

## Renal Dysplasia in an Adult Female - A Case Report

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### ABSTRACT

Renal dysplasia is a rare disorder characterised by abnormal differentiation in kidneys resulting in abnormality in structural organisation along with ill-developed metanephric elements. We present a case of 45 year female presenting with right loin pain. CT scan revealed small right kidney with gross dilation of pelvic calyces & proximal ureter, along with calculus in mid ureter. Histopathological examination of the right kidney showed renal dysplasia with features of chronic pyelonephritis.

**Key words:** Renal dysplasia, Adult female, Pyelonephritis.

### INTRODUCTION

Dysplastic kidney is a rare congenital malformation of the kidney which is classified as unilateral, bilateral or segmental. Bilateral multicystic dysplastic kidney is often incompatible with survival. They result from abnormal induction of metanephric mesenchyme by ureteral bud. [1] Renal dysplasia is common in males and has male to female ratio of 1:4:1. [2] Condition is usually diagnosed during antenatal period due to advances in imaging technology. [3] Dysplastic kidney if persists till adulthood may undergo calcification. Features of chronic pyelonephritis may develop due to ascending infection. We present 45 yrs female presenting with renal dysplasia and chronic pyelonephritis which is rare in occurrence.

### CASE HISTORY

A 45 years female presented to urology department with right loin pain for 6 months. General & systemic examinations were unremarkable. Local examination revealed tenderness in right lumbar region. Ultrasound abdomen showed tubular cystic

lesion extending from right renal fossa upto lower abdomen with kidney not separately seen -likely to be chronic ureteric obstruction sequence with atrophic renal parenchyma. Computerised tomography (CT scan) revealed small sized right kidney with gross dilatation of pelvic calyces and proximal ureter. 3x3mm calculus noted in mid ureter. Left kidney was normal in size. 8x6mm calculus was noted in lower calyx. Pelvicalyceal system and ureter were normal.



Figure 1: Small kidney (left) with cystic areas and dilated ureter (right)

Right sided nephrectomy was done and specimen was sent for histopathological

examination. Grossly we received nephrectomy specimen with attached dilated ureter. Kidney was measuring 3x1.5x1cm & ureter was measuring 12cm. Cut section showed few cystic areas. Cortico medullary junction cannot be made out (Figure 1).

Microscopic examination showed structure of kidney with disorganised architecture along with few normal tubules. Some of the tubules were dilated showing thyroidisation and were lined by flattened epithelium. Few foci show primitive tubules lined by ciliated columnar epithelium with cuffing of mesenchymal tissue (Figure 2). One foci showed island of cartilage (Figure 3). Interstitial stroma showed extensive areas of hyalinization. Few chronic inflammatory cells and congested blood vessels were noted.

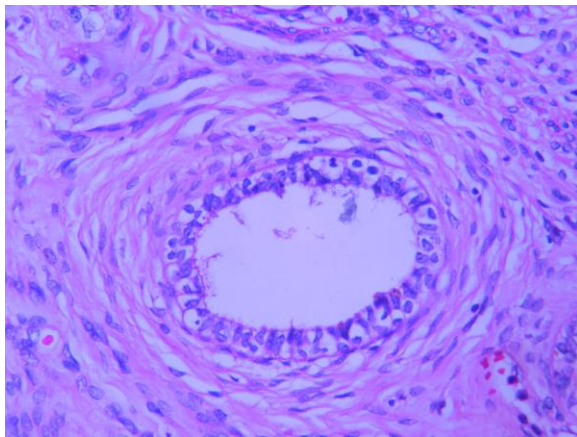


Figure 2: Primitive tubule lined by ciliated columnar epithelium with cuffing of mesenchymal tissue (H&E,X400)

