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Two Stage Resection of a Bilateral Pheochromocytoma and Locally Advanced Pancreatic Tumor in a 26 Years Old Pregnant Women: Case Report

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Abstract **Case Report**

Synchronous occurrence of pheochromocytoma and pancreatic tumors is rare and considered as part of Von Hippel Lindau Syndrome. We herein report a such case that was managed by a two staged resection of the pancreatic tumor along with excision of bilateral pheochromocytomas and also discuss in detail our review of literature about this condition.

Keywords: Synchronous, pancreatic tumors, Von Hippel Lindau Syndrome, pregnanc, pheochromocytomas.

INTRODUCTION

Von Hippel–Lindau disease is a multisystemic tumor predisposition syndrome characterized by benign and malignant tumors, including central nervous system and retinal hemangioblastomas, clear cell renal cell carcinoma, pheochromocytoma, pancreatic neuroendocrine tumors, endolymphatic sac tumor, and epididymal and broad ligament cystadenoma, as well as visceral cysts [1].

Originally described in 1936 by von Hippel [2] and Lindau [3], the incidence of VHL is estimated at approximately one in 36,000 [4].

The clinical diagnosis is confirmed upon identification of a pathogenic variant in the *VHL* gene, the gold standard for VHL diagnosis [5].

Surgical intervention for tumors may be associated with operative/postoperative complications, such as hemorrhage. Historically, substantial mortality was attributable to pancreatic neuro endocrine tumors [6]. Fortunately, these risks have been mitigated in recent years by the institution of comprehensive surveillance paradigms, leading to early recognition of tumors and their multidisciplinary management [7].

CASE REPORT

26 years old pregnant women whose pregnancy reached the end of the first trimester presented during the monitoring consultation of her gestational diabetes high blood pressure figures, in addition she reported the notion of headaches and palpitations.

- Biological studies revealed an increase of methoxylated drifts: Normetadrenaline: 43.98 umol/24h
- An abdominal ultrasound showed a positive fetal cardiac activity
- An MRI revealed the presence of bilateral adrenal gland masses, they measured 67 x 33 mm and 46 x 32 mm respectively, it also revealed a rounded pre-aortic lymph node mass, measuring 50 x 44 mm in diameter.

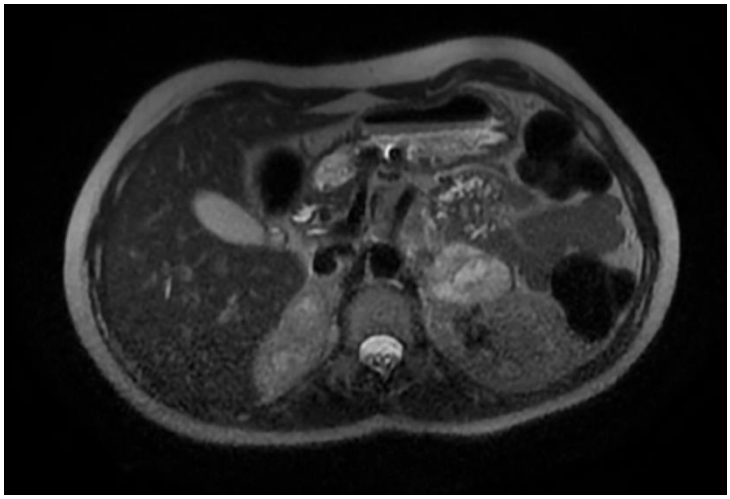

Figure 1: MRI showing the two adrenal glands tumors

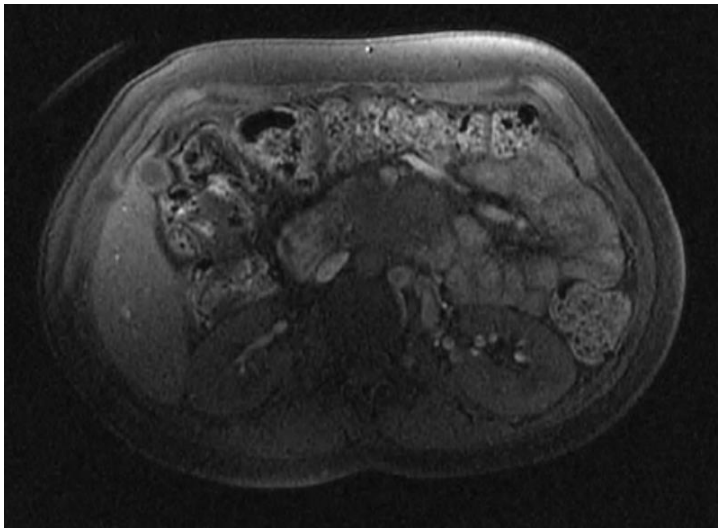

Figure 2: MRI showing the pre aortic mass

A CT scan revealed two bilateral adrenal masses and a voluminous mesenteric mass infiltrating the body of the pancreas delimiting central calcifications. It measured 64x34 mm.

