BILATERAL SYNCHRONOUS RENAL MALIGNANCIES OF VARIED HISTOLOGY (BSRMVH): IPSI-LATERAL RENAL PELVIC UROTHELIAL CARCINOMA AND CONTRA-LATERAL CHROMOPHOBE TYPE RENAL CELL CARCINOMA - A RARE CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Urothelial malignancies of the renal pelvis and renal cell carcinoma are the most common urological malignancies. However, simultaneous bilateral renal malignancies are uncommon. Even more uncommon is synchronous bilateral renal neoplasms of dissimilar histologies.

In our report, we present a very rare case of a 42 year old male, who presented with painless total hematuria with passage of clots and left loin pain. On evaluation, he was diagnosed to have urothelial malignancy of left kidney and renal cell carcinoma of the right kidney. He underwent initial left nephrostomy placement, followed by left radical nephroureterectomy and right radical nephrectomy.

This case is presented for its rarity (first ever such case published in Indian literature), the diagnostic dilemma faced, the challenges that it posed in offering an appropriate treatment and the lessons learnt.

Key words: *Urothelial malignancy, renal cell carcinoma, synchronous malignancy.*

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BACKGROUND

Renal cell carcinoma and transitional cell carcinomas are commonly encountered urological malignancies. It is not uncommon to individually make a diagnosis of renal cell carcinoma and urothelial malignancies. However, the combination, that too synchronously in contra lateral kidneys Bilateral Synchronous Renal malignancies of varied histology (BSRMVH), is a rare entity.

The overall incidence of bilateral renal malignancies is 1 to 4% for renal cell carcinomas and 3.5% for transitional cell carcinomas.[1,2] Cancers of varied histological patterns are extremely rare. So far, only 13 such cases are being reported in literature.

In a large population-based study, one of the most salient and novel finding was that the risk of bilateral renal cell cancer depends profoundly on age at first diagnosis. Patients first diagnosed before age 40years were at a 17-fold higher risk compared with patients first diagnosed at the age of 60 years or older.[3]

Patients with bilateral multifocal renal cell carcinoma are at increased risk of developing locally recurrent or de novo tumors after nephron-sparing procedures. The main reason for recurrence after nephron-sparing surgery is likely to be the presence of multifocal disease, which is identified in 4 to 25% of the radical nephrectomy specimens.[4]

In this report, we present a case of a middle aged obese male who presented with BSRMVH. The rarity of this presentation, the dilemma and challenges faced in diagnosis and management are being discussed here.

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CASE REPORT

A 42-year-old male reported with painless total hematuria with passage of amorphous clots of two months duration. He also had vague pain in the left loin pain for 15 days. The loin pain was dull aching, ill localized, continuous and non-radiating.

On examination there was vague abdominal fullness on the left loin. Left renal angle was full. There was mild tenderness on deep palpation of the left loin. Ultrasound of abdomen showed a well-circumscribed right upper polar tumour and left side grossly dilated hydronephrotic kidney with thinned out cortex.

His hemoglobin levels were 10 gms%. Total count was 13,500 cells/cub mm, with predominant neutrophilia. His temperature at presentation was 100.4F. The serum creatinine was 1.6 mg% and Blood urea levels were 48 mg/dl.

Contrast Enhanced Computed Tomography (CECT) of Kidneys showed left pelvi ureteric junction (PUJ) obstruction with Hydronephrosis and a heterodense enhancing lesion in the right renal interpolar region suggestive of Bosniak type IV cystic lesion with left paraaortic lymphadenopathy (Fig 1). Nuclear scintigraphy showed decreased uptake in the left kidney compared to the right. After an initial diagnosis of left PUJ obstruction

