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Case Report

Radiofrequency ablation in Von-Hippel-Lindau disease with bilateral renal cell carcinoma: a case report

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ABSTRACT

Von Hippel-Lindau (VHL) disease is a familial form of renal cell carcinoma which manifests as bilateral renal cell carcinoma. Radiofrequency ablation is a minimally invasive technique helps in treating renal cell carcinoma with renal function preservation. We reported a case of 29 years old male with bilateral renal cell carcinoma. We treated him with left radical nephrectomy and right side multifocal RCC with Radio frequency ablation. Radiofrequency ablation definitely have a role as alternative treatment modality to nephron sparing surgical procedure for small (<4 cm) localised RCC. Further studies are needed to assess its long-term efficacy.

Keywords: VHL, Bilateral RCC, Radio frequency ablation

INTRODUCTION

Von Hippel-Lindau disease is a familial form of renal cell carcinoma, major manifestations include development of bilateral renal cell carcinoma (RCC), pheochromocytoma, retinal angiomas and hemangioblastomas of the brainstem, cerebellum or spinal cord. Renal cell carcinoma develops in about 50% of patients with Von Hippel-Lindau disease and is distinctive for early age of onset, bilateral and multifocal involvement.[1]

Radiofrequency ablation (RFA) one form of minimally invasive technique helps in treating renal cell carcinoma with renal function preservation and preventing nephrectomy. Since possibility of cancer is there in both kidneys and most of the lesions in opposite kidney are incidentally diagnosed either during evaluation or follow up, local treatment may be an option. A good oncological data is emerging for RFA.[2,3] Indications for RFA are renal cell carcinoma in a solitary kidney, synchronous primary renal tumor, patients with Von Hippel-Lindau disease.[4,5]

We present our recent experience of treating a Von Hippel-Lindau disease patient with bilateral RCC with left radical nephrectomy and on right side by RFA.

CASE REPORT

A 29-year-old male patient was evaluated for hematuria and abdominal pian, CECT abdomen showed multifocal (3 in number) RCC in right kidney (largest 3.4*3.3*3.4 cm) and a 9.3*8.2*7 cm mass lesion in left kidney. ENT and neurological examinations were unremarkable. He underwent laparoscopic left radical nephrectomy. Post op HPE was Type 1 papillary Renal cell carcinoma (grade 2) pT2bNx.

RFA was done for all three lesions in the Right kidney. Post procedure, patients renal function tests were normal and he was kept in regular follow up. 8th month Follow up imaging showed two heterogeneously enhancing lesions in the right kidney, largest at lower pole

measuring 3*2.8 cm. After tumor board discussion repeat RFA was planned. But the patient defaulted the procedure because of COVID-19 pandemic. After 4 months repeat imaging showed increase in size of lesions (largest 3.7*3.2 cm at lower pole), no new onset lesion or metastasis. Repeat RFA was done, post procedure his renal parameters were normal, patient was disease free 1 year and on regular follow up.

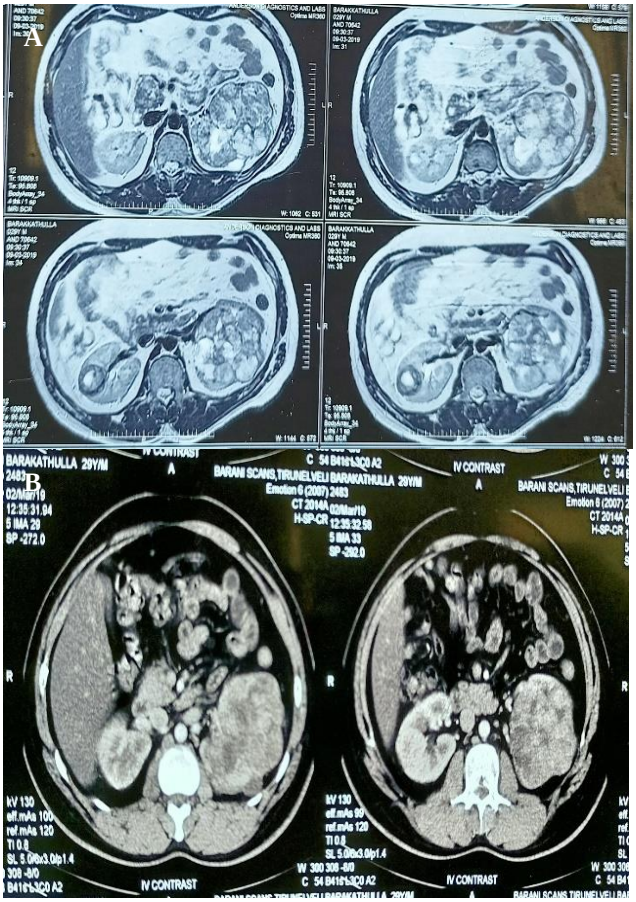

Figure 1 (A and B): Imaging shows bilateral renal cell carcinoma.

