Head of the pancreas mass that turned out to be not a pancreatic cancer

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DESCRIPTION

A 58-year-old woman with nail–patella syndrome, iron deficiency anaemia and haematuria, with no prior history of gastrointestinal disorders was found to have an incidental 3.9 cm × 3.2 cm × 3.0 cm pancreatic head/duodenal mass on a CT scan (figures 1 and 2A), which was done as a part of haematuria work up. The patient was asymptomatic. Further tests for carcinoembryonic antigen, cancer antigen 19–9, C reactive protein and erythrocyte sedimentation rate were unremarkable. Magnetic resonance cholangiopancreatography (MRCP), with and without contrast, was significant for a 2.9 × 3.8 × 2.7 cm predominantly solid, T2 hyperintense, T1 hypointense mass within the head of the pancreas, at its inferior margin, just below the pancreatic duct (figure 2B). This mass demonstrated homogenous enhancement on postcontrast arterial phase imaging. Oesophagogastroduodenoscopy and colonoscopy were

unremarkable. Endoscopic ultrasound (US) with transduodenal fine needle aspiration (FNA) of the mass was consistent with a mass arising from the second duodenal portion (figure 3). Biopsy of the tissue revealed spindle cells with positive staining for c-KIT, DOG-1, vimentin, CD34 and a weak patchy staining for smooth muscle actin with a mitotic count of 0 mitoses per 14 available high-powered fields. These findings supported the diagnosis of a gastrointestinal stromal tumour, and the patient was referred to general surgery for resection. During surgery, an approximately 5 cm mass was found in the second/third portion of the duodenum. The mass was noted to be adherent to the pancreas and originating from the duodenum, part of which was resected. The biopsy of resected mass was concurrent with FNA sample (figure 4).

Gastrointestinal stromal tumors (GIST) located in the duodenum are very rare and represent

Figure 1 CT of the abdomen: (A) coronal view and (B) axial view of the pancreatic head/duodenal mass with the claw sign present on image (B).

Figure 2 (A) CT of the abdomen, axial view of the pancreatic head/duodenal mass with the claw sign present (arrows); (B) magnetic resonance cholangiopancreatography axial view of the abdomen with a mass within the head of the pancreas, at its inferior margin, just below the pancreatic duct.

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Figure 3 Endoscopic ultrasound: (A) with a mass and (B) with normal pancreatic duct.

Figure 4 Microscopic view of (A) the resected mass with duodenal tissue and (B) the tissue revealing spindle cells.

less than 5% of GIST.¹ There are specific features that are required for diagnosis of duodenal GIST: positive CD117/CD34 immunohistochemical staining, oncogenic c-KIT or PDGFA mutation and a tumour arising from duodenal wall.² Currently, these tumours can be stratified based on Fleischer's criteria in very low, low, intermediate and high risk categories based on tumour's size and mitotic activity. Small intestinal GISTs ≤ 5 cm with a mitotic count ≤ 5 per 50 high-power fields have a very good prognosis with only 3%–5% of metastatic risk. In our case, the majority of the imaging was consistent with a mass at the head of the pancreas (figures 1 and 2). However, features of the tumour were not entirely consistent with pancreatic mass and pointed towards GIST. Specifically, the pancreatic duct was not dilated (confirmed by US; figure 3), the tumour was round, smooth and exhibited a characteristic claw sign³ (figures 1 and 2).

Learning points

- ▶ Keeping wide differential is important: as in this case the management was chosen based on the results of complete work up.
- ▶ Even though the CT scan and magnetic resonance cholangiopancreatography suggested the pancreatic head tumour, endoscopic ultrasound was performed to complete work up and was essential in further characterisation of the specific location of the tumour.
- ▶ Claw sign can be used as a tool to differentiate if a tumour is arising within organ (claw sign present).

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