International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Ask-Upmark Kidney: A Report of 2 Cases

Shweta Prasad, Amrit Kaur Kaler*, Shameem Shariff
Department of Pathology, MVJ Medical College and Research Hospital, Bangalore.

*Correspondence Email: amrit_kaler@yahoo.co.in

Received: 15/12//2012 *Revised: 16/01/2013* *Accepted: 25/01/2013*

ABSTRACT

Ask-Upmark Kidney or Renal segmental hypoplasia is a rare congenital kidney disorder associated with hypertension. It was first described by Eric Ask-Upmark in 1929 as congenital unilateral renal hypoplasia. This is a report of 2 cases detected over a 5 year period. The first case is of an 11 month old female who presented with repeated haematuria. Investigations revealed a small, non-functioning right kidney, right vesicouretric reflux, grade 2 and hypertension. A right nephrectomy was indicated. Histopathology revealed renal dysplasia (subtotal/ hypoplastic) with associated uretric atresia. The renal parenchyma showed features of reflux nephropathy. The second case was a post-mortem finding in a 51 year old female who died after a road traffic accident. The lower lobe of the left kidney appeared hypoplastic and segmented with dilated calyces at the hilum, the ureter was dilated. Microscopy showed evidence of early segmental hypoplasia with patchy acute tubular necrosis. Patients usually come to attention due to either hypertension or recurrent urinary tract infections. The condition is more frequent in females and usually unilateral. The kidney is characterised by capsular grooves and fewer pyramids. The hypoplastic areas lack glomeruli and possess thyroid like tubules and thick walled arteries. The reported cases showed similar findings. The pathogenesis is controversial, attributed to vesicoureteric reflux with intrarenal reflux or localised developmental arrest. The scarring could be due to reflux of infected urine or the Tamm-Horsfall protein. Hypertension is attributed to abnormal renin secretion and a nephrectomy usually normalises the blood pressure.

Key Words: Ask-Upmark kidney, Renal segmental hypoplasia, Hypertension

INTRODUCTION

Renal segmental hypoplasia also known as the Ask-Upmark kidney is a congenital kidney disorder associated with hypertension. It was first described by Eric Ask-Upmark in 1929 who reported it as congenital unilateral renal hypoplasia. Habib et al. reported 9 more cases of this rare condition in 1965 and called it "segmental hypoplasia of the kidney". [1] This condition is seen more frequently in females below the age of 12 years. [1] Headache is the leading symptom either in isolation or associated with hypertensive encephalopathy. The present study deals with a report on this rare condition seen in nephrectomy specimens over a period of 5 years.

MATERIAL AND METHOD

Nephrectomy specimens were collected over a 5 year period at the Department of Pathology at MVJ Medical

College and Research Hospital. The gross and microscopic features of these specimens were analysed and correlated with the clinical history. Two cases of Ask-Upmark kidney were identified in the 5 year period.

Case 1

An 11 month old female presented with haematuria. Upon investigation the following results were obtained: WBC count was 10.8 x 10^9 cells/L, examination of the urine showed numerous WBCs and pus cells, RBC +++, urine culture was positive for E. coli. Urea, creatinine levels were normal. The patient was treated with antibiotics. She returned with the same complaints after a period of time. An abdominal ultrasound was done which revealed the right kidney measuring 3.5 cm in length with a thin cortex and no evidence of hydronephrosis. A tortuous and dilated right ureter was seen. The left kidney was 8.3 cm in length with evidence of compensatory hydronephrosis. A micturating cystourethrogram showed right vesicouretric reflux, grade 2. A Dimercaptosuccinic acid (DMSA) scan showed a non-functioning right kidney with a normal left kidney function, a 98% split function. Upon general examination the child was found to be hypertensive.

With the above scan results a right nephrectomy was performed. Post nephrectomy, the blood pressure was normalised.

Macroscopic examination: The excised right kidney was small and atrophic measuring 3x2x1.5cm; it was attached to 11 cm of ureter. The ureter appeared atretic and narrowed in the proximal 3 cm of its length and dilated and tortuous in the lower 8cm.

