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Solid-Pseudopapillary Tumor: Case Report and Literature Review

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Abstract

Solid-pseudopapillary tumor of the pancreas (SPT) or Frantz's tumor is a rare epithelial neoplasm that represents 0.3% to 2.7% of exocrine pancreatic tumors. These tumors occur mainly in young women and have a good prognosis. We present a case of a 19-year-old female patient who presented to the emergency for abdominal pain. Physical examination reveals a left hypochondrium mass. Ultrasound imaging showed an encapsulated caudal pancreatic mass with cystic components. The patient underwent tumor excision and lymph node removal. Macroscopically, the tumor was encapsulated and measuring 12 × 8 × 7 cm. It has a solid-cystic and hemorrhagic appearance inside. Histologically, the tumor had two components: solid and papillary. Tumor tissue showed monomorphic tumor cells radiating around blood vessels. Perineural invasion and vascular emboli were not seen. Three lymph nodes without metastases were observed. The diagnosis of solid-pseudopapillary tumor of the pancreas (SPT) or Frantz's tumor was retained. Solid-pseudopapillary tumor of the pancreas is rare and not always suspected by the physician. The clinical and imaging patterns are not specific. Anatomopathological examination confirmed the diagnosis. The patient underwent curative surgery.

Keywords

Pancreas, Tumor, Solid-Pseudopapillary, Frantz, Antananarivo

1. Introduction

Solid-pseudopapillary tumor of the pancreas (SPT), also known as Frantz's tumor, is an exocrine pancreatic tumor with uncertain potential of malignancy. It

is rare and accounts for 0.3% to 2.7% of pancreatic exocrine tumors [1]. This tumor was first described by Frantz in 1959 [2]. There is no known ethnic predilection [3]. Several names have been used to describe these tumors including solid cystic tumor, papillary cystic tumor, solid and papillary epithelial neoplasm, papillary cystic carcinoma, Hamoudi's tumour, and Frantz's tumour [3] [4]. It mainly affects young women and is characterized by its good prognosis and the polymorphism of the clinico-radiological profile making its diagnosis difficult, requiring an anatomopathological examination. To our knowledge, no Malagasy case has yet been published. We report a case of solid-pseudopapillary tumor of the pancreas in a young Malagasy woman.

2. Observation

The patient was a 19-year-old woman who presented for abdominal pain. There was no particular past history, neither surgical nor medical. The physical examination showed a tender abdomen, with the presence of a left hypochondrium curvature. Abdominal ultrasound revealed a large, corporal-caudal, encapsulated, solid and cystic pancreatic mass measuring 12 cm in long axis. The CT scan did not find any other tumor location. Tumor removal with lymph node dissection was performed and the specimen was sent for anatomopathological examination. The tumor specimen was encapsulated and measured 12 × 8 × 7 cm. The section slices were brownish-white, solid, with cystic and hemorrhagic areas. Microscopic examination showed a tumor proliferation with solid and papillary architecture (**Figure 1**). It was composed of monomorphic, small, cuboidal cells arranged around a conjunctivo-vascular septa (**Figure 1**). The tumor also demonstrated necrosis with cholesterol crystal deposits. No perineural engorgement or endovascular carcinomatous emboli were observed. Three non-metastasized nodes were found. The diagnosis retained was that of a solid-pseudopapillary tumor of the pancreas or Frantz's tumor. The patient underwent a complete resection of the tumor. The evolution was favorable with no recurrence at 12 months.

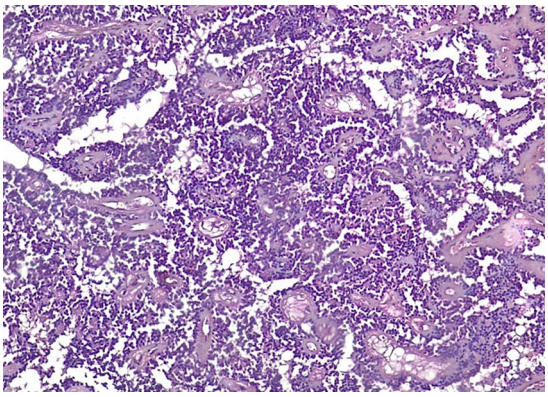

Figure 1. Pancreas, solid-pseudopapillary tumor, HE, ×200. Source: Pathological anatomy and cytology unit of the University hospital center of Joseph Ravoahangy Andrianavalona.

