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Solid Pseudopapillary Tumor of the Pancreas (Frantz Tumor) : A Case Report

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1. Abstract

Pancreatic Pseudopapillary Tumor (PTS) or Frantz tumor is a rare tumor of the exocrine pancreas with a low grade of malignancy. It mainly occurs in young women (20 to 40 years old) with an increased frequency in the Asian population. Their prognosis remains good especially after complete excision. We report the management of a 21-year-old patient for a Frantz tumor, revealed by abdominal pain. The diagnosis is made on the histological study associated with the immunohistochemistry of the caudal spleno-pancreatectomy part.

2. Introduction

Solid Pancreatic Pseudopapillary Tumors (TPPSP) are rare. They represent less than 2% of pancreatic cancers. They mainly affect young women. Their etiopathogenesis remains uncertain. They are characterized by clinical and radiological polymorphism which makes their diagnosis difficult. The only treatment guaranteeing prolonged survival is surgical resection. Their prognosis is excellent. The risk of metastases, mainly hepatic, is low (less than 15%). The risk of recurrence of this tumor, low, nevertheless requires long-term follow-up.

3. Observation

A 21-year-old patient, with no history, was admitted for pain in the left hypochondrium of progressive onset, of a gravity type, without accompanying signs. The physical examination was unremarkable. The abdominal ultrasound showed a tissue and multi-cystic mass between the spleen and the tail of the pancreas. The abdominal CT had concluded in a cystic mass, well limited, finely partitioned of the tail of the pancreas, measuring 7cm (Figure 1). Surgical treatment consisted of a caudal splenopancreatomy removing the cystic mass with partial splenectomy. Morphological study and immunohistochemistry (positive antibodies are: anti-CD10, anti-vimentin, anti-NSE, anti-CD56, anti-RP and anti-synaptophysin) returned in favor of TPPSP with resection complete. The post-operative consequences were simple.

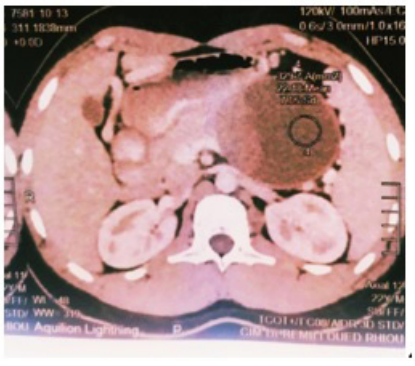

Figure 1: CT scan

4. Discussion

The solid pseudo papillary tumor of the pancreas (TPPSP) was first described by Frantz in 1959 [1]. It is a rare tumor that represents less than 2% of exocrine pancreatic tumors and less than 5% of cystic

pancreatic tumors [2]. It typically affects young women with an average age of 28 and a sex ratio of 10: 1 [3]. However, rare sporadic cases in men and the elderly have also been reported. This tumor affects the head, body or tail of the pancreas as well, with nevertheless a predominance in the corporéo-caudal region (64% of cases) [4]. Rare cases of extrapancreatic localizations are also described (1%), namely retroperitoneal, duodenal, mesocolic, hepatic [4, 5]. The circumstances of discovery are very variable and not very specific: it may be a fortuitous discovery during an imaging examination carried out for another reason, or during the appearance of a palpable abdominal mass, or even by non-specific abdominal pain. Sometimes the tumor, by increasing in size, causes signs of compression of the neighboring digestive, biliary or vascular structures [6]. Complementary examinations generally show a well-encapsulated complex mass with both solid and cystic components. The ultrasound appearance of the tumor varies depending on the size of the cystic areas. But in almost all cases, TPPSP presents as a well-defined cystic mass, with regular contours little or no vascularization, with heterogeneous content and without internal partitions [5]. Abdominal computed tomography shows a well-defined, heterogeneous, solid and cystic mass, which increases slightly or partially at the periphery after injection of the contrast product [8]. MRI is the best way to obtain information on bleeding within the lesion by multi-plane imaging [9]. It also makes it possible to highlight the fibrous capsule and to differentiate between solid and intratumoral cystic components [10]. Data from CT or MRI combined with age and gender should be sufficient to indicate surgery. The only curative treatment is surgery, in which the choice of the operative method depends on the size, the tumor location and a possible invasion of adjacent organs. It consists of a left pancreatectomy with preservation of the spleen if possible, a cephalic duodeno-pancreatectomy, a partial or even total pancreatectomy [17]. The resection should be extended in the event of invasion of neighboring organs, and any nodules of peritoneal carcinoma should be resected [20]. The existence of an invasion of the portal or mesenteric veins should not contraindicate a curative procedure, cases of portal or superior mesenteric resection having been reported with prolonged survival [5]. Associated metastatic lesions should be resected with an acceptable risk, and tumor recurrence should benefit from attempted surgical excision [8]. Lymph node dissection remains controversial. The role of adjuvant chemotherapy or radiotherapy is debatable [8]. The onset of symptoms seems to go back 4 months ago with the appearance of pain in the left hypochondrium motivating the patient to consult a complete abdominal ultrasound with an abdominal CT scan (Figure 1) which revealed a mass of 9 cm long axis which sits at the level of the pancreatic tail. a pancreatic MRI (Figure 2) was performed which targeted a solidcystic mass of the pancreatic tail with invasion of the splenic hilum. we performed a left splenopancreatectomy (Figure 3), the anapathologic piece came back in favor of a frantz tumor [11-14]. The patient has had a good postoperative follow-up. Usually, tumor cells are labeled with anti-CD 10, alpha-1-antitrypsin, vimentin, NSE, E-caderin and beta-catenin antibodies. There is also an anti-progesterone antibody labeling [15]. Positive immunostaining of tumor cells for certain endocrine markers can attest to some endocrine differentiation [16].

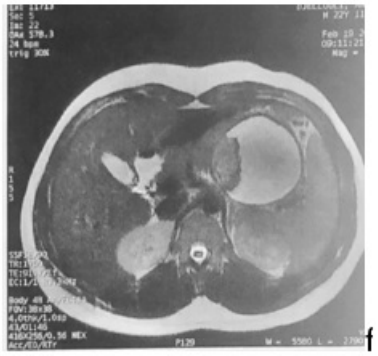

Figure 2: MRI

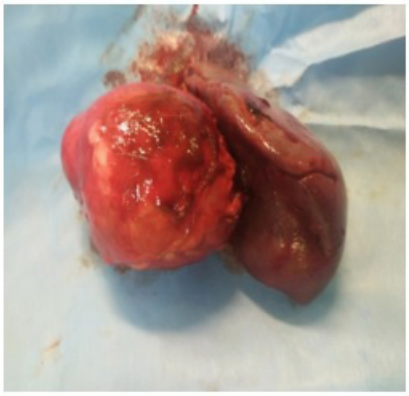

Figure 3: left splenopancreatectomy

5. Conclusion

Solid pseudo-papillary tumor of the pancreas is a rare tumor. Its diagnosis is based on radiology, in particular the abdominal CT scan and MRI coupled with the anapathology study. Surgical treatment is the treatment of choice for this kind of pathology.

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