

Radiopathological correlation of a von Hippel-Lindau syndrome associated pancreatic neuroendocrine tumour with clear cell features

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DESCRIPTION

A 47-year-old woman with von Hippel-Lindau (VHL)-associated cerebellar haemangioblastoma presented for surveillance abdominal MRI. A 3 cm pancreatic tail mass lesion showed T1 signal drop on oppose-phase images, indicating intracellular lipid component (figure 1A,B). It also exhibited intermediate T2 signal and hyperenhancement on arterial phase images, suggesting neuroendocrine tumour (NET) (figure 1C,D). On Gallium-68 (68Ga)-DOTATATE positron emission tomography (PET)/CT scan, this lesion was hypodense on CT and showed intense tracer uptake on PET (figure 1E,F), consistent with a somatostatin receptor positive lesion. No other tracer avid lesion was identified in the remaining torso.

The patient underwent R0 resection via laparoscopic distal pancreatectomy and splenectomy. The 3.0 cm pancreatic mass was successfully removed. Histologically, the tumour cells exhibited cytoplasmic clearing and vacuolisation (figure 2A). Immunohistochemistry demonstrated tumour reactivity for chromogranin and synaptophysin (figure 2B), five mitoses per mm² and Ki-67 proliferative index of 16.2%, consistent with

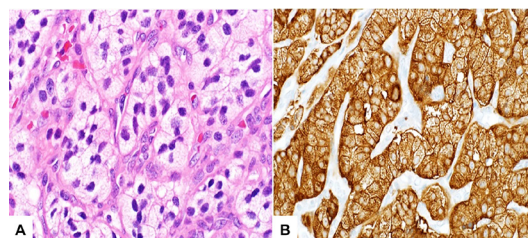


Figure 2 Photomicrography images demonstrate (A) nests of tumour cells with clear vacuolated cytoplasm and nuclear features consistent with neuroendocrine neoplasm (H&E stain, $\times 200$), and (B) synaptophysin reactivity (immunoperoxidase stain, $\times 200$).

well-differentiated pancreatic NET, WHO grade 2, with clear cell features.

VHL syndrome is an inherited autosomal dominant disorder, caused by germline mutation of VHL gene on the short arm of chromosome 3.¹ VHL usually leads to the development of multiple benign and malignant neoplasms, including retinal haemangioblastoma, central nervous system haemangioblastoma, endolymphatic sac tumour, pancreatic NET, pancreatic cystadenoma, pancreatic cyst, clear cell renal cell carcinomas, renal cyst, pheochromocytoma, paraganglioma, epididymal cyst and broad ligament cyst.²

CT, MRI and ultrasound play an important role in diagnosis and surveillance for VHL-associated neoplasms. On CT and MRI, pancreatic NETs have a distinctive appearance, typically a well-defined hypervascular mass on arterial phase images. When located at the tip of the tail of the pancreas, the mass must be distinguished from a benign intrapancreatic splenule. Endoscopic ultrasound allows us to acquire tissue sample of the respective lesion. The new 68Ga labelled somatostatin receptor analogue PET/CT is superior to Indium-111 Octreoscan in detection of low to intermediate grade pancreatic NETs which usually have overexpressed somatostatin receptors on the tumour cell surface.³

In this case, the distinctive signal pattern of a hyperenhancing, lipid rich neoplasm on MRI and positive somatostatin receptor expression on Ga68-DOTATATE PET/CT corresponded to a pancreatic NET. To our knowledge, this is the first report of MRI showing signal dropout on opposed-phase imaging indicating the lipid-rich clear cell feature of a pancreatic NET in VHL syndrome.

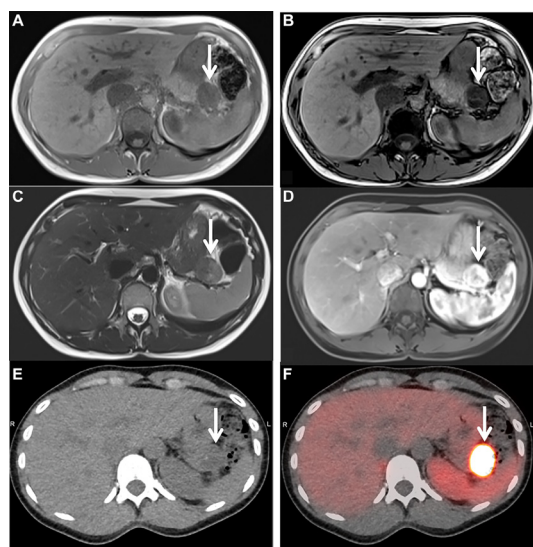


Figure 1 Pancreatic tail mass lesion (arrow) on abdominal MRI: (A) T1 in-phase image, (B) signal dropout on T1 oppose-phase image, (C) T2-weighted image, (D) hyperenhancement on arterial phase. On 68Ga-DOTATATE positron emission tomography (PET)/CT, (E) tumour exhibits hypodensity on axial CT image and (F) intense somatostatin receptor positivity on fused PET/CT image.



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Learning points

- ▶ von Hippel-Lindau (VHL) syndrome is an inherited autosomal dominant disorder manifesting a broad spectrum of multisystem neoplasms.
- ▶ MRI with signal dropout on out-of-phase T1-weighted images shows intracellular lipid from clear cell features in neuroendocrine neoplasm of the pancreas.
- ▶ Combination of abdominal MRI and ⁶⁸Ga-DOTATATE positron emission tomography/CT is useful to identify VHL-pancreatic neuroendocrine tumours.

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elsewhere. The authors' contribution details are: ACH: planning, manuscript preparation, figures selection, reference preparation. JMC: reporting, manuscript preparation, figures selection. CN: planning, patient consent, manuscript preparation. MY: planning, reporting, manuscript preparation and finalising, figures selection and finalising.

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