Endoscopic ultrasonography allowing expectant management of pancreatic haemangioma

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ABSTRACT

Pancreatic haemangiomas are rare benign tumours that can affect both adults and children. They have an unknown incidence and only 15 adult cases have been reported, all from histological examination. Patients present with vague symptoms relating to tumour mass or they are detected incidentally. Cross-sectional imaging is the mainstay of investigation and may reveal arterially enhancing cystic lesions but in the case presented here, it was non-diagnostic. The use of endoscopic ultrasonography confirmed the nature of the benign lesion, allowing a conservative approach as opposed to operative resection.

KEYWORDS

Pancreatic benign lesion – Pancreatic haemangioma – Endoscopic ultrasonography

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Case History

A 78-year-old woman presented with a 3-month history of increasing epigastric pain that was not associated with food and persisted despite proton pump inhibition. She denied any other associated features and had not lost any weight. There were no features of endocrine or exocrine pancreatic insufficiency and no symptoms of obstructive jaundice. Her past medical history included an appendicectomy, a laparoscopic cholecystectomy and ovarian cyst excision. She was a non-smoker who drank occasionally. Abdominal examination was unremarkable apart from some mild epigastric tenderness. She was investigated initially with a gastroscopy, which noted a hiatal hernia, and abdominal ultrasonography. Ultrasonography revealed a possible mass in the pancreatic head and urgent computed tomography (CT) was organised prior to review in the hepatopancreatobiliary multidisciplinary team (MDT) meeting.

CT revealed a 4cm soft tissue mass sitting in the posterior pancreatic head and uncinate process (Fig 1), with long areas of contact with both the portal and superior mesenteric veins. No evidence of extrapancreatic disease was noted. Interestingly, there was no dilation of either the common bile duct or pancreatic duct, suggesting the lesion could represent a neuroendocrine tumour as opposed to an adenocarcinoma. An incidental right renal cyst was noted.

Urgent endoscopic ultrasonography (EUS) and a gut hormone profile were requested. EUS failed to delineate any specific pancreatic mass but a subtle hypoechoic area in the pancreatic head was detected. It confirmed the lack of ductal involvement but did show tiny areas of vessels

with clear flow on Doppler assessment, consistent with a haemangioma. Fine needle aspiration was cautiously performed but this was non-diagnostic from a cytological viewpoint. The gut hormone profile was unremarkable as were subsequent tumour markers and the autoimmune profile, essentially ruling out any previous episodes of pancreatitis.

The patient was discussed again at the hepatopancreatobiliary MDT meeting although a proximal pancreaticoduodenectomy would have been feasible to excise the mass. However, given the non-dilated bile and pancreatic ducts, this would potentially be associated with a higher risk of pancreatic leak and increased morbidity. Repeat CT was requested and the possibility of a laparotomy with taking intraoperative frozen sections of the lesion was mooted to obtain diagnoses prior to resection. Repeat CT at four months and ten months after her original imaging was unchanged, and it was felt that the lesion was highly likely to be a benign haemangioma and that she should therefore be kept under surveillance. Clinical outpatient review at 12 months revealed that she remained well and asymptomatic, with her pain controlled with simple analgesia.

Discussion

Pancreatic haemangiomas are rare vascular neoplasms with an unknown incidence;^{1–4} collectively, vascular neoplasms (including haemangiomas, lymphangiomas, haemolymphangiomas, haemangiopericytomas, haemangioblastomas and haemangiosarcomas) account for 0.1% of all pancreatic

Figure 1 Sagittal and coronal computed tomography showing a large 4cm slightly hypoattenuating mass in the pancreatic head (arrow). There is no arterial enhancement of the lesion or any ductal dilation. Incidental right renal cyst noted.

tumours.⁵ Both adult and paediatric cases have been reported, and they are thought to be more common in infants, with a possible association with Kasabach–Merritt syndrome (haemangioma with thrombocytopenia).^{2,5} Less than 15 adult cases have been described in the literature, all from pathological specimens. There is a female predominance, with age ranges from the third to the seventh decade.^{1–5} Most patients describe non-specific abdominal pain, which may be associated with the presence of an abdominal mass. Additional signs and symptoms include obstructive jaundice, nausea and gastrointestinal bleeding although some lesions have been detected incidentally.^{3,4}

Radiology provides the mainstay of diagnosis, and ultrasonography, CT, angiography and magnetic resonance imaging (MRI) can all be used to assist in diagnosis.^{3,6} Typically, haemangiomas are cystic lesions that enhance strongly in the arterial phase of conventional CT and angiography.^{2,7} However, they are associated with neovascularisation and reduced arterial enhancement may therefore be seen owing to arteriovenous shunting with slow flow.^{3,4,6,7} The vascularity of the tumour can be assessed by comparing the amount of cystic to solid component within it on CT; in addition, MRI may show if blood is the main component of the tumour on a T2 weighted image.⁸ Lack of arterial enhancement on any pancreatic lesion should raise the possibility of other diagnoses. The CT findings in this case did not show an arterial enhancing hypervascular lesion because of presumed neovascularisation causing arteriovenous shunting in the tumour.

No clear consensus exists on the management of adult pancreatic haemangiomas but no treatment is advocated in paediatric cases as they usually regress.² Although a

conservative approach can be recommended owing to the benign nature of the lesion, this strategy needs to be balanced with the risk of bleeding (which may be the presenting symptom) and the possible differential diagnoses.^{2,6} Operative intervention needs to weigh the morbidity and mortality of pancreatic resection against any diagnostic uncertainty. Postoperative survival is excellent given the benignity of the tumour, which allows pathological classification of the lesion. Histological examination may reveal blood filled and cystic spaces, covered by flat endothelium, separated by scanty connective tissue stroma.⁸ The cystic spaces can either be small (capillary haemangioma) or dilated (cavernous haemangioma) and they can be associated with thrombosis or haemorrhage.¹ Immunohistochemical analysis may detect the presence of endothelial markers CD31 and CD34, suggesting a vascular endothelial origin.⁵

Conclusions

Pancreatic haemangiomas are rare benign lesions that present both diagnostic and therapeutic challenges to the clinician. Radiological and endoscopic assessment, even combined with cytology or tissue biopsy, is often inconclusive and raises concerns as to the exact nature of the pancreatic lesion. This diagnostic uncertainty, coupled with the patient's symptomatology, often results in pancreatic excision to provide definitive diagnosis in adults. To date, this is the only documented case of expectant management of this highly unusual tumour in adults, which has been facilitated by the presence of hypervascularity on EUS assessment.

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