CASE REPORT

Intrapancreatic accessory spleen: An eluding diagnosis

Teoh Keat How, MRCS[1], Balraj Singh, MS[1], Navarasi S Raja Gopal, MPath[2]

[1]Department of General Surgery, Penang General Hospital, Penang, Malaysia, [2]Department of Pathology, Penang General Hospital, Penang, Malaysia.

SUMMARY

Intrapancreatic accessory spleen (IPAS) is a benign anomaly of splenic embryology and a rare cause of pancreatic pseudotumour. Here, we report a case of a 70-year-old Malay lady whose IPAS was discovered incidentally during her surveillance computed tomography for her underlying left lower lung fibrosis. Radiologically, the lesion mimicked a neuroendocrine pancreatic tumour and was only diagnosed pathologically as IPAS after surgery. In conclusion, recognising IPAS as a differential for enhancing pancreatic mass allows us to exhaust all non-invasive diagnostic means to diagnose this benign lesion. It will allow the patient to avoid unnecessary surgery and its accompanying complications.

KEY WORDS:
Intrapancreatic; Accessory; Spleen; Pancreas; Surgery

INTRODUCTION

Intrapancreatic accessory spleen (IPAS) is a benign lesion due to an anomaly of splenic embryology and a rare cause of pancreatic pseudotumour.[1,2] Accessory spleen occurs when there is failure in the fusion of the splenic anlage during embryology at the fifth week of foetal life and consists of structurally normal splenic tissue.[3] It is reported in up to 10% of the general population with over 80% of these accessory spleen commonly found in the region of the splenic hilum and vascular pedical.[4] IPAS accounts for almost 20% of all accessory spleen and occurs most frequently in the tail of the pancreas.[1] Here, we report a case of IPAS that mimicked a neuroendocrine pancreatic tumour radiologically and was only diagnosed pathologically after surgery.

CASE REPORT

We report on a 70-year-old Malay lady, with underlying hypertension, bronchial asthma, lung fibrosis and ischemic heart disease, who was referred to us for a solitary mass found at her pancreatic tail during surveillance contrast enhanced computed tomography of her thorax. Her pancreatic protocol imaging confirmed an avidly enhancing lesion at the tail of the pancreas during the early arterial phase, measuring 1.3 x 1.0 cm in size (Figure 1).

She complained of occasional epigastric pain for the past two to three years but denied having constitutional symptoms, jaundice and other associated symptoms. She has no family history of malignancy.

Clinically, she was in good nutritional condition and there was no hepatosplenomegaly or ascites on examination. Her serum carcinoembryonic antigen and CA19-9 were normal at 2.8 µg/L and 6 U/ml respectively. Other blood investigations were unremarkable.

Intraoperatively, a firm distal pancreatic lesion was noted. It was free from the splenic vessels and no lymph nodes were palpable. There was no ascites and the liver and spleen appeared normal. A spleen-preserving distal pancreatectomy was performed with a 1 cm margin.

The pathological examination revealed an encapsulated brownish mass (1.5 x 0.8 x 0.8 cm) with surrounding pancreatic tissues. Microscopically, it was formed by red and white pulps, recapitulating splenic tissue (Figure 2). The immunohistochemical stains showed positive CD45 expression and negative for cytokeratin and neuroendocrine markers (Figure 2). No apparent neoplastic components were found. Thus, IPAS was confirmed as the pathologic diagnosis. The patient's postoperative course was complicated by the development of a pancreatic leak. However, she recovered well after one and a half months of conservative treatment.

DISCUSSION

Generally, IPAS is clinically silent and is often discovered incidentally during investigations done for upper gastrointestinal symptoms.[2] Radiologically, IPAS appears similar to a hypervascular pancreatic tumour like acinar cell carcinoma and neuroendocrine tumour. Such a misdiagnosis, like in our patient, will translate into unnecessary surgical intervention before the correct diagnosis is made.[3,4] Hence, it is important to diagnose IPAS via non-invasive methods as IPAS usually does not pose any clinical threat. The only indications for surgery are when the diagnosis is unclear and is misdiagnosed as malignancy; symptomatic due to torsion, infarct, spontaneous rupture, haemorrhage and cyst formation; and when all functional splenic tissue should be removed for the treatment of haematologic disorders like idiopathic thrombocytopenic purpura.[1,3]

Although there are no clinical or radiographic criteria for the diagnosis of IPAS, there are a few characteristic findings that can help elucidate the diagnosis.[1,3,4] On ultrasonography, IPAS appears as a round, solid, homogenous and hypoechoic mass within the pancreas.[1,3] It also shows posterior acoustic enhancement due to its fibrous capsule.[2,4] Subramanyam et

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Corresponding Author: Teoh Keat How, Department of General Surgery (006), Penang General Hospital, Jalan Residensi, 10990 Georgetown, Penang, Malaysia Email: kito.tkh@gmail.com

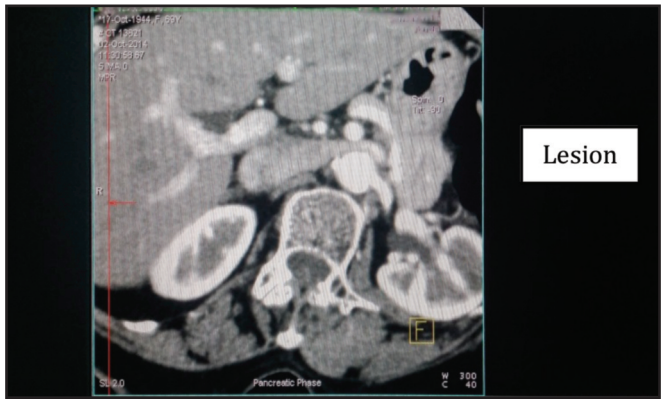

Fig. 1: An avidly enhancing lesion seen at the tail of the pancreas during early arterial phase.

