E-QUID: ANSWER / Gastrointestinal imaging

Solid pseudopapillary neoplasm of the pancreas

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Case report

Ms. K., a 37-year-old patient, consulted the emergency ward for abdominal pain predominantly in the hypochondrium and right iliac fossa. This patient presented a past history of anal sphincter plasty for incontinence following a traumatic childbirth. The clinical examination found a fever at 38.8 °C and diffuse abdominal pain. The lab tests upon admission detected an inflammatory syndrome (leukocytes: 18,000/mm³ and CRP: 150 mg/l) without cytolysis or cholestasis. The urine strip was negative. In view of the intensity of the pain, an abdominal-pelvic CT-scan with injection was carried out (Fig. 1) to search for acute appendicitis or acute cholecystitis. Acute cholecystitis was diagnosed in view of the distension and vesicular parietal thickening. By chance, a tumour of the tip of the cauda pancreatis was discovered and a complementary assessment by MRI was carried out (Fig. 2).
Figure 1. Abdominal-pelvic CT in axial sections: a: axial section in spontaneous contrast; b: axial section injected at portal time.

Figure 2. Pancreatic MRI in axial sections: a: sequence T2; b: sequence T1 after fat saturation; c: sequence T1 EG dynamic after injection at arterial time; d: sequence T1 EG dynamic after injection at portal time.
What is your diagnosis?

After reading the case report, what diagnosis would you choose from the following proposals:
- mucinous cystadenoma;
- pancreaticoblastoma;
- neuroendocrine pancreatic tumour;
- solid pseudopapillary neoplasm of the pancreas;
- mucinous cystadenocarcinoma.

Diagnosis

Solid pseudopapillary neoplasm of the pancreas.

Comments

The injected abdominal-pelvic CT-scan (Fig. 3a and b) reveals a well-defined tumoral mass developed at the expense of the cauda pancreatis. This oblong lesion measures 44 × 30 mm and presents a punctiform calcification with spontaneous contrast (Fig. 3a). The enhancement is heterogeneous associating progressive enhancement of the tissue components and more hypodense, non-enhanced areas corresponding to cystic zones. The pancreatic MRI (Fig. 4a–d) detected this tumoral mass of the cauda pancreatis, measuring 4 × 3 cm in the transverse plane. This lesion presents a heterogeneous T2 hypersignal (Fig. 4a), a global T1 hyposignal with the presence of two spots in spontaneous hypersignal of haemorrhagic appearance (Fig. 4b). The enhancement is heterogeneous, gradual and more marked late (Fig. 4c and d). There are cystic zones of necrotic appearance. There is no peripheral capsule. In view of these characteristics and considering the sex and age of our patient, a solid pseudopapillary neoplasm was diagnosed.

The diagnosis of mucinous cystadenoma is eliminated in view of the presence of an enhanced tissue component and a heterogeneous signal on sequences T1 and T2. The differential diagnosis with mucinous cystadenocarcinoma is difficult. It generally consists of an aggressive tumour that invades the pancreatic parenchyma while the solid pseudopapillary neoplasm pushes back the adjacent structures. In addition, the mucinous cystadenocarcinoma does not generally present intra-lesional haemorrhagic reorganisation. A pancreaticoblastoma is an extremely rare malignant tumour and is specifically paediatric. Jaundice and dilation of the bile ducts are very often associated. The diagnosis of neuroendocrine tumour of the pancreas is eliminated in view of the absence of a greatly enhanced shell around the lesion.

The patient benefited from a cholecystectomy and a caudal pancreatectomy by coelioscopy with sacrifice of the splenic vessels without splenectomy. The extemporaneous anatomo-pathological analysis confirmed the diagnosis and revealed healthy surgical margins. On D2 after the operation, a CT-scan was carried out in view of the appearance of fever associated with epigastric pain and left hypochondrium. The CT-scan revealed infiltration of the pancreatic region and splenic infarction of about 15% of the volume of the parenchyma. The symptoms quickly disappeared and no other post-surgical complications were noted. The diagnosis of solid pseudopapillary neoplasm of the pancreas was definitively confirmed by the histology and immunohistochemistry.

