Pelviureteric Junction Obstruction in Duplex System

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Abstract. An unusual case of a PUJ obstruction in the upper moiety of a duplex kidney is presented. PUJ obstruction in duplex moieties is commonly seen in the lower moiety ureter and is usually due to extrinsic compression by a crossing vessel. In our case there was a long stenotic segment of the ureter draining a functionally insignificant and infected segment of the kidney. The opposite kidney was hypoplastic and non functional. An upper polar hemi nephrectomy was performed. [Indian J Pediatr 2002; 69 (8) : 717-719]

Key words : Pelviureteric junction; Moiety; Ureter

The incidence of Pelviureteric junction (PUJ) obstruction in duplex systems is not known. PUJ obstruction if present is usually confined to the lower moiety and is most of the times due to an extrinsic compression by a lower polar vessel. Occasionally vesicoureteric reflux with secondary PUJ obstruction may be associated. Upper moiety PUJ obstruction is very rare. There is an increased incidence of contralateral structural anomalies. We report an unusual case of PUJ obstruction in the upper moiety of a partially duplicated system of a solitary functioning kidney in an adolescent, whose opposite kidney was hypoplastic.

CARE REPORT

An 11-year-old boy presented with the complaints of intermittent pain in the left renal angle and fever for the last 8 months. Ultrasonography of the abdomen showed multiple gall bladder calculi, absent right kidney and a duplex and hypertrophied left kidney with hydrenephrosis of the upper moiety. Intravenous Pyelography showed no visualisation of function on the right side. The left kidney was found to have a duplex system with PUJ obstruction of the upper moiety. The PUJ was seen as a stenotic segment about 2.5 cm in length. The lower moiety was normal. Both the ureters were united at the level of upper border of sacroiliac joint (Fig 1). A Micturating Cystourethrogram did not show evidence of vesicoureteric reflux. Retrograde pyelography showed normal ureter with hypoplastic kidney on the right side (Fig 2). Left side showed features of obstruction in the upper moiety. DTPA radioisotope scan showed non - function of the right kidney. There was evidence of obstruction and 15% contribution of function by the upper moiety of the left kidney. CT scan showed an intrarenal PUJ with the entire stenotic ureter lying intrarenally (Fig 3). Surgical exploration revealed a large kidney with the upper ureter entering the renal substance in between the fanned out renal pedicle. The cortex of the upper moiety

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Fig 1. Intravenous urography. note the long stenotic segment of upper PUJ
was badly scarred. Aspirate from the kidney was purulent, indicative of persistent infection. Hilar dissection was attempted but it was not feasible to reach the PUJ of the upper moiety because of the deep intrarenal location. An upper moiety heminephrectomy was performed. The ureter was ligated at the level of lower moiety PUJ. Post-operatively, the recovery was uneventful. On follow-up, for the last one year the patient has normal renal functions.

DISCUSSION

Duplication of the renal collecting system is a common upper urinary tract anomaly. PUJ obstruction in the partially or completely duplicated systems is usually seen in the lower moiety. This predilection for the lower moiety is related to: (i) High incidence of reflux in the lower moiety ureter causing secondary PUJ obstruction; (ii) Presence of a lower polar crossing vessel; (iii) Typical architecture of the duplex systems wherein the upper pole subtends a single infundibulum without a true pelvis, and the lower pole accounts for two-thirds of the parenchyma and contains at least two major calyces and a true renal pelvis.

Obstruction of the upper moiety has been rarely reported whereas a case of complete duplication has been reported. The obstruction was found to be due to compression of the PUJ by an artery against the posterior branch of renal vein. Ureterolysis and repositioning of the ureter anterior to the crossing vessel was done. In another case of PUJ obstruction of the upper moiety, a fibrous band was found as the cause of compression. Aaronson and Chir reported a case of an infant with Turner syndrome and an Upper moiety PUJ obstruction. In another report, two patients with upper moiety obstruction had incomplete duplication, while one had complete duplication. The cause of obstruction in these cases was intrinsic narrowing of the ureter.

In the present case we have dealt with a situation where the upper moiety had a PUJ obstruction which had a difficult and unapproachable anatomy at the PUJ. Even with 15% function of this unit, a corrective surgery would have been a preferred option if technically achievable. But the deep intrarenal (Fig 3) location and the long length of the stenotic segment of the PUJ made it impossible for attempting an upper to lower moiety pyelopyelostomy or a pyeloplasty. In view of hypoplasia of the opposite kidney, a better and predominantly functioning lower moiety and an infected and poorly functioning upper moiety, we performed an upper moiety heminephrectomy.

Thus to conclude, this is a rare case of a duplex system with upper moiety PUJ obstruction and hypoplasia of opposite kidney. Corrective surgery such as a pyelopyelostomy or a pyeloplasty was not performed due to associated infection and difficult anatomy and in the presence of poor function of the affected moiety, a heminephrectomy was the most justified option.

REFERENCES

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