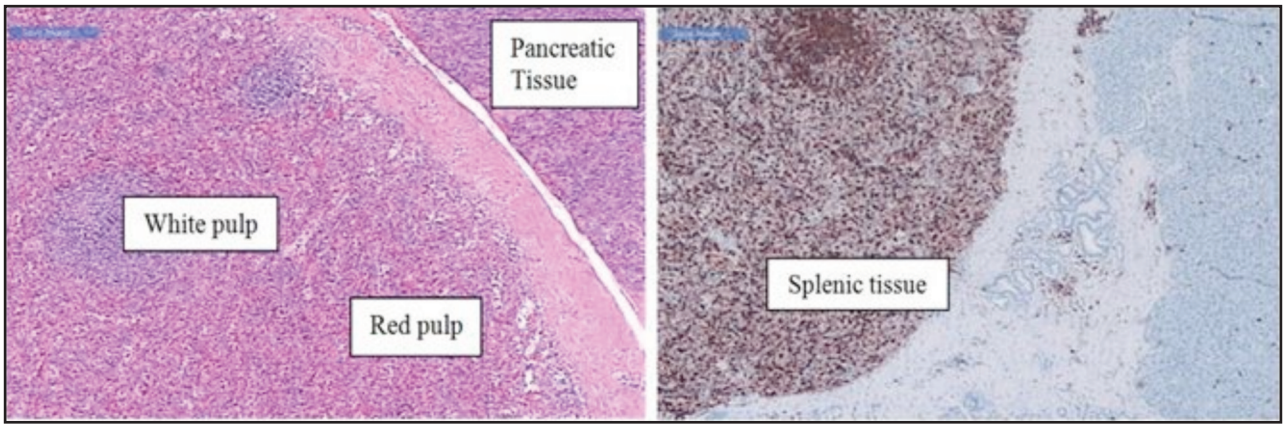

Fig. 2: H&E stained section (left) shows the encapsulated lesion is showing the presence of red pulp and white pulp, in keeping with normal histology of the spleen. The splenic tissue (right) shows strong and diffuse immunoreactivity towards CD45 (Leukocyte Common Antigen), confirming its haematolymphoid derivation. In contrast, the adjacent pancreatic parenchyma shows negative staining.

al. proved that when a vascular pedicle is demonstrated via Doppler ultrasound originating from the splenic vessels entering the lesion, it has a sensitivity of nearly 90% in diagnosing accessory spleen.[5]

On computer tomography (CT), IPAS appears as a well-circumscribed mass with an arciform pattern of enhancement due to varying flow rates of contrast through the red and white pulp.[1,3] However, in our patient's CT, the lesion was only described as an avidly enhancing lesion at the tail of pancreas. Whereas for magnetic resonance imaging (MRI), IPAS is shown as a low signal intensity mass on T1-weighted images and a high-signal intensity mass on T2-weighted images.[1,3]

Recently, the liberal use of CT for diagnosis and screening has increased the detection of asymptomatic pancreatic neoplasms. With the majority (60%-75%) of these incidentalomas been reported as malignant or premalignant lesions, other imaging techniques like 99mTechnetium heat-damaged red blood cells (99mTc-HDRBC) scintigraphy and superparamagnetic iron-oxide (SPIO)-enhanced MRI are needed to differentiate IPAS from malignancy.

99mTc-HDRBC scintigraphy is highly sensitive and specific in detecting functional splenic tissue as up to 90% of 99mTc-HDRBC is sequestered in it. However, visualisation can still be difficult when there is only minimal functioning splenic tissue and its spatial resolution is still inferior compared to CT or MRI.[3] Therefore, 99mTc-HDRBC scintigraphy is ideally used in conjunction with other cross-sectional imaging techniques.

SPIO-enhanced MRI offers higher spatial resolution.[3] Its principle is similar to 99mTc-HDRBC scintigraphy but it targets the reticuloendothelial system cells.[3,4] With its high tissue specificity for reticuloendothelial system, it significantly decreases MRI signal intensity for splenic tissue, but not for tumours.[3,4] However, in our setting, these two imaging modalities were not available. Hence, the decision was made to proceed with distal pancreatectomy based on her CT findings.

Endoscopic ultrasonography guided fine needle aspiration (EUS-FNA) biopsy is also an alternative. The predominant cytological features suggestive of IPAS are polymorphous mixed lymphoid populations, traversing thin-walled

vascular structures and positive CD8 immunostaining in cell block sections.[2,4] CD8 immunocytochemical staining is characteristic of endothelial cells present in splenic sinuses as it does not stain systemic endothelial cells and haemangioma.[2,4] However, there are cases of false positive EUS-FNA, where IPAS were mistakenly diagnosed as pancreatic neuroendocrine tumour.[4] Hence, in order to reduce diagnostic error, EUS-FNA should be performed by an experienced endoscopist.

CONCLUSION

IPAS is a challenging diagnosis to make but is being detected more often now with the advancement of medical imaging. Recognising IPAS as a differential diagnosis for enhancing pancreatic mass allows us to exhaust all non-invasive diagnostic means to avoid surgical intervention and its associated complications. However, if the diagnosis is in doubt, surgical intervention to achieve definitive diagnosis is still unavoidable.

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