Letters to the Editor

Solid pseudopapillary neoplasm of the pancreas or Frantz's tumour: Report of two cases with different locations

Key words: Pancreas tumour. Solid pseudopapillary tumour of the pancreas. Diagnosis. Treatment.

Dear Editor,

Solid pseudopapillary tumour of the pancreas (SPTP) was described by Frantz in 1959 (1). It is an uncommon neoplasm with an incidence estimated at between 0.2 and 2.7 % of all pancreatic tumours, although it may account for 50% in paediatric patients.

Case reports

We report 2 cases in women aged 30 and 27 years, who are currently under follow-up. Both presented with non-specific clinical features such as epigastric pain, vomiting, diarrhoea, fever or weight loss over the previous months. Ultrasound disclosed heterogeneous masses in the tail and head of the pancreas, respectively. The study was completed in both women with computed tomography and magnetic resonance, which revealed heterogeneous lesions with solid and cystic components. They underwent surgery with a preoperative suspicion of SPTP: Distal splenopancreatectomy in the patient with the tumour in the tail of the pancreas and cephalic pancreatoduodenectomy with pylorus conservation in the woman with the tumour in the head of the pancreas. In both women, the pathological anatomy and immunohistochemistry, with positivity for CD10, CD56, vimentin, neuronal enolase and progesterone

receptors, and mild positivity to chromogranin, established a final diagnosis of SPTP.

SPTP occurs 90-95 % of the time in young women in their 3rd decade of life (2) and has a preference for Asians (3). The most common location is the body and tail and on rare occasions it is found in the retroperitoneum, ectopic pancreatic tissue, mesentery or liver (4). Its growth is slow, which is why patients are asymptomatic (5) or have non-specific clinical features and why diagnosis is usually casual in 15 % of cases (6). Imaging tests are usually typical: well-delimited heterogeneous mass due to its haemorrhagic necrotic solid and cystic components (7). Fine-needle puncture-aspiration is controversial, as there is no clear efficiency for differential diagnosis and it may be confused with a pseudocyst when only necrotic material is collected. The origin of SPTP is still uncertain, although a theory gaining strength is that it originates in pluripotential pancreatic cells which would be favoured by genetic factors, and hormone stimuli due to a greater frequency in women and the presence of hormone receptors (8). Immunohistochemistry usually shows a positive reaction

Fig. 1. Abdominal computed tomography showing abdominal mass in tail of pancreas measuring 16 cm maximum with solid and cystic areas.

to neuron-specific enolase, vimentin, alpha1-antitrypsin, somatostatin, glucagon or insulin.

Depending on the location of the SPTP a type of surgery must be performed (distal or cephalic excision) with complete resection in order to obtain a good prognosis and a low rate of local recurrence and metastases; the 10-year survival rate is good and a long-term follow-up is recommended (9). When the lesion is superficial or complete resection is not possible, an enucleation or partial resection can be performed with acceptable outcomes. No clear indication for treatment with radiation therapy or chemotherapy has been shown. Metastases are found at the time of diagnosis in 10-15 % of patients, especially in the liver and usually as the only metastasis (10). In these cases of local recurrence or metastases, the surgical approach may not be recommended due to the long survival rate.

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