid pseudopapillary tumors of the pancreas in young women: Case report

Taoufik El Abbassi, Abdessamad Elazhary *, Mohamed Ouchane, Anas El Wassi, M. Rachid Lefriyekh

Department of General Surgery I, CHU Ibn Rochd, Casablanca, Morocco
Faculty of Medicine and Pharmacy of Casablanca, Hassan II University, Morocco

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ABSTRACT

Introduction and importance: Solid pseudopapillary tumor of the pancreas (SPTP) is a rare tumor of the exocrine pancreas, with undetermined etiopathogeny, which most often affects young women. The clinical and physical signs are non-specific and despite the progress of complementary examinations, Confirmation is usually anatomopathological. Surgical resection is the only curative treatment.

Case presentation: We report the case of a 17-year-old girl, consulted for a left hypochondrium mass which imaging concluded to be a corporal-caudal tumor mass of the pancreas for which a complete surgical excision was performed and whose anatomopathological study confirmed a pseudo-papillary and solid tumor of the pancreas. The positive diagnosis of SPTP remains difficult and is usually made on pathological analysis with immunohistochemical study.

Discussion: Solid pseudopapillary tumor of the pancreas is a rare anatomic-clinical entity, first described by Frantz in 1959. The clinical manifestations of SPTP are not specific. Biologically, no signs are predictive of SPTP and imaging usually shows a well encapsulated mass with both solid and cystic components. The curative treatment of SPTP is exclusively surgical and consists of a complete removal of the tumor with its capsule because of its degenerative potential SPTP has a good prognosis with a recurrence rate of 10–15% and 95% survival at 5 years.

Conclusion: Solid pseudopapillary tumor of the pancreas is a rare tumor of the exocrine pancreas. Its evolution is slow. Preoperative diagnosis remains difficult despite the progress of complementary examinations. Surgical resection is the only curative treatment. Its prognosis remains excellent.

1. Introduction

Pancreatic pseudopapillary solid tumor (SPTP) is an extremely rare epithelial tumor, accounting for less than 2% of pancreatic exocrine tumors and less than 5% of cystic pancreatic tumors [1]. It is a tumor with a low malignant potential and its etiopathogeny is still uncertain [2]. It mainly affects young women and their diagnosis is still based on immunohistochemical study despite the progress in imaging [3]. Radical treatment is surgery requiring complete removal of the tumor mass [4].

Our patient was 17 years old, consulted for a mass of the left hypochondrium for which a complete surgical resection was performed. The confirmation of a solid pseudo-papillary tumor of the pancreas was done by anatomopathological and immunohistochemical study of the surgical specimen. This article is respecting the SCARE Checklist guidelines [5].

2. Observation

A 17-year-old woman, operated on for acute appendicitis 4 years ago, had been presenting for one month with pain in the left hypochondrium and epigastrium of progressive installation, associated with vomiting without digestive hemorrhage or transit disorder, all evolving in a context of apyrexia and conservation of the general state.

Abdominal examination revealed a 20 cm mass occupying the left hypochondrium and epigastrium, firm in consistency, mobile in the superficial plane and fixed to the deep plane. The biological check-up, including hemoglobin and renal function, was normal and the tumor markers CA 19.9 and ACE were negative.

A thoracoabdominal CT scan showed a necrotic tissue mass in the omental bursa measuring 20 cm, which could have originated from either the gastric or pancreatic (Fig. 1).

* Corresponding author at: Faculty of Medicine and Pharmacy of Casablanca, Hassan II University, Morocco.
E-mail address: elazhary55@gmail.com (A. Elazhary).

