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PUJ BLOCK WITH DUPLEX COLLECTING SYSTEM IN HORSESHOE KIDNEY

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ABSTRACT

Though horseshoe kidney is a common congenital anomaly, duplication of collecting system in both kidneys is rare. Only 5 cases of the duplex collecting system in horseshoe kidney have been reported till date. PUJ obstruction is common in a horseshoe kidney. This is the first reported case of PUJ obstruction of the duplex collecting system in a horseshoe kidney.

KEYWORDS: Fused Kidney (MeSH unique ID: D000069337), Pelviureteric Junction Obstruction (MeSH unique ID: C537373), Hydronephrosis (MeSH unique ID: D006869), Kidney (MeSH unique ID: D007668).

INTRODUCTION

Amongst the different renal anomalies, horseshoe kidney is the most common congenital anomaly of urinary tract with an incidence of 1 in 400 births. [1] It is found in 90% of all renal fusion anomalies and is noted in 0.25% population. [2] Duplex collecting systems are one of the common congenital anomalies with an incidence of 0.8%. [3] PUJ obstruction is most commonly associated finding in horseshoe kidney seen in up to 35% patients. [4] Duplex collecting system in horseshoe kidney and PUJ obstruction in upper pole moiety in horseshoe kidney with the duplex collecting system is exceedingly rare.

CASE REPORT

A 35 year old female patient presented with pain in the left lumbar region with vomiting. Ultrasonography of abdomen showed horseshoe kidney, moderate to severe hydronephrosis in left kidney with thinning of renal

parenchyma. Multi detector computed tomography of the abdomen showed horse shoe kidney with normal renal parenchyma connecting lower pole of both kidneys across midline (Figure 1a, b). Both kidneys showed double moiety (Figure 2a, b). Calyces in the lower pole moiety of both kidneys were directed medially. Upper pole of left kidney showed marked hydronephrosis with dilated extra-renal pelvis of size 27 x 23 mm suggestive of PUJ block (Figure 1a, 2, 3b). Renal parenchyma in the upper pole of the left kidney was thinned out due to marked hydronephrosis. It showed slight delayed function with puddling of contrast in hydronephrotic sac. Otherwise, both kidneys showed normal function. Single ureter was noted on either side with normal insertion at urinary bladder. Diagnosis of horse shoe kidney with double moiety in both kidneys with PUJ block in the upper pole moiety of the left kidney with marked hydronephrosis was given.

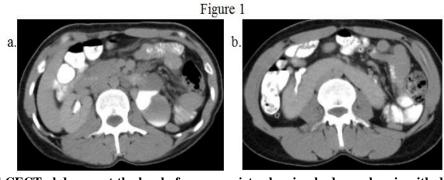


Figure 1a: Axial CECT abdomen at the level of upper moiety showing hydronephrosis with dilated extrarenal pelvis due to PUJ block in upper pole moiety of left kidney with puddling of contrast. Figure Figure 1b: Axial CECT abdomen at the level of lower moiety showing normal renal parenchyma connecting lower pole of both kidneys anterior to aorta and IVC across midline.

www.ejpmr.com 476

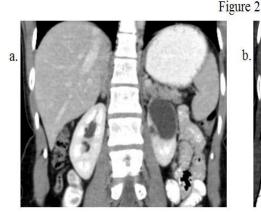




Figure 2a: Coronal CECT abdomen (nephrographic phase) showing double moiety in both kidneys with hydronephrosis in upper pole moiety of left kidney. Figure 2b: Coronal CECT abdomen (excretory phase) showing double moiety in both kidneys with hydronephrosis in upper pole moiety of left kidney showing delayed excretion and puddling of contrast.

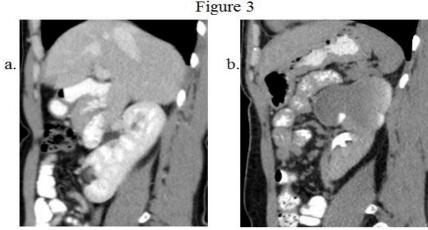


Figure 3a: Sagittal CECT abdomen (nephrographic phase) showing double moiety in right kidney. Figure 3b: Sagittal CECT abdomen (excretory phase) showing double moiety in left kidney with PUJ block in upper pole moiety.

DISCUSSION

Amongst the different renal anomalies, horseshoe kidney is the most common congenital anomaly of urinary tract with an incidence of 1 in 400 births. [1] It is found in 90 % of all renal fusion anomalies and is noted in 0.25 % population. [2] It also shows male predominance. [1] It consists of two different functioning kidneys on either side of the midline lying vertically, connected by an isthmus of functioning renal parenchyma or infrequently a band of fibrous tissue at their lower poles across the midline of the body. [2] Fusion in these cases may be in the midline (symmetrical, 10% cases) or lateral (asymmetrical para-midline, 90% cases) position. 95 % cases show fusion through the lower poles.

Duplex collecting systems are one of the common congenital anomalies with an incidence of 0.8%. Duplex system is defined as the kidney with two pelvicalyceal systems, which may have either solitary or bifid ureter (partial duplication) or dual ureter draining distinctly into the urinary bladder (complete duplication), with a solitary renal parenchyma which is drained by two

pelvicalyceal systems. $^{[5]}$ In 20% cases it is bilateral and more prevalent in women. Only 5 cases of the duplex collecting system in horseshoe kidney have been reported in literature. $^{[6]}$

Axis of kidneys in horseshoe kidneys are more vertical than normal with their lower poles deviated medially than upper poles. Thus, axis and rotation of fused kidneys are abnormal. Renal pelvis and ureters are located more anteriorly or laterally than normal. Ureters cross anterior to the isthmus.^[1] Ureter usually inserts high on renal pelvis and passes anterior to the isthmus and lower pole probably due to incomplete renal rotation with resultant abnormal upper ureteral angulation. Hence, the incidence of hydronephrosis, PUJ block, infection and stone formation is high. However, lower ureter usually inserts into the urinary bladder normally and is rarely ectopic. [2] PUJ obstruction is most commonly associated finding in horseshoe kidney seen in up to 35% patients.^[4] This is the first reported case of the PUJ block with the duplex collecting system in a horse shoe kidney.

www.ejpmr.com 477

CONCLUSION

Horseshoe kidney, duplex kidneys and PUJ block are usually clinically silent. They may present with recurrent UTI and abdominal pain. PUJ block in upper pole moiety with marked hydronephrosis and thinning of renal parenchyma may mimic simple renal cyst. Awareness of PUJ block in the duplex collecting system in horseshoe kidney is hence essential.

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www.ejpmr.com 478