3. Discussion

SPT is a tumor with low malignant potential, classified as a borderline tumor of the pancreas. It was first described by Frantz in 1959 [2]. The terminology "pseudopapillary and solid tumor" was adopted by the WHO in 1996 [5]. It is a rare tumor and represents 0.3% - 2.7% of pancreatic tumors [1]. In the literature, most authors have reported isolated cases [6] [7] [8]. According to Papavramidis T in 2005, about 718 cases were reported in the English literature, and these were mostly isolated cases [9]. In our case, it was a 19-year-old woman. Almost all cases reported in the literature were female patients [10]. The average age of discovery varies between series, for most authors it is often around 22 years old [9]. Arzu Neşe Yener in Türkiye and Nabil Jakhlal in Morocco reported a case of 20 and 21 years of age respectively. Richard Azagoh-Kouadio [7] in Ivory Coast reported a case of SPT in an 11-year-old child. Elderly people are rarely affected [11]. The circumstances of discovery are variable and non-specific. Abdominal pain was the clinical manifestation in our patient. It is the most reported symptomatology in the literature [6] [8], followed by abdominal mass [10] [11]. The patient of Richard Azagoh-Kouadio [7] in Ivory Coast complained of an abdominal mass. SPT can also be discovered incidentally during a routine examination or for another reason. Other reported signs include vomiting, nausea, anemia, anorexia, weight loss and fever [10]. In all cases, imaging (ultrasound, CT, and MRI) showed an encapsulated tumor consisting of solid and cystic components, often without dilatation of the pancreatic duct [11]. None of these radiological features are specific to SPT, and they are also found in other pancreatic tumors, particularly in cystic neuroendocrine tumors and pancreatoblastoma. According to the topography, the tumor was corporal-caudal in our case, which is identical to the case reported by Richard Azagoh-Kouadio *et al.* in Ivory Coast [7]. In their series of 553 Chinese cases, Peng-Fei Yu *et al.* [10] found that the most frequent site was the head of the pancreas (39.8%), followed by the tail (24.1%), body and tail (19.5%), and the body of the pancreas (11.2%) [8]. Extra-pancreatic locations accounted for 1.8%. Tumors were nonmetastatic in 91.7% of cases. The macroscopic appearance of the tumor section slices varies with size. Larger tumors are friable because with growth, cystic degeneration and bleeding territories occurs. Smaller tumors tend to be more solid. In our case, the sections were solid and polycystic, with hemorrhagic remodeling. Peng-Fei Yu *et al.* [10] reported in their series that 60.12% were solid and cystic, 24.25% purely solid, and 15.63% purely cystic. Our patient's tumor was 12 cm. The cases of Arzu Neşe Yener [8] from Turkey and Nabil Jakhlal [6] from Morocco had 6 and 7 cm long axis, respectively. According to the literature, the average diameter was 7 cm, but varied from 1 cm to 25 cm [10] [11]. Histologically, according to WHO, the tumor is composed of small, cuboidal or polygonal, monomorphic cells. They are arranged in several layers around the fibro-vascular septa, giving a pseudopapillary structure. Mitoses and cytonuclear atypia are exceptional. Clusters of foamy histiocytes and giant cells around cholesterol crystals could be

also found. These characteristic features of SPT [12] were found in our patient as well as in the literature [6] [7] [8] [9] [10]. Histological criteria of malignancy can be seen in 10% - 15% and should lead to the diagnosis of pseudopapillary and solid carcinoma [12]. In our patient, we observed no invasion of adjacent structures, no vascular emboli, no perineural invasion, and no lymph node or distant metastases. On immunohistochemical examination, tumor cells show nuclear and cytoplasmic labeling for β-catenin with loss of E-cadherin from the cytoplasmic membrane [10] [11], positivity for progesterone receptor (PR), and alpha-1 antitrypsin and CD 10. Tumor cells also sometimes express neuroendocrine markers [10] [12]. Our patient and those of Nabil Jakhlal [6], Richard Azagoh-Kouadio [7], and Arzu Neşe Yener [8] underwent corporo-caudal pancreatectomy removing the tumor. The only curative treatment for SPT is surgery. The choice of operative method depends on the size, tumor location, and possible invasion of adjacent organs [6]. Local extension and metastasis are not a contraindication to surgery. The evolution of these tumors is usually favorable, even with metastasis. The overall 5-year survival rate of patients is approximately 95% [10]. Recurrences are less than 10%, and usually occur within 4 years after surgery [10].

4. Conclusion

Solid-pseudopapillary tumor of the pancreas is rare and not well known. It is not always suspected by the physician given that clinical and imaging findings are often non-specific, as observed in other pancreatic tumors. Therefore, anatomopathological examination is essential to confirm the diagnosis. Surgery is the only curative treatment.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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