CASE REPORT

Retroperitoneal ectopic pancreas: imaging findings

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ABSTRACT. Ectopic pancreas is relatively uncommon and usually occurs in the stomach or duodenum. Retroperitoneal ectopic pancreas has not previously been documented. We report the case of a 48-year-old man with retroperitoneal ectopic pancreas that imitated bilateral adrenal tumours on ultrasound and MRI. Subsequent CT-guided biopsies confirmed an ectopic pancreas. The lesions remained stable during follow-up for 7 years. In retrospect, the similarity in signal intensities and enhancement pattern between the retroperitoneal masses and the pancreas may have been a clue to the diagnosis of this rare entity.

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Ectopic pancreas is an uncommon finding, with an estimated incidence of 0.55–13.7% according to autopsy analyses [1]. Most ectopic pancreatic lesions are found in the gastrointestinal tract and frequently involve the stomach (26–38% of cases), duodenum (28–36%), jejunum (16%) and, less commonly, Meckel's diverticulum or the ileum [1–3]. Although rare, ectopic pancreas in the lung and the mediastinum has also been documented [4, 5]. However, ectopic pancreas in the retroperitoneum has not been reported. Herein, we describe the imaging features in a case of retroperitoneal ectopic pancreas with an extremely unusual presentation — imitation of bilateral adrenal tumours.

Case report

A 48-year-old man visited our hospital because of mild flank soreness of 3 months' duration. A physical examination showed no abnormal findings. His pulse rate was 68 beats per minute and his blood pressure was 116/78 mmHg. Laboratory investigations revealed no abnormal findings. Abdominal ultrasound showed bilateral suprarenal well-defined masses nearly isoechoic to the liver (Figure 1a). Intravenous urograms showed a mass effect in the bilateral suprarenal regions with downward displacement of both kidneys. The chest radiograph was normal. Abdominal MRI revealed retroperitoneal masses (7 cm on the right, 4.5 cm on the left) in the bilateral suprarenal regions. The signal intensities of the lesions were parallel to those of the pancreas on T_1 and T_2 weighted images, and the enhancement of the lesions was also similar to that of the pancreas (Figure 1b,c). The initial impression included bilateral adrenal adenomas, lymphoma and metastasis. The serum adrenocortical hormone and aldosterone levels

and the 24 h urine vanillylmandelic acid level were normal. Ultrasound-guided biopsy of the left retroperitoneal mass was performed, and histopathological examination showed that the core biopsied specimen was composed of pancreatic acini. Follow-up amylase and lipase levels were normal, and there was no evidence of biopsy-related pancreatic injury. CT-guided biopsy (Figure 1d) of both retroperitoneal masses was performed 1 week later, and histopathological examination of the biopsied specimens also revealed pancreatic acini (Figure 1e). Chest CT revealed no lung mass or enlarged thoracic lymph node. A diagnosis of bilateral retroperitoneal ectopic pancreas was made. Surgical resection was suggested but the patient refused, and thus he was managed conservatively with regular follow-up. Over the past 7 years, the bilateral retroperitoneal masses have remained stable and there has been no evidence of malignancy elsewhere or any condition associated with a hyperfunctioning adrenal tumour in this patient.

Discussion

Most cases of ectopic pancreas are found in the upper gastrointestinal tract; 70.0–86.5% of cases affect the stomach, duodenum or jejunum [1, 2]. Unusual locations that have been described for an ectopic pancreas include the gallbladder, bile ducts, splenic hilum, umbilicus, fallopian tubes, mediastinum, oesophagus, colon, omentum, lung, mediastinum and lymph nodes [1–7]. To our knowledge, this is the first reported case of ectopic pancreas occurring in the retroperitoneum.

The embryologic origin of ectopic pancreatic tissue is controversial; hypotheses include (i) the anomalous separation of the developing pancreatic anlagen, with bowel wall penetration and subsequent distant transport, or (ii) differentiation of the totipotent endodermal cells in the intestinal tract into pancreatic tissue [3, 4]. However, ectopic pancreas found in the lung, mediastinum and

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Figure 1. (a) Abdominal sonogram shows a well-defined suprenal mass (arrows) that is isoechoic to the liver. (b,c) Coronal T_1 weighted and fat-suppressed enhanced T_1 weighted images show well-defined bilateral suprenal masses (black and white arrows) with a signal intensity and enhancement similar to that of the pancreas (open arrows). (d) Axial scan of a CT-guided biopsy confirms the correct position of the biopsy needle within the right suprenal mass (arrows). (e) Photomicrograph shows typical pancreatic acini in the biopsied specimen (haematoxylin and eosin stain, original magnification $\times 200$).

lymph nodes, as well as our case of retroperitoneal ectopic pancreas, suggests that differentiation of totipotent cells into pancreatic tissue in extraintestinal locations is plausible [3–6].

An ectopic pancreas usually measures 0.5–2.0 cm and, occasionally, can measure up to 5 cm [1, 2]. Most cases of

ectopic pancreas in the gastrointestinal tract are asymptomatic, although complications such as stenosis, ulceration, bleeding, intussusception, pancreatic cancer and acute pancreatitis may develop [1–3]. In contrast, our patient presented with mild flank soreness, which might be related to the retroperitoneal mass effect.

Ectopic pancreas typically appears on a radiographic barium study or endoscopic examination as a submucosal mass with central umbilication [2, 3]. On CT, an ectopic pancreas most frequently appears as a well-defined oval or round mass with smooth or serrated margins in the gastric antral wall or intestinal wall, or as a mesenteric mass similar to a gastrointestinal stromal tumour or carcinoid tumour [2, 3, 7]. Our case manifested as incidental retroperitoneal masses mimicking bilateral suprarenal masses on ultrasound and MRI. Therefore, the initial differential diagnoses included metastasis, lymphoma, pheochromocytoma, adenoma and haematoma [8–11]. The lack of hypertension and hyperfunctioning adrenocortical conditions was helpful in excluding pheochromocytoma and functional adenoma [8–10]. The lack of any trauma history or signal intensities of haemorrhage on MRI was helpful in excluding haematoma [11]. However, definitive differentiation of metastasis and lymphoma on non-invasive imaging may be difficult, and thus image-guided biopsy was inevitable [8–10]. In this particular case, the ultrasound-guided biopsied specimen revealed pancreatic tissue and we initially suspected that the biopsy specimen was wrongly taken from the pancreas rather than the left retroperitoneal mass, but follow-up serum lipase and amylase levels showed no evidence of biopsy-induced pancreatic injury. Nevertheless, subsequent CT-guided biopsies, which allowed accurate localisation of both retroperitoneal masses, were performed, and bilateral retroperitoneal ectopic pancreas was histopathologically confirmed. Owing to the mass effect, with obliteration of the normal adrenal glands, and the patient's refusal to undergo surgical resection, it could not be determined whether the lesions originated from the adrenal glands or the para-adrenal soft tissues. Even so, retroperitoneal ectopic pancreas could be diagnosed definitively. In retrospect, the similarity in signal intensities and enhancement pattern between the retro-

peritoneal masses and the pancreas may have been a clue to the diagnosis of this rare entity, for which CT-guided biopsy allowed a definitive diagnosis. In light of this case, ectopic pancreas should be included as a rare differential consideration for bilateral retroperitoneal or suprarenal masses.

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