CLINICAL CASE

Massive postoperative ascites following pancreatic cysts fenestration in a patient with von Hippel-Lindau disease

Ascite postopératoire après fenestration de kystes pancréatiques chez un patient atteint de maladie de von Hippel Lindau

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Summary Pancreatic lesions in von Hippel Lindau disease (VHLD) are frequent and mainly consist of cystic lesions, which should not be resected because of their benign evolution. Solid lesions, mostly pancreatic endocrine tumors (PET), are rare and usually occur in combination with cystic lesions. We report a case of a patient with VHLD who underwent PET enucleation in a polycystic pancreas requiring fenestration of multiple adjacent cysts, to ensure complete resection with free resection margins. The postoperative course was complicated by massive ascitic fluid effusion, probably related to pancreatic-cyst fenestration. Although this complication is well-known after liver-cyst fenestration, it has not been reported after pancreatic-cyst fenestration. This observation emphasizes that morbidity from surrounding pancreatic polycystic disease should not be underestimated in pancreatic surgery for VHLD.

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Abbreviations: VHLD, von Hippel-Lindau disease; PET, pancreatic endocrine tumor.

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Résumé Les lésions pancréatiques dans la maladie de von Hippel-Lindau sont fréquentes, le plus souvent des lésions kystiques devant être respectées, du fait de leur évolution bénigne. Les lésions tissulaires, plus rares, sont principalement des tumeurs endocrines et en général associées aux lésions kystiques. Nous rapportons ici le cas d’une patiente atteinte de maladie de von Hippel-Lindau, ayant eu l’énucléation d’une tumeur endocrine pancréatique, avec la nécessité, pour une résection R0, de fenêtrer de multiples kystes pancréatiques adjacents. Les suites postopératoires ont été marquées par une abondante ascite, attribuée à la fenestration des kystes. Si cette complication est rapportée après le traitement chirurgical des polykystoses hépatiques, aucun cas n’avait été jusqu’à présent publié après la fenestration de kystes pancréatiques. Cette observation souligne que dans la chirurgie pancréatique dans la maladie de von Hippel Lindau, la morbidité liée à la fenestration des kystes, ne doit pas être sous-estimée.

Von Hippel-Lindau disease (VHLD) is an autosomal dominant inheritable cancer syndrome, characterized by the development of severe benign or malignant tumors in many organs. Visceral features include renal cysts and clear cell carcinomas, pheochromocytomas or paragangliomas, pancreatic cysts and endocrine tumors, as well as epididymal and broad ligament cystadenomas.

We report the case of a young woman with VHLD, who underwent combined resection of a para-aortic paraganglioma and enucleation of a pancreatic endocrine tumor (PET) from a polycystic pancreas. The postoperative course was complicated by massive ascitic fluid effusion, probably related to fenestration of multiple adjacent pancreatic cysts.

Case Report

A 36-year-old black woman was admitted for abdominal pain and flushing, in association with weight loss of 5 kg in the past three months. She had a past history of high blood pressure with no familial history of inherited disease. Helical computed tomography and MRI (Figs. 1 and 2) showed a 55 × 40 mm para-aortic solid lesion without vascular involvement or enlarged para-aortic lymph nodes, associated with multiple cystic lesions in the pancreatic gland and a 24 mm solid lesion in the head of the pancreas with strong enhancement on contrast arterial injection. The solid pancreatic lesion was at a distance from the main pancreatic duct, but closely surrounded by multiple cystic lesions. Meta-iodobenzylguanidine (MIBG) scintigraphy showed unique and intense uptake of the para-aortic area with normal uptake of adrenal glands, associated with high levels of serum noradrenaline (three times normal values) and urinary normetanephrine (12 times normal values). The levels of pancreatic hormonal peptides (gastrin, glucagon, insulin, somatostatin, vasoactive intestinal peptide) were normal. The association of para-aortic paraganglioma, non-functional PET and multiple pancreatic cysts suggested VHLD.