Discussion

Formerly known by different names: Frantz tumour, solid and cystic epithelial neoplasm of the pancreas, papillary and cystic neoplasm of the pancreas, in 1996, the WHO renamed this lesion solid pseudopapillary neoplasm of the pancreas [1]. This is a rare tumour (about 1% of tumours of the exocrine pancreas) that presents a low potential of malignity and can benefit from surgical treatment [2]. This tumour occurs mainly in young (mean age: 24 years) women (90%) [3]. Clinically, more than 50% of the cases involve an

Figure 3. Abdominal-pelvic CT in axial sections: a: axial section in spontaneous contrast: well-defined tumour of the cauda pancreatic with a central punctiform calcification (arrow); b: axial section in portal time: heterogeneous lesion enhancement with hypodense cystic area in the periphery (tip of arrow).
asymptomatic tumour that is discovered by chance [2]. The most common symptoms are abdominal pain, nausea and vomiting, loss of weight and a heavy sensation or weight on the stomach [2,3]. More rarely, it’s possible to observe intestinal occlusion, anaemia, jaundice, pancreatitis, an obstruction of the bile ducts or a traumatic rupture with haemoperitoneum [2]. In the child, the most common symptom is a palpable abdominal mass with a significantly higher tumour size (8 cm versus 6 cm in the adult) [2]. In men, the mean age of the diagnosis is significantly higher (37 years versus 26 years in women) [3]. There is no difference between the sexes as regards the size of the tumour, the clinical symptoms, the imaging characteristics and the location of the tumour [3].

In the present case, it was discovered by chance during an exploration carried out for an acute abdominal syndrome with acute cholecystitis.

Histologically, macroscopically, the lesion is separated from the healthy parenchyma by a thin fibrous capsule. Microscopically, the cystic zones are surrounded by solid tumoral tissue presenting characteristic pseudopapillary structures (Fig. 5a–c). With the progression of the tumour, necrotic and haemorrhagic zones develop. The incidence of a malignant transformation of these tumours is about 15% and occurs either by local invasion or by metastatic extension at a distance [2].

On the immunohistochemical level, the tumoral cells express a great many immunomarkers at their surface, both those of exocrine tumours (α1-antitrypsin, α1-antichymotrypsin...) as well as those of endocrine pancreatic tumours (neurone-specific enolase and synaptophysine). A recent hypothesis mentions that solid pseudopapillary neoplasms arise from undifferentiated multipotent stem cells [2].

In the imaging, the sonogram may reveal a well-defined, rounded, oblong or polylobate, encapsulated, heterogeneous mass with an echogenic tissue component and an anechoic cystic component. It may reveal peripheral calcifications and posterior reinforcement. A mass effect may exist on the adjacent structures [2]. In the CT-scan, the mass appears to be heterogeneous and hypodense. It is possible to search for calcifications. After injection, there is a peripheral capsular enhancement. The limits of the CT-scan remain the lack of tissue resolution [2]. The pancreatic MRI

![Figure 4](image-url)

**Figure 4.** Pancreatic MRI revealing a caudal pancreatic tumour: a: in hypersignal in T2 weighting with fat saturation; b: hyposignal T1 with punctiform centro-lesional haemorrhagic hypersignals (arrows); c, d: the EG T1 VIBE sequences after dynamic injection of gadolinium revealing, at arterial time (c) peripheral enhancement late (d) a tendency for homogenisation of the enhancement of the tissue components.
is currently the choice non-invasive diagnostic tool for cystic masses of the pancreas. The MRI reveals the cystic components, the intra-lesional haemorrhagic rearrangements and the presence of a capsule, in the form of a hypodense halo on weighted T2 sequences, characteristic of solid pseudopapillary neoplasms of the pancreas [4,5]. In addition, the lesion is generally well-defined, voluminous, of mixed tissue and cystic content, the origin of a heterogeneous signal in hyper or hyposignal T1 and a heterogeneous signal rather in hypersignal T2 [4]. Nevertheless, it should be noted that the concept of hypersignal T1 in the cystic zones, classically attributed to haemorrhagic phenomena, is only the expression of an increase in the protein concentration that is not synonymous with the heme of haemoglobin, a myxoid transformation that may provide the same images. After the injection of Gadolinium, there is peripheral enhancement with progressive and heterogeneous lesion filling [1,4]. The preferential location is the cauda pancreas. The lesion pushes back the adjacent structures without invading them [2]. Canal dilation upstream may be present [2,6]. The presence of these signs in imaging indicates the diagnosis in a young woman since 80% of all solid pseudopapillary neoplasm of the pancreas present typical characteristics in imaging (TDM and/or MRI) [3]. Recent studies have shown that over 50% of the lesions were diagnosed before surgery [2].

