CASE REPORT

Intrapancreatic accessory spleen: a misleading diagnosis

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SUMMARY

Intrapancreatic accessory spleens are congenital malformations that occur in roughly 2% of individuals. Most of them are innocent until found. Lately, there have been a few case reports of intrapancreatic spleen misdiagnosis leading to unnecessary pancreatic surgery. We report the case of a 64-year-old woman who had a hypervascular pancreatic nodule diagnosed on dynamic CT and MRI after an episode of acute pancreatitis. The patient's progress was followed for 18 months, repeated the CT and MRI examinations and an endoscopic ultrasonography with fine needle aspiration was performed. Neoplastic cells were not identified on cytology. Despite the stability of the lesion, a distal pancreatectomy was performed to exclude a neuroendocrinal tumour. An intrapancreatic accessory spleen was ultimately identified on histological examination.

BACKGROUND

Intrapancreatic accessory spleen is a common congenital defect, with a prevalence of 2% found in an autopsy series of 3000 people.¹ It is usually an asymptomatic and benign lesion but it mimics pancreatic malignant tumours. For this reason, it can be a cause of unnecessary surgery as has been recently reported.²⁻³

It is of the utmost importance to be aware of this entity and to know that it is not only frequent but also possible to diagnose by dynamic CT or MRI, without further invasive procedures.

CASE PRESENTATION

A woman, 64 years old, with a history of hydatid cysts in the liver and lung for which she had been previously submitted to surgery and a parathyroid adenoma already removed by parathyroidectomy.

She had mild acute hepatitis 2 years ago and imaging examinations were performed for routine evaluation. A mass in the tail of the pancreas was incidentally discovered.

INVESTIGATIONS

CT documented a well-defined nodular lesion, with 1 cm of diameter, on the tail of the pancreas, showing contrast enhancement in the arterial phase (figure 1), and isodense in the venous and delayed phases. The hypervascular features raised the diagnostic hypothesis of a neuroendocrinal tumour of the tail of the pancreas. The MRI (figure 2) showed a solid lesion, with low signal intensity on T1 and high signal intensity on T2-weighted images, with persisting enhancement on the late phases of dynamic MRI study.

To help in the characterisation of this mass, an endoscopic ultrasound (figure 3) with fine needle aspiration (EUS-FNA) biopsy was performed. The cytology and immunohistochemical panel did not show neoplastic cells.

The patient had normal glycaemia, Ca 19.9 (cancer antigen 19.9), CEA (carcinoembryonic antigen) and chromogranin levels.

DIFFERENTIAL DIAGNOSIS

Imaging findings of a solid hypervascular mass on the pancreatic tail, in a contrast-enhanced CT, may suggest several diagnoses like neuroendocrinal tumours, solid and papillary epithelial neoplasm, pancreatic adenocarcinoma and even metastases.

One must have a high degree of suspicion to think of an intrapancreatic accessory spleen (IPAS) and rely on spleen-like dynamic behaviour on CT or MRI in order to have certainty in the diagnosis and obviate surgery.

In this case, the diagnosis was not considered unequivocal. And most of all, after several procedures and concerns, the patient wanted to be reassured that she had no serious condition and desired a definitive solution.

TREATMENT

Despite the lesion being stable for 18 months, the patient underwent a distal pancreatectomy to exclude malignancy.

OUTCOME AND FOLLOW-UP

Histological examination of the surgical specimen described a brown mass, capsulated, with 12 mm (figure 4), compatible with an accessory spleen.

Figure 1 Axial contrast-enhanced CT, in the arterial phase, shows a solid hypervascular nodule (yellow arrow) in the tail of the pancreas.

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Figure 2 (A) Axial T2-weighted MRI (and B, with fat suppression) demonstrates a mild-to-intermediate hyperintense nodule in the pancreatic tail, with signal intensity similar to that of the spleen (white arrowhead). (C) In transverse T1-weighted MRI (and D, with fat suppression) the nodule (yellow arrow) is hypointense, similar to the adjacent spleen (white arrowhead); the surrounding pancreas (orange arrow) is hyperintense.

The patient recovered from surgery without complications and stays symptomatic, as before.

DISCUSSION

Accessory spleen, also referred to as splenunculi, consists of a common congenital defect, seen in 10–30% of patients, in autopsy.^{4–6} It results from a failure in the fusion of mesenchymal cells that form the splenic anlage.^{5,7}

Around 80% of the accessory spleens are located in the splenic hilum and 17% on the pancreatic tail.¹ Most of these lesions are asymptomatic.⁸

In the majority of the cases described in the literature, the finding of an IPAS was incidental during an unrelated imaging

study and most of them were diagnosed only after surgery, which is performed for suspected pancreatic neoplasia.

An accessory spleen usually poses no clinical problems and no treatment is necessary unless in the case of torsion, haemorrhage, cyst formation or in the presence of haematological disorders, such as idiopathic thrombocytopenic purpura.

It is crucial to make the diagnosis of IPAS non-invasively, in order to avoid unnecessary surgery and eventually reduce possible patient morbidity and mortality.^{9,10}

Intrapancreatic accessory spleens are easily misdiagnosed as neuroendocrine tumours because of their hypervascular appearance, but besides its location on the tail of the pancreas, the key to suspect of IPAS is similar appearance of the lesion to the spleen in different imaging modalities, including ultrasound, CT and MRI on all contrast-enhanced phases.⁹

The diagnosis can be confirmed using superparamagnetic iron oxide (SPIO)-enhanced MRI, Levovist (Bayer, Berlin, Germany)-enhanced US and scintigraphy using Tc-99m-labelled sulfur colloid or Tc-99m-labelled heat-damaged red blood cells. The mechanism of these different techniques is essentially the same: trapping of the contrast material by reticuloendothelial cells.

Scintigraphic techniques are the most specific imaging methods for diagnosing ectopic splenic tissue; however, they offer far inferior anatomic resolution to CT or MRI.^{11,12}

EUS-FNA may reveal a predominance of small lymphocytes with a subset of histiocytes, conspicuous eosinophil and plasma cells and CD8-positive immunostaining of endothelial cells in cellblock sections.¹²

Thus, it is important to consider the possibility of an accessory spleen when an asymptomatic intrapancreatic mass is incidentally visualised on examination, and the imaging characteristics of the lesion match those of the spleen.

Figure 3 Endoscopic ultrasound shows a 12 mm, oval-shaped, homogeneous, hypoechoic lesion (yellow arrow) with well-defined borders, located in the pancreas.

Figure 4 Histopathology of the surgical specimen confirming intrapancreatic accessory spleen. (A) H&E stain of cell block showing pancreatic acinar tissue (white arrow) with splenic tissue in the middle (yellow arrow). (B) H&E stain of intrapancreatic accessory spleen showing normal red and white pulp.

Learning points

- ▶ Intrapancreatic accessory spleen is a common congenital defect (2%).
- ▶ It is usually asymptomatic and benign.
- ▶ It is a hypervascular nodule that can mimic a pancreatic tumour.
- ▶ It may be safely diagnosed with careful radiological investigation avoiding unnecessary surgery.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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