On sectioning the kidney showed a solid appearance with no obvious cysts or dilatation of the pelvicalyceal system. Distinction between cortex and medulla was ill defined.

On passing a probe through the lower part of the ureter it could not be passed beyond a stricture situated 1.2 cm from the pelviureteric junction. The dilated portion of the ureter showed a mucosal fold which corresponded with the tortuosity.

Microscopic examination: The sections showed renal tissue with glomeruli, tubules and patchy dense areas of lymphocytic infiltration. Capsular thickening and scarring was observed. The parenchyma in the areas of scarring showed many thick walled blood vessels in a tortuous fashion extending through the entire thickness of cortex and medulla. Glomeruli showed normal appearances, sclerosis and periglomerular fibrosis. The tubules were normal in most places with focal calcification, elsewhere they were dilated with eosinophilic casts. The medulla contained concentric collars of mesenchymal tissue around ductular structures which were lined by cuboidal to stratified epithelium. The ureter showed proximal constriction and distal dilatation along with inflammation of the mucosa.

Impression: Renal dysplasia (subtotal/ hypoplastic) with associated uretric atresia. The renal parenchyma showed features consistent with reflux nephropathy.

Case 2

A female patient aged 51 years was brought to the hospital after a road traffic accident and was proclaimed dead within a few minutes. Her relatives were motivated to donate her kidneys. Both kidneys were removed. The right kidney appeared normal in size, shape and measurement while the left showed the lower pole to be hypoplastic with a dilated ureter. The right kidney was taken for transplantation. The examination of the left kidney is as follows.

Macroscopic examination: The kidney measured 10x6x3 cm, weighed 140 g and appeared flattened. Hilum showed a dilated vein 10cm long, a dilated ureter 4.5cm long

and an adjacent artery. Lower pole of the kidney appeared segmental. Cut section showed dilated calyces at the hilum.

Microscopic examination: The renal parenchyma appeared dilated and thickened blood vessels are seen traversing it with perivascular fibrosis. The arteries showed prominent and thickened intima and media, the veins showed thickening of tunica media. The renal tubules showed patchy acute tubular necrosis (ATN), a few showed thyroidisation with inspissated secretions. There was no evidence of inflammation.

Impression: The kidney showed evidence of early segmental hypoplasia with patchy ATN.

DISCUSSION

Ask-Upmark kidney was first reported in 6 patients, 5 of whom were adolescents and presented with malignant hypertension and congenital segmental hypoplasia of the kidney. [2]

Most of the patients draw medical attention because of their hypertension [1-6] or sometimes a history of recurrent urinary tract infections. This condition is seen more frequently in females. [1] Although the condition is usually unilateral, bilaterally asymmetrical segmental hypoplasia has also been reported. [7]

Grossly the hypoplasia is characterised by capsular grooves and decrease in number of pyramids. [2] Similarly our cases presented with small, atrophic and segmented kidneys.

On microscopy the hypoplastic areas contain no glomeruli but have thyroid like tubules and thick walled arteries. [2] Our cases showed similar findings with thickened vessels and thyroidisation of the tubules.

The pathogenesis of the Ask-Upmark kidney is rather controversial. Some authors attribute the principle cause to vesicouretric reflux (VUR) with intrarenal reflux, however not all patients demonstrate a VUR at the time of diagnosis. [3,7] The possibility of localised developmental arrest was also suggested in earlier descriptions with the presence of renal dysplasia in the abnormal segments; however this alteration in metanephric development could probably be a consequence of an intrauterine reflux. [3,7]

