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Robotic Partial Nephrectomy for Complex Lesions of Renal Cell Carcinoma in Patients with Von Hippel-Lindau's Disease - Case Report

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Introduction

Von Hippel-Lindau (VHL) is a neurocutaneous syndrome that affects 1 in 36,000 people. The gene related to this disease is called VHL, an autosomal dominant gene with variable penetrance. It is a tumor suppressor gene located on the short arm of chromosome 3. [1] The syndrome can be diagnosed based on clinical findings. [2]

VHL most commonly causes tumors such as cerebellar hemangioblastomas and retinal angiomas. Other tumors such as pheochromocytomas and liver, renal, pancreatic or genital tract cysts are common. The risk of developing renal cell carcinoma is progressively higher with advancing age, reaching 70% at 60 years. However, manifestations tend to occur

between the ages of 10 and 30. [3] [4] [5]

The treatment of Von Hippel-Lindau's involves surgical removal of tumors such as pheochromocytomas and renal cell carcinomas. [6]

However, there is a pharmacological treatment, wich is called is Belzutifan, an HIF-2 α inhibitor, indicated for adult patients with renal cell carcinomas, central nervous system hemangioblastomas, or pancreatic endocrine tumors that have no indication for immediate surgical removal. [7]

Case Report

JRSM, 25 years old male patient, with obesity, arterial hypertension and glucose intolerance, with no use of regular medications, went through a brain MRI due to headache in 2017. The imaging showed a 7mm pituitary adenoma (08/23/17).

Computed tomography (CT) of the abdomen realized in 2019 showed renal nodules and complex cysts (bosniak iii and iv), multiple pancreatic cysts and cistoadenomas in both epididymis. Renal nodules are described above:

1. Solid lesion in the middle and lower 1/3 of the right kidney, projected to the renal sinus with 45mm
2. Endophytic confluent lesions in the lower 1/3 of the right kidney with 55mm

3. Cystic lesion with thick septa at the upper pole of the partially exophytic right kidney with 50mm

No changes were identified during the 2021 follow up.

Control CT performed in 2022 showed topical kidneys, with increased dimensions and lobulated contours. Several predominantly solid nodular lesions, with intense and heterogeneous contrast enhancement bilaterally. The largest lesion was on the right side, measuring 4.5 cm in the upper third. Another lesion was found the middle third with exuberant cystic component, measuring 6.3 cm, insinuating to the renal sinus and displacing the lower calicinal grouping. In the left kidney, the largest lesion, measuring 4.0 cm, was located in the predominantly endophytic lower third, insinuating itself into the renal sinus. The appearance of the lesions founded were compatible with primary neoplasms (renal cell carcinoma).

On 11/16/2022 he underwent lesions resection on the right kidney by robotic laparoscopy. On the firt sample, called A, were found four lesions, the greater one measuring 3.2x3.0 cm. On the second sample, called B, one lesion was found, measuring 3.3x3.2cm. The latest sample, called C, showed a single 4.3x4.0 cm lesion. The histopathological study showed a multifocal clear cell type renal carcinoma, all of them with the same characteristics. No sarcomatous pattern were detected , no capsule invasion, no perirenal fat invasion, no renal sinus fat invasion, al lesions grade 1 and with free margins. Staging pt1b nx.

Postoperative control computed tomography showed signs of right partial nephrectomy, with resection of three lesions in the middle and lower thirds of the right kidney, one of them previously composed of three clustered nodular formations. In the remaining renal bilateral parenchyma were still found cystic and solid hypervascularized nodules, with characteristics of primary neoplasia (probable clear cell type - RCC), measuring about 3.2 cm (Image x), one of them near to the nephrectomy margin. Besides, endophytic complex cyst with thickened beams and irregular solid componentes, measuring 6.4 cm (Bosniak IV), was found in the middle and upper third of the right kidney, insinuated in the renal sinus. Another cystic lesion had a hemorrhagic component with thin bilateral septa, measuring 1.3 cm (Bosniak II).

Multiple sparse cystic formations persisted throughout the pancreatic parenchyma, the largest in the pancreas body, measuring 3.1 cm (stable relative to 11/10/2022 tomography).

The solid hypervascularized formations located in the head, neck and tail of the pancreas remained similar.

