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Case Report

Unusual cause of chronic epigastralgia in a young girl: solid pseudopapillary tumor of the pancreas

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Abstract: Solid pseudopapillary tumors of the pancreas (SPTP) are rare tumors, predominant in young women with etiopathogenesis still unclear, first described by Frantz in 1959. Their prognosis is generally good, treatment is based on surgery. Here we report the case of an 18-year-old girl with SPTP. The symptomatology was chronic evolving for 6 months made of atypical epigastralgia. The physical examination objectified a solid mass extended from the epigastrium to the left hypochondrium. The abdominal scanner showed a tissue mass at the expense of the body of the pancreas measuring 21x12x11cm. A pancreatectomy was performed. The diagnosis was confirmed by histological examination and immunohistochemistry.

Keywords: Pancreas, solid pseudopapillary tumor, Frantz, surgical resection.

INTRODUCTION

Solid pseudopapillary tumors of the pancreas (SPTP) are rare. Described by Frantz in 1959. They represent less than 2% of exocrine pancreatic tumors (Canzonieri, V. *et al.*, 2003). They mainly affect young women. Their etiopathogenesis remains uncertain. They are characterized by clinical and radiological polymorphism, most often discovered by abdominal pain, bulky mass, without alteration of the general state. Treatment is based on surgical resection, usually with a good prognosis.

The aim of this work is to report a new case and to recall the main data concerning these tumors according to the data in the literature.

CASE REPORT

We report the case of a 18 year old girl, she had no medical history, admitted for chronic epigastric pain evolving for 6 months, type of gravity, without accompanying signs. The physical examination showed a bulky mass of the epigastrium and the left hypochondrium solid and irregular. The abdominal computed tomography had concluded with a tissue mass at the expense of the body of the pancreas, with defined regular contours measuring 21x12x11cm without signs of locoregional infiltration (Figure 1).

Surgical treatment consisted of a total pancreatectomy. The operating suites were simple. The morphological and immunohistochemical histological study was in favor of a solid pseudopapillary tumor of the pancreas (positive antibodies are: AC anti CD10, AC anti chromogranin, AC anti-vimentin).

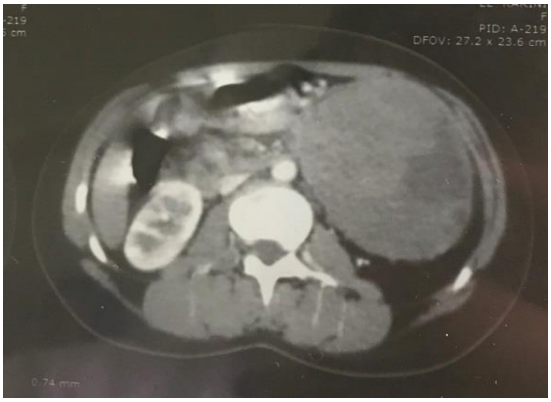

Figure 1: Scanner image showing a limited heterogeneous tissue mass of the pancreas

DISCUSSION

The pseudo-papillary and solid tumor of the pancreas was first described by Frantz in 1959. It is a rare tumor which represents less than 2% of pancreatic exocrine tumors and less than 5% of cystic tumors of the pancreas (Canzonieri, V. *et al.*, 2003). It generally affects young women with an average age of 28 years

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and a sex ratio of 10/1 (Yu, P. F. *et al.,* 2010). This tumor sits equally well in the head, body or tail of the pancreas, with a predominance in the corporeal-caudal region (64% of cases) (Denis, M. A. *et al.,* 2005). Rare cases of extrapancreatic localizations are also described (1%) namely retroperitoneal, duodenal, mesocolic, hepatic (Denis, M. A. *et al.,* 2005; & Podevin, J. *et al.,* 2003). It is characterized by clinical and radiological polymorphism, most often discovered by abdominal pain or mass, without alteration of the general state. Sometimes the tumor, increasing in size, causes signs of compression of the neighboring digestive, biliary or vascular structures (Wang, D. B. *et al.,* 2009). It can be revealed during a complication, namely a rupture or an intra-tumor hemorrhage (Sugito, K. *et al.,* 2010).

Ultrasound, CT and MRI allow the description of cystic, solid and mixed forms (Zhang, H. *et al.,* 2006; & Chen, S. *et al.,* 2007). They usually show a very limited mass, poorly vascularized and which develops preferentially in the corporocaudal region of the pancreas (64% of cases) (Denis, M. A. *et al.,* 2005). Ultrasound can show hypoechoic, homogeneous or heterogeneous images depending on the size of the cystic areas. The abdominal computed tomography shows a heterogeneous, hypodense lesion partially enhanced at the periphery (Abid, M. *et al.,* 2009; & Moholkar, S. *et al.,* 2005).

Magnetic resonance imaging is the most effective examination; it shows hyperintense hemorrhagic foci in T1 and T2, surrounded by a capsule often hypo-intense on the T2 sequences (Abid, M. *et al.,* 2009). Echo-endoscopy provides greater precision on small pancreatic lesions with a diameter less than 2 cm, which are usually undetectable by standard imaging techniques (ultrasound, CT, MRI) (Deest, G. *et al.,* 2008).

The differential diagnosis is mainly made in adults with neuroendocrine tumors and pancreatic pseudocysts (Abid, M. *et al.,* 2009).

The only curative treatment is surgical, the choice of which operating method depends on the size, the tumor location and a possible invasion of the adjacent organs. It consists of a left pancreatectomy with conservation of the spleen if possible, a cephalic duodeno-pancreatectomy, a partial or even total pancreatectomy (Abid, M. *et al.,* 2009). However, the low degree of malignancy of these tumors and the presence of a dense fibrous capsule led several surgeons to attempt simple enucleation (Papavramidis, T., & Papavramidis, S. 2005), especially in the absence of capsular invasion (Bahri, I. *et al.,* 2001). The excision must be extended in the event of invasion of the neighboring organs, and possible nodules of peritoneal carcinosis must be resected (Guedira, M. *et al.,* 2006). The existence of an invasion of the portal or mesenteric veins should not contraindicate a curative gesture, cases of portal resection or superior mesenteric having been reported with prolonged survival (Podevin, J. *et al.,* 2003).

Associated metastatic lesions must be resected with an acceptable risk, and tumor recurrences must be the subject of an attempted surgical excision (Cheng-hong, P. *et al.,* 2006). Lymph node dissection remains controversial. The place of chemotherapy or adjuvant radiotherapy is debatable (Cheng-hong, P. *et al.,* 2006). The same is true for hormone therapy, used because of the positivity of certain tumors at the progesterone receptors, but without real effectiveness (Abid, M. *et al.,* 2009). The prognosis for TPPSP is good. The recurrence rate is 10 to 15% (Guedira, M. *et al.,* 2006 ; & Cheng-hong, P. *et al.,* 2006). Cases of prolonged survival have been reported even in the presence of hepatic or peritoneal metastases or in case of incomplete surgery (Guedira, M. *et al.,* 2006).

CONCLUSION

The solid pseudopapillary tumor of the pancreas is a rare tumor, must be evoked in front of any pancreatic tumor in young women, the confirmation is histological and immunohistochemical, the treatment is purely surgical, generally of good prognosis but the rate of recurrence is not negligible.

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