Solid pseudopapillary tumor of the pancreas in a 14-year-old girl

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Tumeur solide pseudopapillaire du pancréas chez une fille de 14 ans

Case

A 14-year-old girl was admitted complaining of episodic epigastric pain, vomiting, and an abdominal mass that had been growing for a year. Clinical examination revealed a mass of the left abdominal and epigastric regions. Ultrasound (US) examination showed a tumor of the pancreatic body and isthmus region, measuring 6.8 × 6.3 cm. Laboratory findings were normal. Abdominal computed tomography (CT) (figure 1) showed a well-defined homogenous soft tissue mass at the pancreatic body. T1-weighted abdominal magnetic resonance imaging (MRI) with G-DTPA enhancement showed a tumor of the pancreatic body with homogenous low signal intensity, and the T2-weighted MRI revealed a well-defined pancreatic tumor with slightly higher signal-intensity than the pancreatic parenchyma. Before resection the tumor measured 8 cm in diameter (Figure 2) and no metastases were seen. A central pancreatic resection was performed, and the pancreatic stump was fastened to the stomach. The histological examination showed a solid pseudopapillary tumor of the pancreas (SPTP) (figure 3). No adjuvant therapy was performed. The postoperative course was uneventful, and
the follow-up one year later showed no signs of tumor recurrence or endocrine or exocrine insufficiency.

Discussion

The SPTP is a rare neoplasm [1], accounting for only 0.2-2.7% of all pancreatic tumors [2]. It is known for its low malignant potential and its indolence, both important for its good prognosis [3,4]. Recent advances in diagnostic imaging and methods, including US, CT, MRI, and fine-needle aspiration biopsies have led to good characterization of its pathological features [1]. SPTP is a rare tumor that occurs preferentially in young women (mean age of 25 to 30 years) [1–3]. Neither its etiology nor its exact pathogenesis is known, but its striking predilection for women suggests a hormonal influence. Clinically, the abdominal mass may cause discomfort or pain, or it may be found incidentally on routine physical examination [2,3]. US reveals a well encapsulated tumor [1,2,4], cystic and solid masses, and sometimes a pure-solid mass or calcifications. CT may help to differentiate SPTP from other cystic neoplasms, whereas MRI may help to differentiate it from islet cell tumors. MRI may also be superior to CT in the correlation between radiological and clinicopathological findings.

Fine-needle aspiration may play an important role in preoperative planning by helping to distinguish SPTPs from other pancreatic lesions with a significantly different prognosis and treatment [1,5].

This tumor is typically localized on the body or tail of the pancreas [1,3]. Macroscopically, it is solid and well encapsulated. Histologically, the tumor is typically solid and pseudopapillary with intensive vascularization and cellular degeneration. Expression of tumor markers is highly variable, and most SPTPs are immunoreactive for vimentin, α 1-antitrypsin, and α 1-antichymotrypsin [6]. Its prognosis is almost uniformly good, as more than 95% of patients with SPTP are cured after complete resection of the tumor [1,2,3]. The treatment is based on complete resection with preservation of as much pancreatic tissue as possible. Resection techniques do not appear to influence survival. Up to now there is no clear evidence that chemo- or radiotherapy is indicated [4].

In conclusion, SPTP is a rare tumor with a favorable prognosis. Clinical correlations, radiological findings and histologic features together make accurate diagnosis of this tumor possible.

Conflicts of interests : none
References