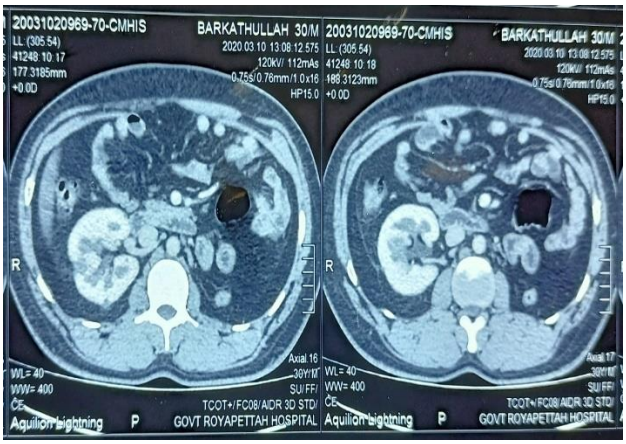

Figure 2: CT scan showing recurrent multifocal RCC in right kidney (post left radical nephrectomy status).

DISCUSSION

Renal cell carcinoma is the commonest cancer of the kidney which accounts for 3% of all cancers in adults.[6] The incidence of renal cell carcinoma has increased over the past decade due to wider use of radiological imaging and it is also related to increased cigarette smoking and obesity in the population.[3]

The familial form of RCC is von Hippel-Lindau (VHL) disease. It occurs with a frequency of 1 per 36,000 population. Major manifestations of VHL disease include the development of RCC, pheochromocytoma, retinal angiomas, and hemangioblastomas of the brainstem, cerebellum, or spinal cord. RCC develops in about 50% of patients with von Hippel-Lindau disease and is distinctive for early age at onset (often in the third, fourth, or fifth decade of life) and bilateral and multifocal involvement.[1]

Molecular genetic linkage studies in patients with von Hippel- Lindau disease eventually led to the identification of the VHL tumor suppressor gene.[7] The role of this gene at chromosome 3p25-26 as a tumor suppressor for both sporadic and familial forms of clear cell RCC has been confirmed. A critically important function of the VHL protein complex is to target the hypoxia-inducible factors 1α and 2α (HIF-1α and HIF-2α) for ubiquitin-mediated degradation, keeping the levels of HIFs low under normal conditions. Accumulation of HIF-2α leads to a severalfold upregulation of the expression of vascular endothelial growth factor (VEGF), the primary angiogenic growth factor in RCC, contributing to the pronounced neovascularity associated with clear cell RCC.[8]

Current consensus states that small renal cell carcinomas that is less than 4 cm should be treated with minimally invasive techniques preserving renal function to avoid unnecessary nephrectomy, because histological behaviour of the lesions may not be aggressive and there will be protracted natural course of events, the option of bilateral nephrectomy and renal transplant may not be attractive.[9] Radio frequency ablation has proved to be safe and an effective treatment option for small renal tumors and a good oncological outcome data emerging for radio frequency ablation.[2,3]

Percutaneous radio frequency ablation uses a high frequency alternating current within the targeted tissue to cause ionic agitation generating frictional heat which results in Cancer cell destruction when temperature exceed greater than 60°C.[4,5,10] Patients who undergo RFA require a long-term imaging follow up

The indications for RFA treatment were: non-surgical candidates (American Society of Anaesthesiologist's (ASA) score of >3, stage T1 renal tumours, renal tumours in a solitary kidney, synchronous primary renal tumour, patients with Von-Hippel-Lindau disease.[4,5]

Operator experience as well as tumor factors such as size and location play a major role in the success of RFA Overall technical success rates according to literature is around 90% according to Poon et al.[11] One of the major complications of RFA is Haemorrhage.[12] No such complication occurred during the procedure in our patient. Ureteric stricture rate is 2-3%, more chance of ureteric stricture if the tumor is located adjacent to the ureter / pelvi ureteric junction.[12,13]

Chance of bowel injury is very minimal. it is reported in literature about use of hydro dissection, which is becoming increasingly routine when the distance between treatment margin and bowel is less than 1cm.[12,14] There is 3.1% change in GFR after RFA when compared to GFR before treatment. In our patient renal function tests after procedure were normal.

5-year Overall survival rate of 85% after RFA was shown in studies by Tract et al and Zagoria et al 66 %.[15,17] Need of repeat RFA seen in 3%.[16] Our patient had recurrence 8 months after the RFA, after tumor board discussion repeat RFA was done. Zagoroa et al have shown 12 % of local pregression and 7% metastatic progression.[17]

Overall, it is reassuring to confirm that the present longer term survival outcome is within the reported range of the 'gold standard' partial nephrectomy, where the 5-year metastasis free survival after partial nephrectomy for T1 renal tumours is 86-97% and the local recurrence rate after partial nephrectomy is 1-3%.[18-20]

CONCLUSION

RFA definitely have a role as alternative treatment modality to nephron sparing surgical procedure for small (<4cm) localised RCC with good oncological outcome, reduced morbidity and faster recovery without altering renal functions. Particularly it is an attractive option in patients with early-stage bilateral disease or patients with early-stage disease in solitary kidney. Further studies are needed to assess its long-term efficacy. Anyhow the option of Radical nephrectomy with curative potential should be kept open.

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