Major pancreatic surgery for von Hippel-Lindau disease (VHL) 3 cases and literature review

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Background

VHL gene abnormality - 1 / 36,000 individuals

Renal cell carcinomas and pancreatic NETs are true cancers and can result in mets and death

- Pancreas is involved in 15% of patient with VHL
- Major pancreatic surgery for VHL is recommended if

- symptomatic large (>5 cm) serous cystadenomas or
- neuroendocrine pancreatic tumor (pNENs) > 2 cm appear

Aim

To assess the demographics and results of major pancreatic surgery in patients with VHL

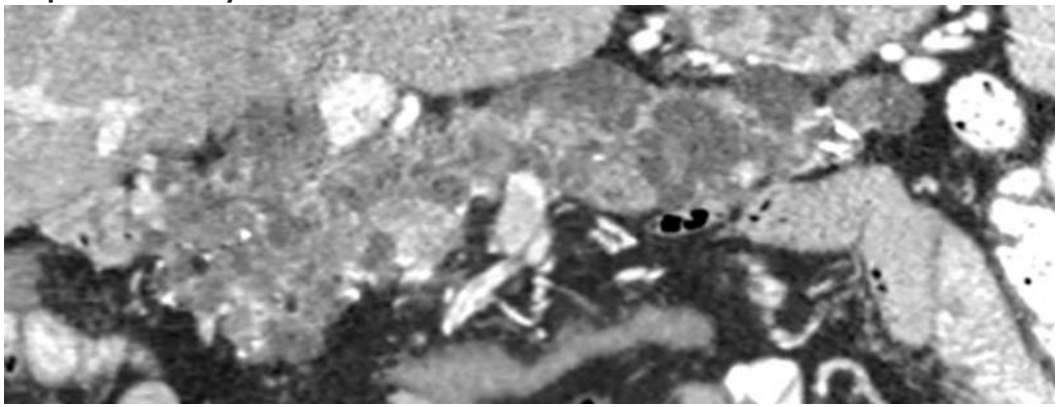
Methods

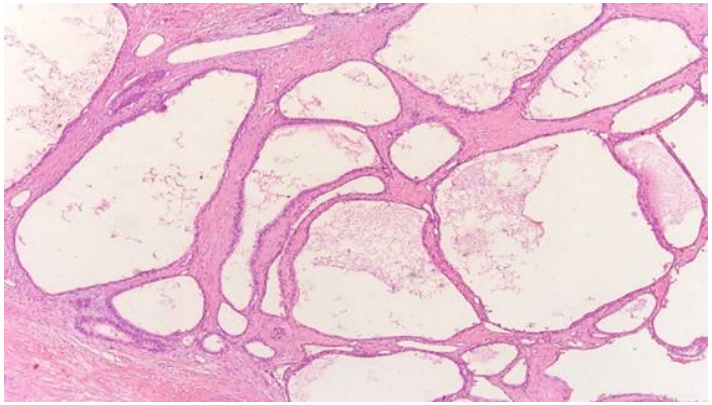
Retrospective analysis of 5th MCH Department of Surgical Oncology database 2013 – 2015