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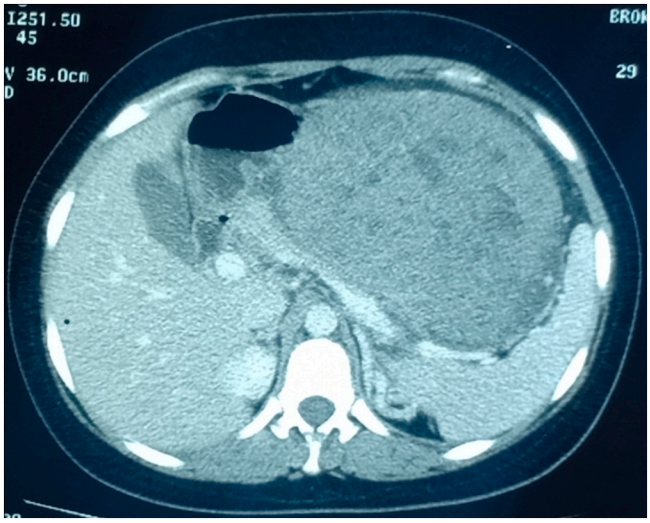

Fig. 1. CT image showing a large heterogeneous tumor mass, developed at the corporal-caudal region of the pancreas.

An echo-endoscopy was performed, showing the presence of a heterogeneous solid mass, arriving in contact with the corporo-caudal part of the pancreas without loss of separation line, laminating the portal vein and the spleno-mesenteric venous confluence. The patient was operated and the surgical procedure consisted in a corporo-caudal resection of the pancreas removing the entire tumor mass with a safety margin of 1 cm on the healthy pancreas (Fig. 2).

The histological study found an encapsulated operative specimen (Fig. 3) weighing 2300 g, white-beige in color with soft consistency, with many cystic formations of hemorrhagic content in places, the tumor proliferation corresponded to a pseudo papillary structure in solid mass.

The tumor cells diffusely expressed anti-vimentin antibody, beta-Catenin and CD 10 and a local positivity of synaptophysin, compatible with a solid pseudo papillary tumor of the pancreas with complete resection. Postoperative follow-up was simple with a one-year follow-up.

3. Discussion

Solid pseudopapillary tumor of the pancreas is a rare anatomic-clinical entity, first described by Frantz in 1959 [6]. It usually affects young women, however, rare cases in men and elderly people have been reported. It is a tumor that can develop on the head, body or tail of the pancreas, with a clear predominance of the corporal-caudal region with a rate of 64% [7]. Rare cases of extra-pancreatic localizations have also been described with a rate of less than 1%, retroperitoneal, duodenal, mesocolic and hepatic [8]. The clinical manifestations of SPTP are not specific, it can be revealed by atypical abdominal pain, or the appearance of a palpable abdominal mass on clinical examination or an accidental finding during an imaging examination performed for another reason [9]. The increase in size of the tumor mass may lead to signs of digestive, biliary, or vascular compression [10]. More rarely, the tumor is discovered following a spontaneous bleeding complication or secondary to abdominal trauma [11]. Biologically, no signs are predictive of SPTP and imaging usually shows a well encapsulated mass with both solid and cystic components. On ultrasound, the echogenicity of these tumors is variable depending on the size of the cystic areas. CT scan shows a large, heterogeneously dense, solid-cystic pancreatic mass, surrounded by a capsule that enhances especially in the late stage [9–11]. Magnetic resonance imaging (MRI) is the most efficient examination, which shows hyper intense lesions in T1 and T2, of intracystic hemorrhagic remodeling surrounded by a capsule often in the form of a hypo intense border on T1 sequences [12]. Echo-endoscopy can be hampered by the voluminous nature of the lesions, however, the lesion is echogenic, heterogeneous, with a hypoechoic peripheral halo [13].

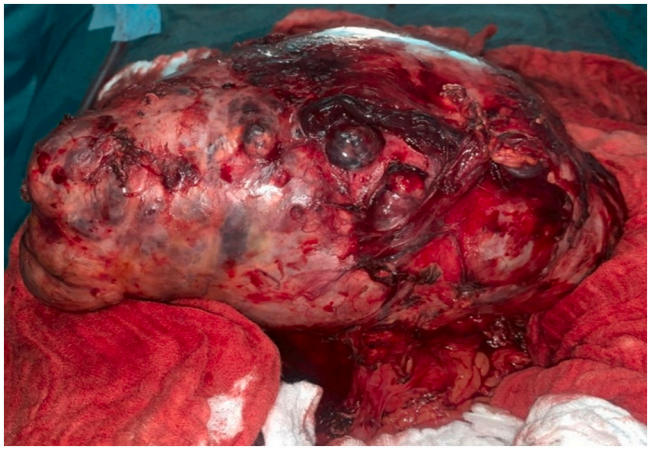

Fig. 2. Intraoperative image of the tumor mass with peripheral vascularization and areas of intratumoral hemorrhage.