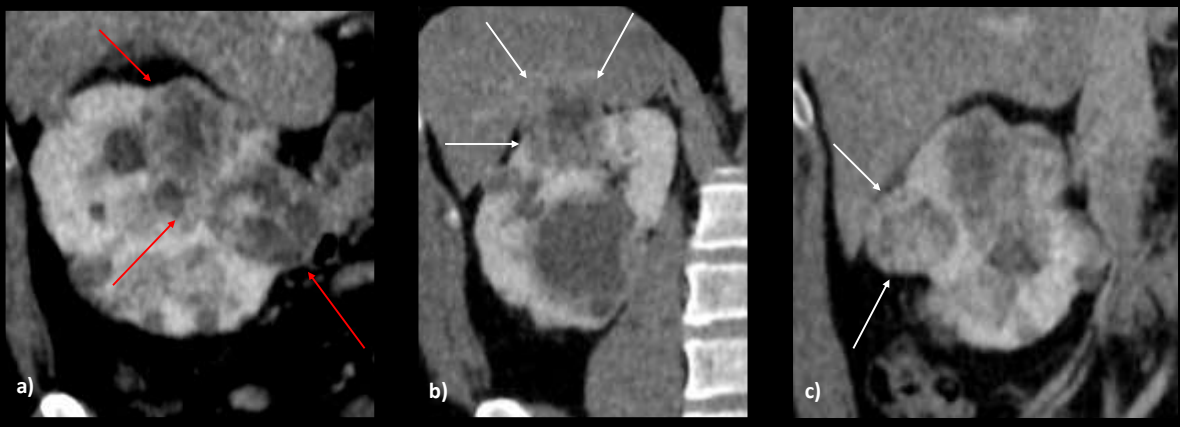

Figure 1. **Multifocal Clear cell RCC in a 25-year-old man with a diagnosis of VHL disease.** (a) Enhanced CT scan shows endophytic confluent mass (red arrows) and hypervascular exophytic nodules (white arrows) suggestive of a renal cell carcinoma.

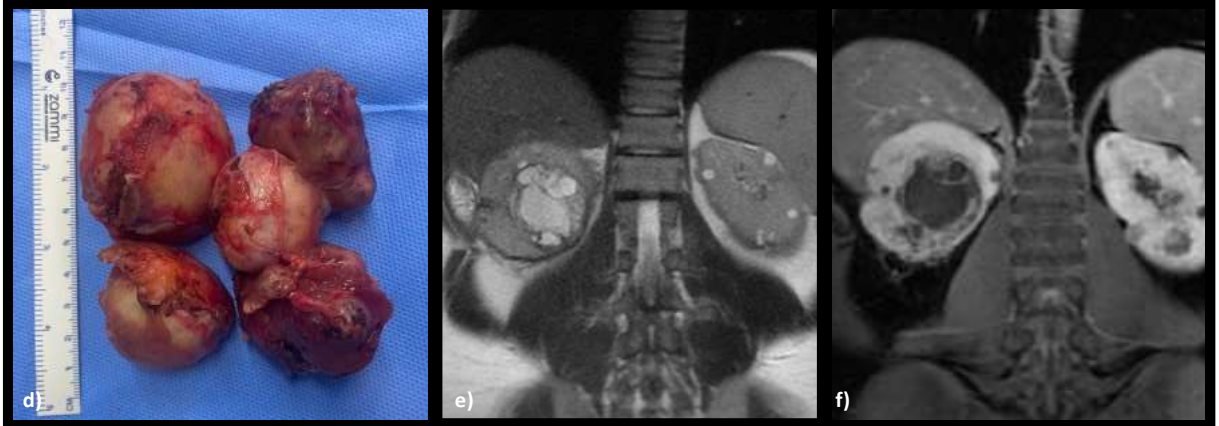

Figure 1. **Multifocal Clear cell RCC in a 25-year-old man with a diagnosis of VHL disease. (d)** Macroscopic aspect of the resection piece. **(e)** and **(f)** Postoperative MRI images showed signs of right partial nephrectomy and scar tissue in the postoperative bed.