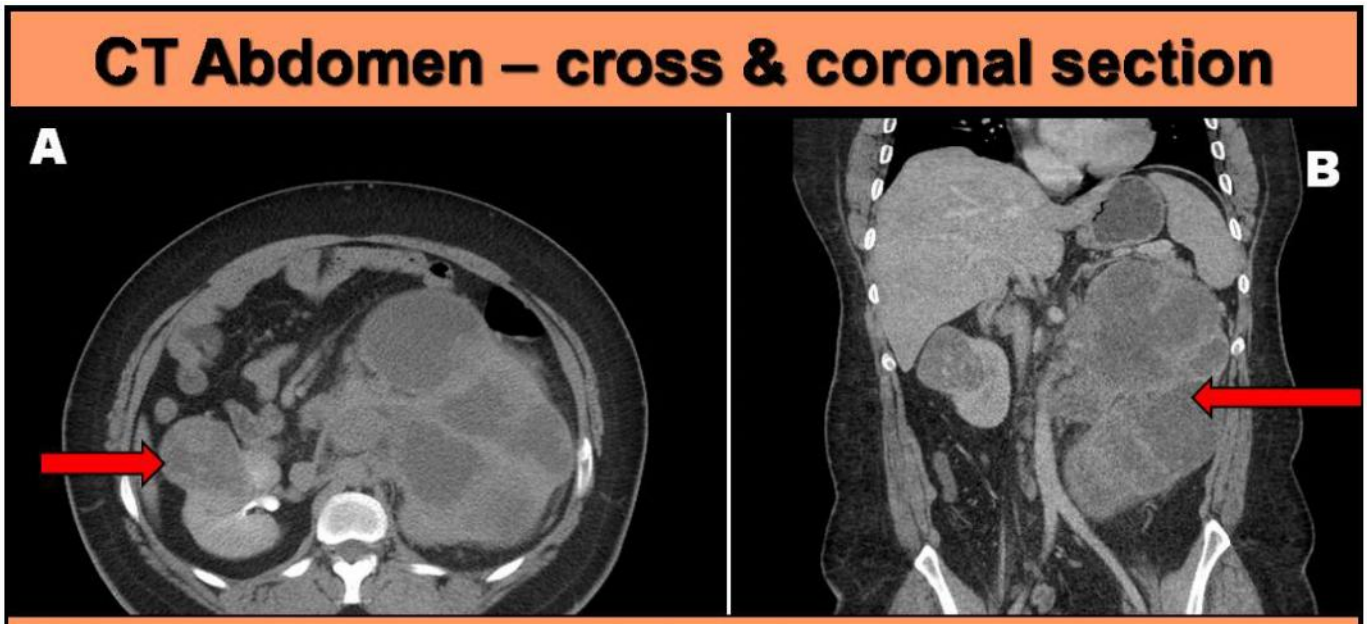

Fig 1:
A – solitary renal cell carcinoma of right kidney
B- Hydronephrotic left kidney with transitional cell carcinoma

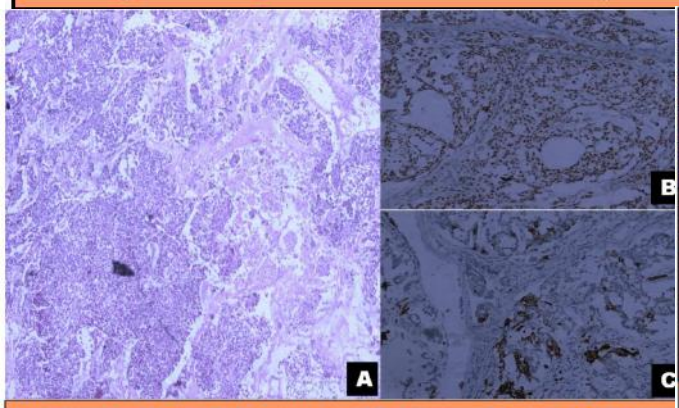

Fig 2: A – Urothelial Carcinoma High power (H & E 200 X)
B – P63 Positive
C – CD 10 negative