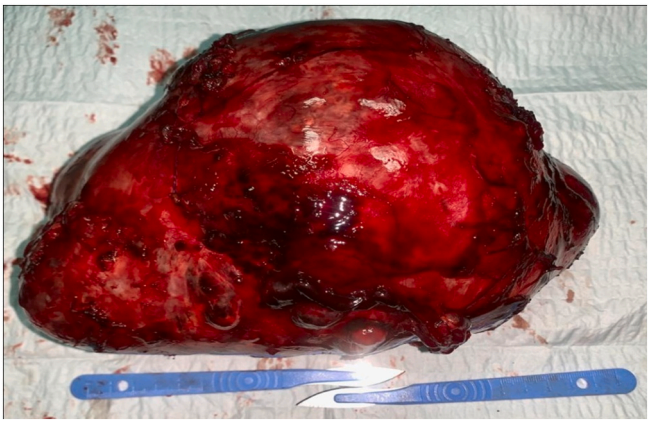

Fig. 3. Surgical specimen image of solid pseudopapillary tumor of complete resection pancreas.

Preoperative biopsy under radiological or endoscopic control can lead to complications such as bleeding, pancreatic fistula and biliary fistula with the risk of tumor dissemination on its way, extra pancreatic diffusion of the tumor, and transformation of a well localized tumor, with a good prognosis, into an aggressive tumor [9].

The positive diagnosis of SPTP remains difficult and is usually made on pathological analysis with immunohistochemical study [6]. Macroscopically, it is a generally bulky tumor, round or oval in shape, surrounded by a fibrous capsule [14]. The tumor consists of peripheral solid patches and central papillary structures. Mitoses are usually very rare [4]. The tumor cells are small, monomorphic, cuboidal or polygonal and often arranged around fibro-vascular septa. The stroma is usually endocrine-like, rich in blood capillaries. Vascular emboli are rare [3]. Immunohistochemical study is essential for diagnosis, and several specific markers have been identified; immunostaining with anti-vimentin antibody, a marker for germline cells, is positive in more than 90% of cases. Anti-alpha-1-antitrypsin antibody and anti-neuron specific enolase (NSE) antibody are positive in about 50% of cases [15].

The differential diagnosis is mainly with pancreatic pseudocysts, cystic neuroendocrine tumors, and pancreatic acinar cell tumors [16].

The curative treatment of SPTP is exclusively surgical and consists of a complete removal of the tumor with its capsule because of its degenerative potential. This removal ranges from a simple enucleation to a partial or even total pancreatectomy depending on the topography of the tumor, and must be extended in case of invasion of loco regional organs

[17]. Chemotherapy, radiotherapy and hormonal therapy are rarely used; their indication is discussed particularly in non-localized forms [18].

SPTP has a good prognosis with a recurrence rate of 10–15% and 95% survival at 5 years. Cases of prolonged survival have been reported even in the presence of liver or peritoneal metastases or in cases of incomplete surgery [10].

4. Conclusion

Solid pseudopapillary tumor of the pancreas is a rare tumor of the exocrine pancreas, with an attenuated malignancy that affects young women. Its evolution is slow. Preoperative diagnosis remains difficult despite the progress of complementary examinations. Surgical resection is the only curative treatment. Its prognosis remains excellent.

Consent

Informed consent was obtained from the patient for publication of this case report and accompanying image. A copy of the written consent is available for review by the Editor in Chief of this journal on request.

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Ethical approval

This case report is exempt from ethical approval at our institution.

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El Abbassi Taoufik: Corresponding author writing the paper and operating surgeon
DR EL Azhari Abdessamad: writing the paper and operating surgeon
Ouchane Mohamed: writing the paper and operating surgeon
El Wassi Anas: writing the paper
Mohamed Rachid Lefriyekh: study concept, correction of the paper.

Declaration of competing interest

The authors declare that they have no conflicts of interest in relation to this article.

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