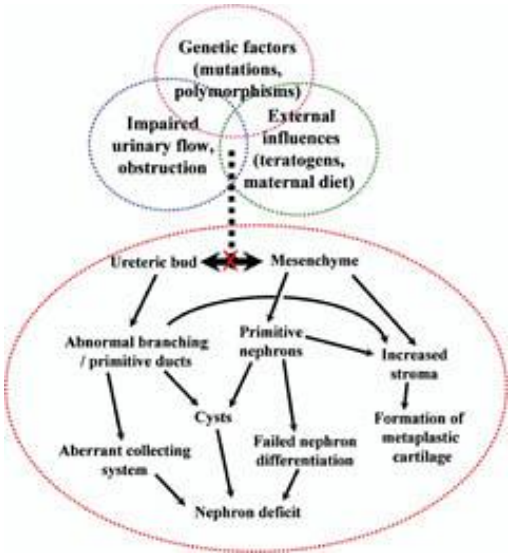

Fig 1: Mechanisms of developmental kidney disease

The deep scars seen in the Ask-Upmark kidney tend to be in the midzone of the kidney, as they represent intrarenal reflux into a single lobe. The scarring can be either due to reflux of infected urine or due to Tamm-Horsfall protein in case of sterile intrarenal reflux. The protein may induce obstruction as well as its influx into the renal interstitium may set up a chain of reflux nephropathy. [9]

The hypertension frequently seen in patients with Ask-Upmark kidney is proposed to be due to abnormal renin secretion. [2-5] A nephrectomy of the affected kidney usually normalises the blood pressure. [1-6] The child in our case had a similar result.

Ask Upmark kidney need to be differentied from chronic pyelonephritis or ectopic segmentation of vascular origin. The

reduction in the size and weight of the affected kidney are rarely associated with inflammation or vascular change. The renal artery is small and this is not secondary to atherosclerotic changes, since the walls of the renal artery in this nephropathy are always normal. The segments affected may be in the cortex and medulla of one or several renal lobes, but they do not have the fan-like distribution characteristic of chronic pyelonephritis and renal infarctions. [1]

CONCLUSION

The association of a unilateral small kidney with hypertension is attributed to pyelonephritis or primary hypoplasia and the early description of Ask Upmark and Habib et al should be taken into consideration.

REFERENCES

1. Rosenfeld JB, Cohen L, Garty I, Ben-Bassat M. Unilateral renal hypoplasia with hypertension (Ask-Upmark kidney). BMJ.1973; 2:217–219.
2. Sobel JD, Hampel N, Kursbaum A, Adler O, Gellet B. Hypertension due to Ask-Upmark Kidney. Br J Urol. 1977 Nov; 49(6):477-80.
3. Babin J, Sackett M, Delage C, Lebel M. The Ask-Upmark kidney: a curable cause of hypertension in young patients. J Hum Hypertens 2005 Apr;19(4):315-6.
4. Sugimoto T, Tanaka Y, Nitta N, Uzu T, Nishio Y, Kashiwagi A . Renal segmental hypoplasia, Ask-Upmark kidney, in a patient with adult-onset hypertension. Intern. Med. 2006;45(19):1101–2.
5. Dumas R, Baldet P, Rolin B, Jean R. Hypertension and segmental renal hypoplasia causing a syndrome of haemolysis and uraemia. Arch Dis Child 1981;56:403-404.
6. Shindo S, Bernstein J, Arant BS. Evolution of renal segmental atrophy (Ask-Upmark kidney) in children with vesicoureteric reflux: Radiographic and morphologic studies. J Pediatr. 1983;102:847–854.
7. Marwali MR, Rossi NF. Ask-Upmark kidney associated with renal and extrarenal arterial aneurysms. Am J Kidney Dis 1999;33:e4.
8. Barness-Gilbert E, Lacson A. Developmental abnormalities of the kidney. In: Mills SE, Carter D, Greenson JK, Oberman HA, Reuter VE, Stoler HM. Sternberg's Diagnostic Surgical Pathology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins; 2004. p. 1851-52.
9. Hodson CJ. Reflux Nephropathy: A Personal Historical Review. AJR 1981;137: 451-62.

How to cite this article: Prasad S, Kaler AK, Shariff S. Ask-Upmark kidney: A report of 2 cases. Int J Health Sci Res. 2013;3(1):61-64.

****