The problem of the differential diagnosis is above all raised with neuroendocrine tumours of the pancreas and mucinous cystadenocarcinomas where the prognosis is less favourable [1]. The cystic forms of neuroendocrine tumours present a localised or circumferential hypervascular thickening of the wall after injection [5]. They affect older adults (mean age: 53 years), without preference as to sex, often in a context of multiple endocrine neoplasma [1]. The presence of enhanced tissue elements associated with cystic lesions is a criterion in favour of mucinous cystadenocarcinoma [1]. In the child, pancreaticoblastoma is also a differential diagnosis. This is an extremely rare malignant tumour, sometimes containing cystic zones [7]. The diagnosis with certainty is based on the histology. All of these differential diagnoses are an indication for surgical treatment. It is therefore useless to carry out a puncture-biopsy as this may favour cell dissemination, the cause of a local recurrence of metastatic diffusion [8].

Surgical treatment provides a cure in over 95% of the cases with full resection and microscopically healthy mar-

**Figure 5.** Pathological anatomy: a: macroscopy: solid well-defined nodular lesion presenting cystic components with haemorrhagic content; b: photomicrography (hematoxyline-eosine-safran × 12): revealing a heterogeneous, partially encapsulated tumour, presenting cell areas and cystic zones with haemorrhagic content; c: photomicrography (hematoxyline-eosine-safran × 100) revealing pseudopapillary structures (asterisk).
gins. As for tumours of the body of the pancreas or cauda pancreatis, the most common, a distal pancreatectomy with preservation of the spleen is recommended. For tumours of the head of the pancreas, a cephalic duodeno-pancreatectomy with preservation of the pylorus may be carried out. For tumours of the isthmus, a central pancreatectomy with pancreaticojejunostomy or pancreatico-gastric anastomosis is recommended. For small tumours, far from the main pancreatic duct, tumour enucleation may be considered. Lymphadenectomy is not recommended since lymph node metastases are extremely rare. In a study on 292 patients, Mao et al. found that only five (2%) of the patients presented lymph node invasion [9]. This data is confirmed by more recent studies that do not note any lymph node metastasis in all of the patients included [3, 10]. The surgical resection syndrome of metastases is justified by the excellent long-term prognosis even in the presence of secondary locations [2]. In men, the tumour is more aggressive with a higher incidence of invasion of the portal vein that requires more radical and less conservative surgical resection [3]. Globally, the rate of morbidity remains close to 60% with, in particular, the occurrence of post-surgical pancreatic fistulae, infectious complications and intestinal occlusion [3].

The place of adjuvant or neo-adjuvant treatments has not been defined. The rate of recurrence is estimated at between 10 and 15% after resection over a mean follow-up period of about 7 years. It is therefore necessary to monitor these patients over a relative long period of time [2]. In men, close and prolonged monitoring is recommended [3]. The rate of mortality is estimated at 2% [3].

Conclusion

In conclusion, solid pseudopapillary neoplasms of the pancreas mainly affect young women. Abdominal pain is the most common symptom although the chance discovery by imaging accounts for over half of all cases. Due to the excellent tissue resolution, the pancreatic MRI is the best imaging tool to characterise cystic tumours of the pancreas. The diagnosis of solid pseudopapillary neoplasm of the pancreas will be based on the MRI in view of the presence of an encapsulated tissue lesion comprising cystic zones and intra-tumoral haemorrhagic rearrangement. It is a tumour with a low malignancy potential, a good prognosis after surgical treatment although it requires monitoring due to the potential recurrence.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References