Discussion

Renal lesions screening with abdominal imaging is recommended every two years, starting at 15 years of age. Renal cell carcinoma (RCC) rarely develops during childhood. In a study of 99 patients diagnosed with VHL's disease in childhood, no RCC was diagnosed before the age of 18. [8] The mean age of development of RCC is around 37 years old. However, it's occurrence at the age of 16 has already been described.[9] The reported case presented with lesions suspected of RCC at 22 years of age.

Because of the accuracy of abdominal MRI, it is the choice method for the surveillance of abdominal lesions. According to the latest recommendations, MRI every two years, starting of the age of 15, is the preferred approach in asymptomatic individuals. [9] Once a suspicious kidney injury is diagnosed, a urologist should perform renal surveillance based on size, characteristics, and growth of the lesion . [9]

In order to reduce the number of times a kidney is submitted to surgical procedures, the indications for surgery should be judicious. In the literature, there is a very well established correlation between tumor size and risk of metastases in patients with VHL. Active surveillance is recommended in lesion up to 3 cm on its greatest axis, suspected of CCR. When identified, treatment with nephron-sparing therapies should be indicated when it is technically feasible. Radical nephrectomy should only be performed in non-functioning kidney or in non-feasible partial nephrectomy lesions [10]. In those cases, Belzutifan could be considered (Jonasch et al., 2021).[11]

Surgery with robotic assistance has provided possibility of performing surgeries of a great complexity. In the pre-robotic era, this patient would undoubtedly be submitted to right radical nephrectomy. With the resources of technology, including reconstruction in three dimensions and perioperative ultrasonography, it was possible to resect the three largest malignant renal lesions of the right kidney.

Conclusion

Surgical removal is the key to treating most VHL tumors. The time of surgery and the choice of surgical method vary according to the clinical presentation and surgeon experience.

Challenging surgeries, such as the presented, are feasible with the support of technologies. For a young patient with a no cure genetic syndrome, the renal function preservation has a great impact on survival and life quality.

Video: https://drive.google.com/file/d/1lvrRjwjBElJ9OPvSWWLynypdGeojVC_9/view

References

1. GOSSAGE, L.; EISEN, T.; MAHER, E. R. VHL, the story of a tumour suppressor gene. Nat. Rev. Cancer, v. 15, n. 1, p. 55-64, 2015.

2. DECKER, J.; NEUHAUS, C.; MACDONALD, F.; BRAUCH, H.; MAHER, E. R. Clinical utility gene card for: von Hippel-Lindau (VHL). Eur. J. Hum. Genet., v. 22, n. 4, 2014.

3. BINDERUP, M. L. et al. Prevalence, birth incidence, and penetrance of von Hippel-Lindau disease (vHL) in Denmark. Eur. J. Hum. Genet., v. 25, n. 3, p. 301-307, 2016. DOI: 10.1038/ejhg.2016.173

4. BINDERUP, M. L. M. von Hippel-Lindau disease: Updated guideline for diagnosis and surveillance. Eur J Med Genet, v. 65, n. 8, p. 104538, 2022. DOI: 10.1016/j.ejmg.2022.104538. EPub 2022 13 jun.

5. MAHER, E. R. Von Hippel-Lindau disease. Curr Mol Med, v. 4, n. 8, p. 833-842, 2004. DOI: 10.2174/1566524043359827

6. GLASKER, S. et al. Von Hippel-Lindau disease: current challenges and future prospects. OncoTargets Ther., v. 13, p. 5669-5690, 2020.

7. GLASKER, S. et al. Von Hippel-Lindau disease: current challenges and future prospects. OncoTargets Ther., v. 13, p. 5669-5690, 2020.

8. LAUNBJERG, K. et al. von Hippel-Lindau development in children and adolescents. Am. J. Med. Genet., v. 173, n. 9, p. 2381-2390, 2017.

9. CHAHOUD, M. et al. Evaluation, diagnosis and surveillance of renal masses in the setting of VHL disease. World J. Urol., v. 39, n. 7, p. 2409-2415, 2020.

10. BAUSCH, B. et al. Renal cancer in von Hippel-Lindau disease and related syndromes. Nat. Rev. Nephrol., v. 9, n. 9, p. 529-538, 2013.

11. JONASCH, E. et al. Belzutifan for renal cell carcinoma in von Hippel-Lindau disease. N. Engl. J. Med., v. 385, n. 22, p. 2036-2046, 2021.