Surgical exploration revealed a typical para-aortic paraganglioma, which was removed with lateroaortic lymphadenectomy. Examination of the pancreas by both manual and intraoperative ultrasound showed an enlarged polycystic pancreatic gland with a well-limited solid lesion. The latter was removed by enucleation (Fig. 3) and fenestration of 10 to 15 adjacent pancreatic cysts (from 8 to 17 mm)
was required, to ensure complete resection with free resection margins. An abdominal drain was inserted close to the pancreatic enucleation area. From postoperative day 1 to 5, drain production was above 1000 mL/24 h of clear ascitic fluid. Measurement of pancreatic enzymes in the ascitic fluid was normal (inferior to 25 UI/L), as well as white cell count (110 per mm$^3$), lymphocyte count (< 5%), albumin level (13 g/L), cholesterol, chylomicron and triglycerides, so that the diagnosis of chylous ascitic effusion from lateroaortic lymphadenectomy could be excluded. There was no evidence of right-heart failure or chronic-liver disease and the portal vein was patent on Doppler ultrasound. The abdominal drain was removed at day 7, but resulted in persistent ascitic oozing (> 2000 mL/24 h between day 7 and 10) through the abdominal wall defect. Ascitic infection developed and the patient subsequently developed fever, abdominal tenderness, high white blood-cell count and positive culture of ascitic fluid for Escherichia coli. Abdominal CT scan disclosed multilocular ascites requiring a second intervention at day 11 for several reasons including poor clinical status, the impossibility of performing effective percutaneous radiological drainage and suspected surgical complications. Surgery failed to demonstrate pancreatic or intestinal complications and showed upper mesocolic multiloculated infected ascites. Treatment included abdominal washing and drainage. The drain was removed on day 19, because of prolonged ascitic fluid production with the same biochemical profile and the patient was finally discharged on day 22. Pathological examination identified a malignant parangangioma of 60 mm. The PET showed a well-differentiated pattern without vascular invasion and both antichromogranin A and anti-synaptophysin antibodies were positive on immunohistochemistry. The percentage of Ki-67 positive cells was 2% and there were less than two mitosis per 10-high power fields. Genetic testing confirmed the diagnosis of VHLD. At follow-up (16 months), the patient is doing well without recurrence or evidence of endocrine or exocrine insufficiency.

Discussion

The prevalence of pancreatic lesions in VHLD has been reported to vary between 55 and 70% and seems to depend on family and genotype [1, 2]. Most of these lesions remain asymptomatic and mainly consist of cystic lesions: isolated or multiple cysts in around 91% and serous cystadenomas in 12% [1]. Most solid lesions observed in VHLD are PET, usually observed in combination with cystic lesions in 11 to more than 45% of the cases. The risk of malignancy is estimated to be between 8 and 25%, correlated to the size of the tumor (with a risk close to zero is tumors less than 3 cm), genotype of the disease and tumor doubling time [3], but seems to be less important that in sporadic PET. In view of the predominantly benign lesions, we believe that resection of PET in VHLD patients should favor limited pancreatectomy or enucleation when feasible. However, the presence of polycystic-pancreatic disease can modify the anatomy of the pancreatic gland and increase morbidity at surgery. In this case, pancreatic enucleation required fenestration of 10 to 15 adjacent cysts, to ensure complete resection of PET with free resection margins. This resulted in massive ascitic fluid effusion, secondary complication by multiloculated infected ascites, leading to a second intervention for drainage and washing. Repeat biochemical measurements and white blood counts in ascitic fluid (amylase level, cholesterol, triglycerides, chylomicrons, albumin) failed to demonstrate pancreatic or chylous leakage secondary to pancreatic enucleation or lymphadenectomy. A complete medical work-up did not identify other causes of postoperative ascites, that is, portal hypertension, renal failure or right ventricular insufficiency. Ascending infection along the abdominal drain left in place because of high ascitic output might have caused multiloculated infected ascites. Massive ascitic effusion has previously been reported after multiple fenestration of polycystic-liver disease [4–7] and has been attributed to fluid secretion from cyst epithelium, when fluid production from fenestrated cysts exceeds the resorption capacity of the peritoneum. We believe that a similar phenomenon might have been responsible for the massive ascitic effusion following enucleation of PET from a polycystic pancreatic disease.

Very few clinical reports have focused on enucleation of PET in VHLD disease. To our knowledge, this is the first description of massive ascitic fluid effusion after pancreatic enucleation, requiring multiple fenestrations of adjacent cysts.

In conclusion, indications for pancreatic surgery in VHLD are limited and should be restricted to PET taking into account the size of the lesion, the symptoms, risk of malignancy and the presence of other intra-abdominal tumors. Enucleation or limited pancreatic resection should be performed whenever possible, but surgical morbidity due to surrounding pancreatic-polycystic disease should not be underestimated.

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References


