Abstract:
Renal anomalies are the commonest of all congenital anomalies of urinary tract. Supernumerary kidney is an extremely rare anomaly. A unique presentation in our case made diagnosis difficult preoperatively. We report this extremely rare anomaly and its recognition and subsequent management.

Keyword: supernumerary kidney, unique presentation, rare congenital anomaly

Introduction:
The supernumerary kidney is an accessory kidney with its own collecting system, blood supply, and distinct encapsulated parenchymal mass. Till now approximately only 100 cases have been reported in literature[1]. It represents a very rare anomaly of the urinary system [2]. We here present a 25 year old man who presented with this very rare anomaly in an unique manner. This case is presented because of its very rare nature and its unusual presentation.

Case History:
A 25 year old male presented to our department with complaints of haematuria for the past 1 month, intermittent in nature, total and not associated with clots. On eliciting history he had no other urological complaints. Clinical examination of the patient including external genitalia was completely normal. Routine blood and urine investigations were also normal.

Investigations:
On ultrasonography, he was found to have huge anechoic lesion with thin walls and septae occupying almost the whole of the abdomen, which was totally unexpected [figure1]. CT scan was done. It revealed a huge cystic lesion with thin septae occupying almost the whole abdomen, predominantly the retroperitoneum, displacing all bowel loops and displacing left kidney and altering its axis [figure2]. The CT urogram revealed a blind ending ureter, like a Y shaped diverticula arising

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from the right ureter [figure3]. We did a cystoscopy which showed bilaterally normal ureteric orifices. Retrograde ureterogram was done on the right side which showed a normal right ureter with blind ending diverticula. With these findings we had provisional diagnoses of supernumerary kidney or a huge retroperitoneal cyst with blind ending diverticula.

Treatment:
We decided to explore the patient and did a midline laparotomy. On exploration there was a huge cystic lesion in the retroperitoneum. It was found to have a common ureteric stem with right ureter and a separate blood supply [figure 4]. It was displacing and compressing the left kidney. The cystic or the hydronephrotic kidney was completely mobilized from the surrounding structures and to our surprise easily too, expect for a small portion adherent to the left kidney, which was left behind and edges marsupialised. Complete excision or nephrectomy was done [2][3]. Histopathological examination showed presence of a few nephrons and fibrous tissue. So this cystic lesion had a separate vascular supply from the ipsilateral kidney and a distinct parenchymatous mass with histopathological evidence of nephrons, also without any decrease in the number of calyces in the right kidney. With these evidences we confirmed the diagnosis in this patient to be a supernumerary kidney with probably pelviureteric obstruction causing gross hydronephrosis.

Discussion:

Literature:
The supernumerary kidney is a definitive accessory organ with its own collecting system, blood supply, and distinct encapsulated parenchyma. It may be either completely separate or loosely attached to the kidney on the ipsilateral side [1]. The ureteral inter-relationships on the side of the supernumerary kidney can be variable either bifid or completely duplicated in a 1:1 ratio. It has a higher predilection for the left side. The supernumerary kidney is reniform but generally smaller than the main ipsilateral kidney. In almost half of the reported cases, the collecting system is severely dilated with thin parenchyma suggesting obstruction [4][5]. Although this anomaly is present at birth, it is rarely symptomatic until early adulthood. The average age at diagnosis was 36 years. Pain, fever, hypertension, and a palpable abdominal mass are the usual presenting complaints. Urinary infection or obstruction, or both, are the major conditions that lead to an evaluation. Usually it is diagnosed by imaging. Occasionally it is not accurately diagnosed until the time of surgery or at autopsy [1]. This case is being presented for its very rare nature and unusual presentation with haematuria probably due to compression and congestion of the left kidney.

References:
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figure1- usg showing large anechoic le-
son with thin septae occupying almost 
the whole abdomen

figure2- ct image showing a large 
hypodense lesion with thin septae 
displacing bowel loops and shifting 
left kidney

figure3- ct urogram showing y 
shaped bifid ureter with displaced, 
rotated kidneys

figure4- operative pictures

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