

Synchronous Bilateral Renal cell carcinoma in Von Hippel-Lindau syndrome; treatment with Partial nephrectomy and Radio frequency ablation

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Introduction

Renal cell carcinoma (RCC) can be sporadic or familial. Von Hippel-Lindau (VHL) disease accounts for 1.6% of unselected RCC patients. Around 5% of RCC patients will have bilateral synchronous tumors (1). Fifty percent of VHL patients get RCC which is often bilateral and multifocal (2). RCC in VHL occurs at a younger age (20 - 40 years), compared to sporadic cases (40 - 60 years) (2).

Case report

A 26 year old patient had presented with sudden onset ataxia following a two month history of headache. He had blurred vision with mild weakness of the right upper and lower limbs. His brother had been diagnosed with Von Hippel-Lindau syndrome recently.

On examination he had been found to have hypertension. A Magnetic Resonance Imaging (MRI) scan of the brain had shown a haemangioblastoma in the right cerebellar hemisphere with mass effect causing obstructive hydrocephalus and tonsillar herniation.

Craniotomy and excision of the posterior fossa space occupying lesion had been done and the patient's ataxia had improved.

As the patient remained hypertensive a contrast enhanced CT of the abdomen was done which indicated the presence of three heterogeneously enhancing solid renal mass lesions suspicious of renal cell carcinoma. Two were in the right kidney measuring 1.7 cm × 1.9 cm × 2 cm and 2 cm × 2.1 cm × 2 cm (T1a N0 M0) postero-medially in the interpolar region and the lower pole respectively. One was in the lower pole of the left kidney measuring 3.3 cm × 3.9 cm × 4 cm (T1a N0 M0).

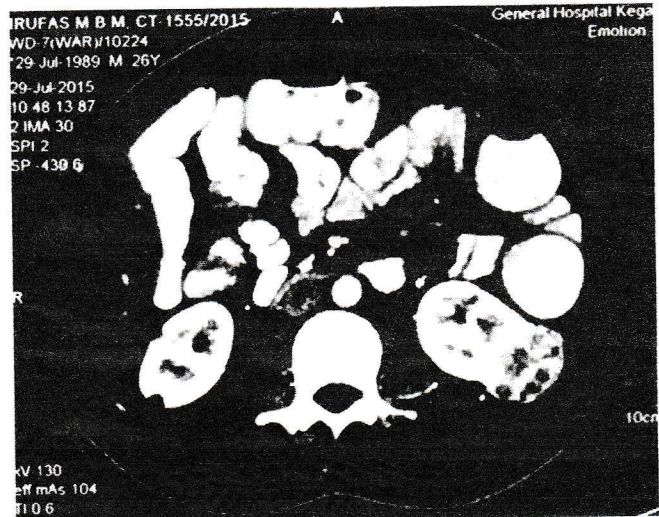


Fig. 1. Contrast enhanced CT scan of the abdomen showing bilateral renal tumors.

The left tumour was removed with a partial nephrectomy under general anaesthesia and the histology was one of a clear cell renal carcinoma of Fuhrman grade 1 with a 0.5 mm resection margin.

The patient was referred to the Consultant Radiologist for radio frequency ablation (RFA) of the right renal tumors which were carried out under local anaesthesia with Ultra Sound guidance. Repeated contrast enhanced CT scan showed residual tumors in the right kidney and the patient underwent repeated RFA and both tumors were ablated completely.

Discussion

Renal Cell Cancer is the third most common genito-urinary malignant disease.

Patients with small renal tumors have similar perioperative morbidity and outcome regardless of whether they are treated with partial or radical nephrectomy.

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