Acinar Cell Cystadenoma – a Rarity in Advanced von Hippel-Lindau Disease: A Case Report

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Case Report

A 40-year-old woman suffering from VHL disease presented for evaluation of the findings of a positron emission tomography/magnetic resonance imaging (PET/MR) which was performed to monitor an osteodestructive sacral haemangioblastoma. The results of the imaging showed numerous cystic tumours and a single solid tumour of the pancreas as well as a retroumbilical mass. Consultant radiological opinion considered the solid tumour to be a pancreatic neuroendocrine tumour (pNET) with a retroumbilical metastasis.

Abdominal examination revealed a palpable supraumbilical resistance without tenderness. Computed tomography showed hyperperfused lesions in the pancreatic head which were evaluated as either haemangioblastomas or neuroendocrine tumours (fig. 1). The known sacral haemangioblastoma had grown, and there was no sign of the retroumbilical mass. Additionally, magnetic resonance cholangiopancreatography revealed a large amount of cystic lesions throughout the entire pancreas (fig. 2).

We decided to explore the site laparoscopically. The retroumbilical mass corresponded to a tumour sized 2 × 3 cm in the greater omentum, which was resected. Pathological evaluation was difficult due to an uncommon arrangement of immunohistochemical markers. The mass was categorized as either a metastasis of a solid pseudopapillary tumour or as a haemangioma. It was agreed to resect the pancreatic lesion to be able to compare the particular histologic findings and to exclude a possible malignancy.

Therefore, an uneventful pylorus-preserving hemipancreatectoduodenectomy was performed.

Histological evaluation revealed a neuroendocrine tumour which was 1.3 × 1.2 × 1.2 cm in size. The TNM classification was pT1, pN0 (0/19), M0, L0, V0, Pn0, R0, G1. The proliferation rate was measured by MIB-1 and accounted for <1%. Considering these results, the umbilical lesion was re-evaluated and despite the initial assessment classified as a haemangioma. A Sister Mary Joseph node could be excluded.

The additional pancreatic cysts were lined with a single-layered epithelium, with the largest one measuring 4.5 × 3.6 × 1.5 cm. These lesions were evaluated as acinar cell cystadenomas, although cysts are equivalent to serous cystadenomas in VHL disease (fig. 3).

Discussion

VHL disease is inherited, and its genetic locus is on the short arm of chromosome 3 [1]. In early stages the disease often remains
asymptomatic; later on, blindness, neurological complications, or death may occur [3].

Pancreatic involvement is diagnosed in 50–80% of patients with VHL disease [4]. In about 8% of cases pancreatic manifestations occur solely and are either cystic or solid tumours. Solid masses are mostly neuroendocrine tumours, while cystic lesions are rather simple cysts or serous cystadenomas [5]. Therefore, most pancreatic lesions are benign, but malignant or premalignant tumours such as intrapancreatic mucinous neoplasms or metastases of renal cell carcinoma need to be excluded [6].

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5–17% of VHL disease patients suffer from pNET [7]. These neoplasms are malignant in 17% [8]. Most neuroendocrine tumours do not influence physiological processes by secretion of hormones, but there is a risk of dissemination to distant organs or of local advancement. At the time of diagnosis, up to 25% of the neoplasms have already metastasized [9].

Surgical treatment is recommended for tumours >2 cm. If the tumour is <2 cm, resection should only be performed in case of at least G2 grading or if symptomatic, otherwise surveillance by annual MRI scans is advised. Whenever possible, oncological resection is to be aimed for [10]. Advanced stages of a pNET can be treated by chemotherapy, targeted drugs, or peptide receptor radionuclide therapy. For poorly differentiated tumours, G3 platin-based cytostatic agents are used [11].

Conclusion

pNETs are found in <17% of patients with VHL disease. Generally, guidelines recommend a surgical treatment if the tumour is >2 cm. In our case, however, a hemipancreatectoduodenectomy needed to be performed despite a pNET tumour size <2 cm, because metastatic spread could not be excluded in histopathological testing. Additional pancreatic lesions were lined with epithelium and considered as acinar cell cystadenomas.

Disclosure Statement

The authors declare no conflict of interest.

References