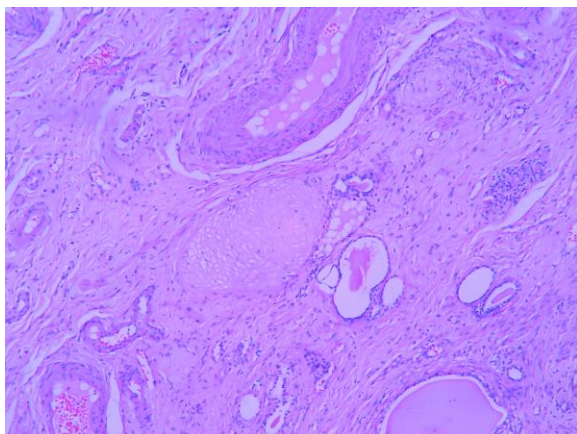


Figure 3: Renal parenchyma with thyroidisation of tubules and a foci showing island of cartilage (H&E, X100)

## DISCUSSION

Renal dysplasia is abnormal differentiation of renal parenchyma that is

characterised by undifferentiated tubular structure surrounded by primitive mesenchyme. Sometimes accompanied by divergent differentiation resulting in heterotopic elements such as cartilage. Distinct patterns of dysplasia include multicystic dysplasia, hypoplastic dysplasia, Aplastic dysplasia, obstructive dysplasia & diffuse dysplasia.

Multicystic renal dysplasia is not caused by genetic abnormality & is rarely familial. Aplastic dysplasia & diffuse dysplasia are hereditary and accompanied by congenital abnormality in multiple organs. The gene involved is Hepatocyte growth factor and PAX2. [4]

Multicystic dysplastic kidneys are large and are detected as flank mass immediately after birth. But the small multicystic kidneys are not apparent for many years and are detected during post-mortem examination. Unilateral renal multicystic dysplasia occur in approximately 1 in 5000 live births and are detected in 0.5 percent of autopsies. [5] Diffuse dysplasia usually is bilateral and often forms a component of syndromes of congenital anomalies having an autosomal recessive inheritance. Obstructive dysplasia and sometimes hypoplastic dysplasia may manifest as urinary retention in neonates with large hypertrophied bladder. Obstructive renal dysplasia is associated with congenital urinary tract obstruction along with the anomalies like posterior urethral valves, urethral atresia, mega ureter syndromes & prune belly syndrome. Hydronephrosis & pyelonephritis are frequent complications. Aplastic renal dysplasia presents as rudimentary dysplastic kidneys. [6]

Grossly dysplastic kidney may be abnormally large or very small. Cut section reveals either loss of lobular architecture with absent or distorted calyces & medullary pyramids. Unilateral multicystic dysplasia may undergo involution overtime. Microscopically the presence of primitive collecting ducts or tubules in a loose mesenchyme in medulla is the most

common and consistent histological feature. Epithelium of the duct can be columnar or ciliated columnar. A condensation of mesenchyme collars about the ducts is often-striking feature. In such collars smooth muscle cells with fibrils can also be presents. In the medulla, islands of undifferentiated blastema similar to that seen between the lobules of the fetal kidney may be present.

Differential diagnosis includes multicystic nephroma and ureteropelvic junction obstruction. In our case the patient was elderly female with renal calculi. She had the features of chronic pyelonephritis, probably due to obstruction and ascending infection. Mesenchyme in collagenous tissue of a pyelonephritis scar in which scattered tubules are present can sometimes simulate dysplasia elements. However the epithelium of such tubules will be more flattened, regenerative type & the tubules will be surrounded by collagen. [7] In our case we had island of cartilage also along with primitive tubules which helped in the diagnosis.

The treatment of choice is nephrectomy. Antibiotics should be given if the patient has vesicoureteral reflux in the contralateral kidney to prevent the development of pyelonephritis. Patient should have lifelong follow up as the patient may develop renal insufficiency proteinuria and hypertension in later life. If the patient has only one functional kidney, then dietary modifications like Low salt diet and low fat diet is advised. [8]

## CONCLUSION

We present a case of renal dysplasia due to its rarity of occurrence in an adult

female. This was an incidental finding in patient who has undergone surgery for non functioning right kidney probably due to obstructive pathology. Early diagnosis is essential as it may prevent the damage to the other kidney. Lifelong follow up of patient is needed.

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