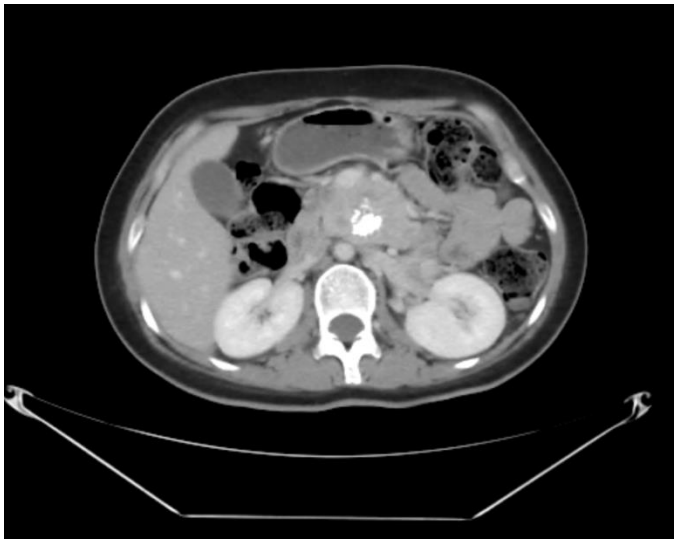

Figure 3: CT scan showing the voluminous calcified pancreatic mass

The patient underwent embolization of the three tumor masses and the patient was operated the next day.

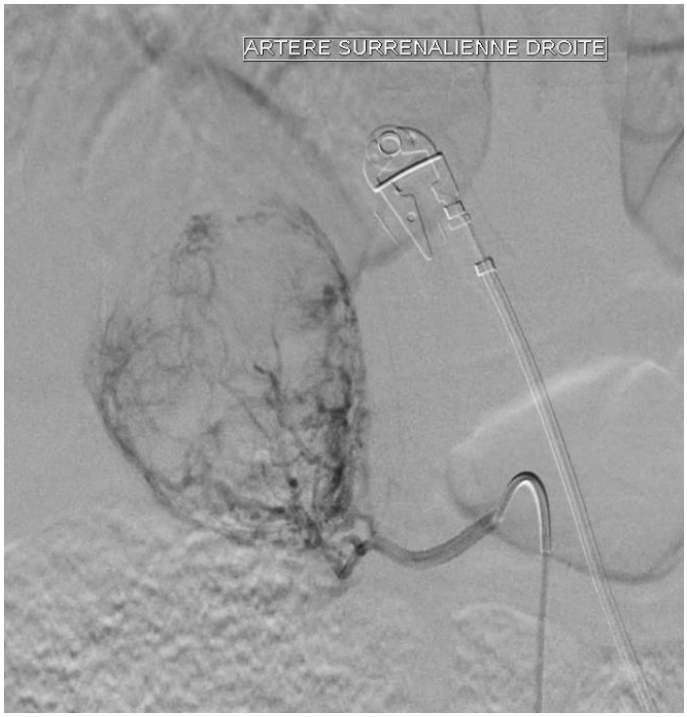

Figure 4: Embolization of the right adrenal artery

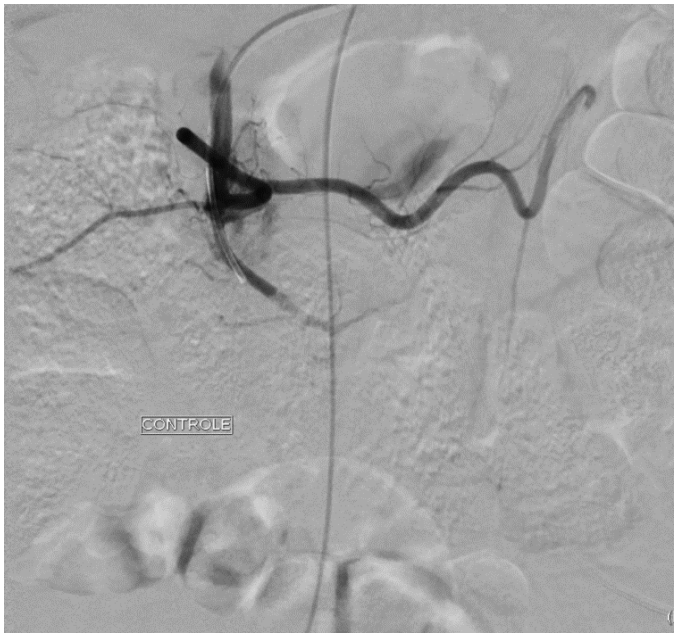

Figure 5: Picture after embolization of gastro duodenal artery

We proceeded through a midline laparotomy. We found a voluminous tumor of the pancreas that invaded the mesenteric axis.

We realized a two-stage resection of the pancreatic tumor through a cephalic duodeno pancreatectomy followed by a bilateral adrenal gland resection.

Continuity was restored by a pancreatico-gastric, hepatico-jejunal and gastro-jejunal anastomosis.