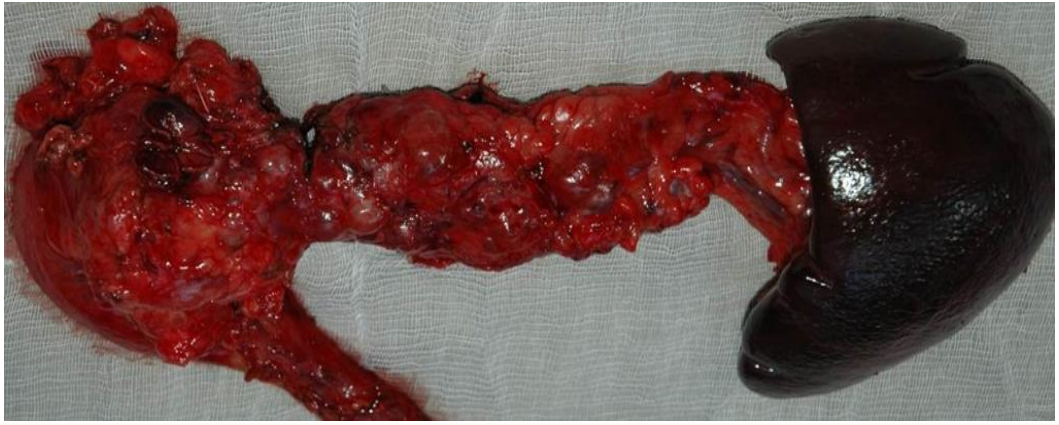
Results

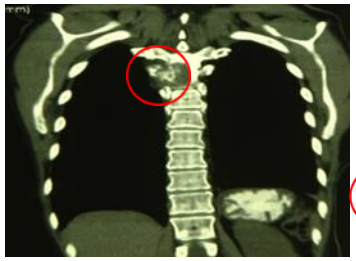
	Total pts, n	M,n	F,n	Age,yrs	MHS,d
Under Surveillance	10	3	5	42	
Major pancreatic resections	3	1	2	48	
Working ability		All patients			

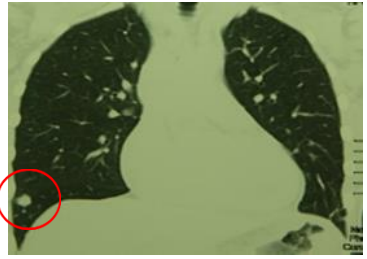
Case 1. F, 54: VHL type 1 (mutation c.551T→C (p.L184P) : head & tail pNETs on total serous cystadenoma pancreatic involvement ; history of RCC treated by right-sided nephrectomy





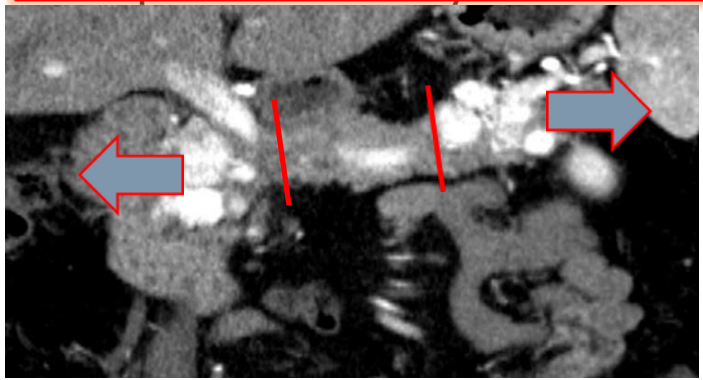


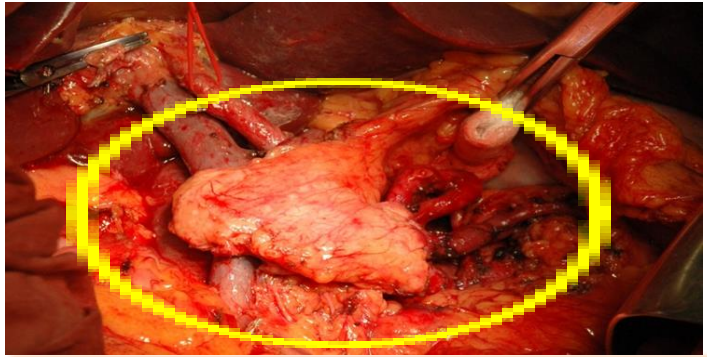




Died 8 months after surgery due to RCC mets

Case 2 F, 51: VHL type 2b (no detectable mutations): multiple CNS hemangyoblastomas; head & body& tail G1 pNETs; history of partial resection of the left adrenal gland for Pheochromocytoma (1989) and right adrenalectomy for Pheochromocytoma (1992); polycythemic syndrome Somatic mosaicism. Diagnosis had been proved clinically