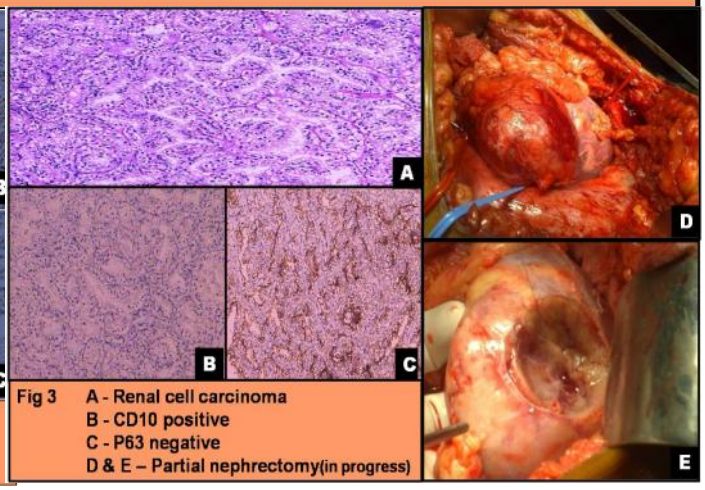

Fig 3 A - Renal cell carcinoma
B - CD10 positive
C - P63 negative
D & E – Partial nephrectomy(in progress)

and right renal cell carcinoma was made, patient was posted for left double J stenting and right partial nephrectomy. During left side stenting, retrograde pyelogram revealed an irregular upper ureteric margin and a tight narrowing at the pelvi ureteric junction that was impossible to be negotiated with a hydrophilic guide wire. Diagnostic ureteroscopy revealed only a red glow at the level of upper ureter and the renal pelvis could not be entered. Hence left percutaneous nephrostomy (PCN) was done as a temporary external diversion procedure. On PCN placement, about 1200 ml of thick brownish, gelatinous fluid was drained. Initially an infected PUJ obstruction was thought of, but the urine culture was sterile. Cytology of the drained fluid was positive for urothelial malignancy.

He subsequently underwent Left nephroureterectomy with excision of the nephrostomy tract. Histopathology revealed a high grade urothelial malignancy of renal pelvis (Fig 2 A,B,C). Patient was explained about the need of residual ureteral stump removal along with the cuff of bladder on left side, but he was not willing for the same. One month later he underwent nephron sparing surgery on the right side. In the immediate post op period his serum creatinine rose upto 1.6mg/dl, later it reached a nadir of 1.1mg/dl. Histopathology revealed chromophobe Renal cell carcinoma T1bN0 Fuhrman Grade 1 (Fig 3 A - E). Patient subsequently was offered chemotherapy using Paclitaxel and Gemcitabine.

DISCUSSION

Bilateral synchronous renal malignancies of varied histology are extremely rare. So far, all inclusive, by a thorough medline search, only a very few cases are reported in literature. If we exclude those diagnosed by autopsy, to the best of our knowledge, we are presenting the eighth overall and the first ever such case being

reported from our country. A majority of those reported earlier were from Japanese literature.

The first case of BSRMVH was reported by Villegas et al in 1967, where the diagnosis was made at autopsy.[5] Gillis et al reported the first case of BSRMVH in a live patient in 1971.[6] Joczi reported a similar case in 1976 where a radical nephrectomy was done for renal cell carcinoma and a partial nephrectomy was performed for transitional cell carcinoma.[7] However, the patient survived without any recurrence for the next 8 years. Sung Kyu reported a case of a 63 year old male who presented with left renal pelvic carcinoma and right renal cell carcinoma and subsequently underwent left nephrectomy and lymphadenectomy and planned for angio infarction of the right renal mass.[8]

Smoking has been the most important and significant predisposing factor for development of renal cell carcinomas and urothelial malignancies. Various occupational hazards, industrial chemicals, ingestion of large amounts of phenacetin and family history have all been considered to be contributory factors for development of urothelial malignancies.[9,10] However synchronous occurrence of malignancies of varied histology has been a rare entity.

The biological behavior of tumors of varied histology, the various grades and stages at which they present, presence of other co-morbid and predisposing factors and the need for a long term follow-up in such cases pose a significant challenge in prognosticating as well as managing such tumors.[11]

Our patient had a left radical nephroureterectomy and a right partial nephrectomy done. However, he needed long term follow up for the residual left ureteral stump and bladder recurrences.

The purpose of this case report is to highlight the rarity of our presentation, need for a high index of suspicion whenever any form of an abnormal fluid is drained from the kidney and the diagnostic dilemmas and challenges that one could face in making an appropriate decision regarding management.

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