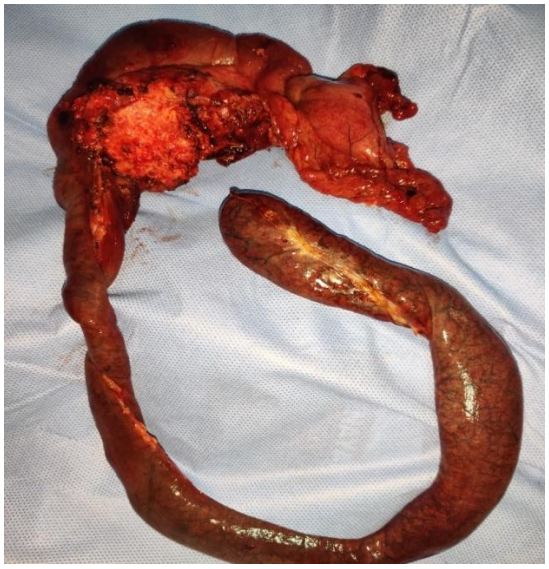

Figure 6: Anterior view of the resected pancreatic tumor

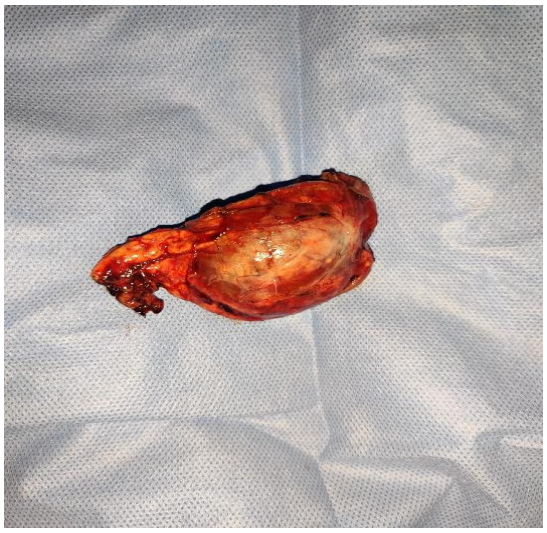

Figure 7: Left adrenal gland

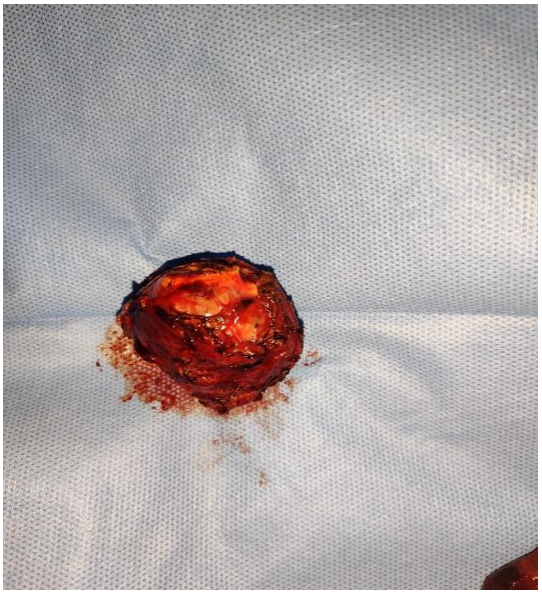

Figure 8: Right adrenal gland

DISCUSSION

VHL disease is an inherited familial cancer syndrome caused by a germline mutation in the VHL tumor suppressor gene and predisposing to a variety of benign and malignant neoplasms most frequently retinal, central nervous system and spinal hemangioblastomas, renal cell carcinoma (RCC), pheochromocytoma, and pancreatic tumors [8]. The frequency of pheochromocytoma in VHL syndrome is about 10 to 20% [9].

Families with VHL disease have been divided into two subtypes: VHL type 1 and VHL type 2, based on the likelihood of developing a pheochromocytoma. The presence of pheochromocytoma defines types 2 VHL disease.

Pheochromocytomas are rare catecholamine-secreting tumors, in VHL disease they tend to be seen at a young age and are more frequently multifocal and may be extra-adrenal [10]. In most published cases, the mean age at presentation was about 30 years, but very young cases have been described [11].

About 10% of pheochromocytomas are malignant with evidence of metastatic disease or invasion of adjacent organs [12].

Symptoms are present in approximately 50% of patients with pheochromocytoma, and they are typically paroxysmal. The classic triad of symptoms in patients with pheochromocytoma consists of episodic headache, sweating, and tachycardia [13, 14].

The diagnosis of pheochromocytoma is made based upon biochemical confirmation: Measurement of plasma or urinary metanephrines and normetanephrines is the gold standard in diagnosing pheochromocytoma [15]. followed by identifying the tumor with imaging studies.

Treatment of pheochromocytoma requires a multidisciplinary team approach that typically involves a physician (endocrinologist), surgeon, and anesthetist who are experienced in its management. If not contraindicated, surgical removal, the only curative procedure, should be performed expeditiously.

CONCLUSION

We reported a rare case of simultaneous pheochromocytoma accompanied with a pancreatic tumor. A multidisciplinary team approach followed to plan the management along its course of progression.

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