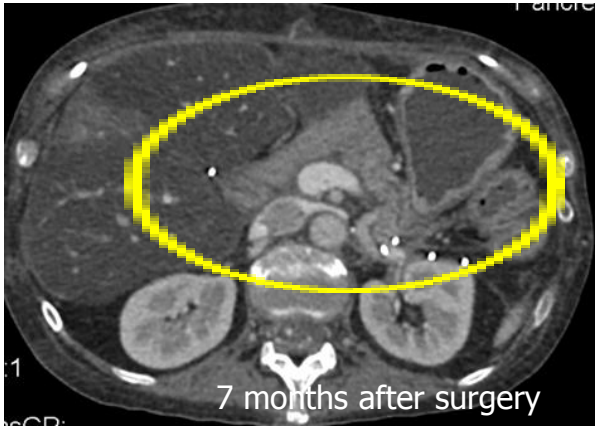




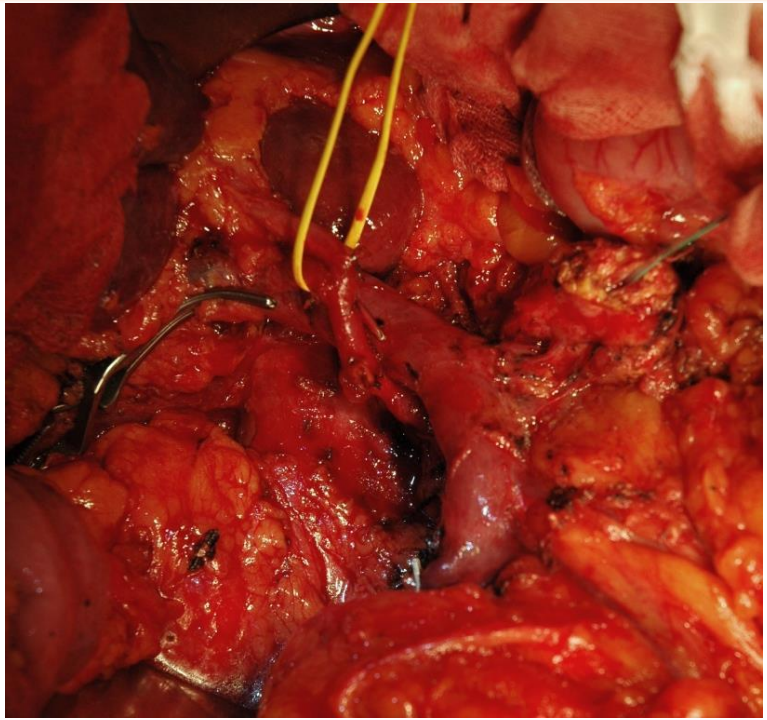
Middle-preserving pancreatectomy for NEN & left adrenalectomy for pheochromocytoma

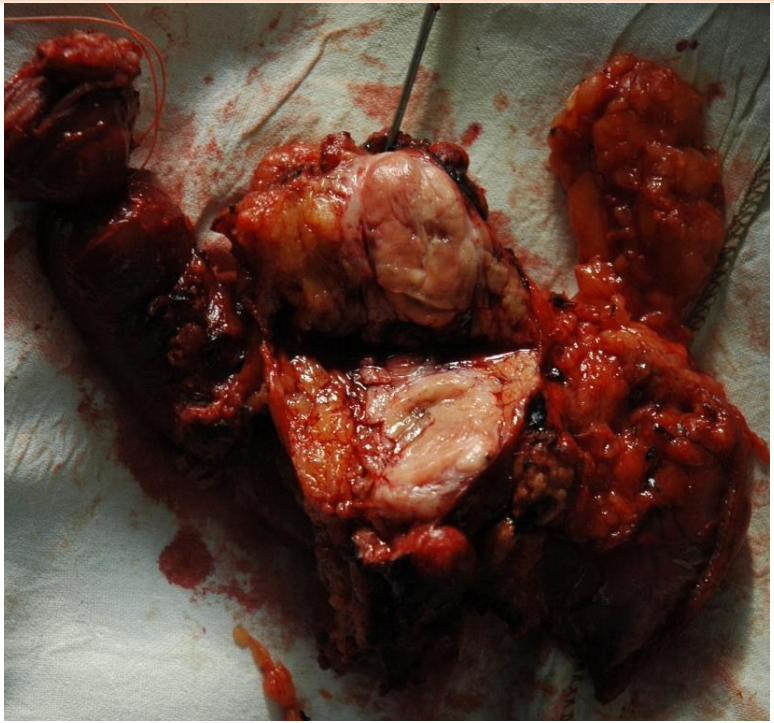
Non-complicated postoperative period

Surveillance time = DFS = 7 months



Case 3 M, 45: VHL type 2b (mutation 695 G → A (p.R161Q): spine hemangyoblastomas; head pNEC G2p$T_3N_{1(3/13)}M_0$, L1P1, R0 IIB ; history of bilateral adrenalectomy for Pheochromocytoma and nephrectomy





Whipple procedure, Surveillance time = DFS 10 months

All the patients were discharged and two latter at the moment are functional, working and fully compensated

Conclusion

Timely and possibly parenchima-sparing pancreatic resections are the operations of choice for pNETs in VHL patients