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Proximal ureteral atresia in a duplex kidney with preserved renal function: A case report and review of literature

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ABSTRACT

Ureteral atresia is a rare congenital anomaly and is usually associated with renal dysplasia. Distal atresia is more common and other associated urinary anomalies are rare. We report a case of an eight year old boy who presented with left flank pain and vomiting. Ultrasound abdomen and pelvis and renal scintigraphy were suggestive of hydroureteronephrosis with obstructed drainage. He was suspected to have ureteric stricture and taken up for surgery. Cystoscopy and retrograde pyelogram revealed an incomplete duplication with lower moiety ureter ending blindly. Exploration of left renal fossa showed a duplex anomaly with normal upper moiety and hydronephrosis of lower moiety with a ureteric stump. Ureteropelvic anastomosis was done between the dilated renal pelvis and the blind ending ureter. Patient was doing well post operatively with good function in both moieties. Review of literature shows only two previously reported cases of proximal ureteric atresia. Preservation of renal function in an older child has also been reported only once. Ureteral atresia is usually diagnosed intra-operatively and various reconstruction options are available in patients with preserved function.

Key Words: Ureteral atresia, proximal atresia, duplex kidney, hydroureteronephrosis.

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Introduction

Ureteral atresia is a rare congenital anomaly, predominantly affecting the distal ureter and resulting in renal dysplasia [1]. Herein, we present a case of proximal ureteral atresia in the lower moiety of a duplex kidney with renal function preserved on that moiety. To the best of our knowledge this is the first reported case of proximal ureteral atresia in a duplicated kidney.

Case report

An eight year old boy presented with complaints of intermittent episodes of left flank pain for three months. It was associated with low grade fever and non-bilious vomiting. Clinical examination and blood investigations were normal.

The abdominal and pelvic ultrasound showed left hydroureteronephrosis with the proximal ureter dilated up to the level of iliac vessels (Fig. 1).

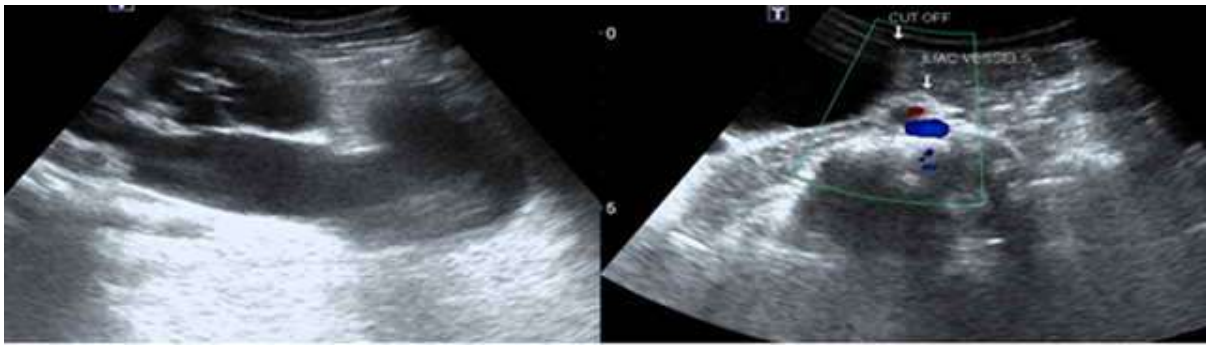

Fig. 1. The left hydronephrosis with proximal ureter dilated upto the level of iliac vessels.

Voiding cystourethrogram showed no reflux and diuretic renal scintigraphy showed a hydronephrotic the left kidney with preserved differential function of 45 % and partially obstructed drainage (Fig. 2). Child was taken up for surgery with a suspicion of left mid ureteric stricture.

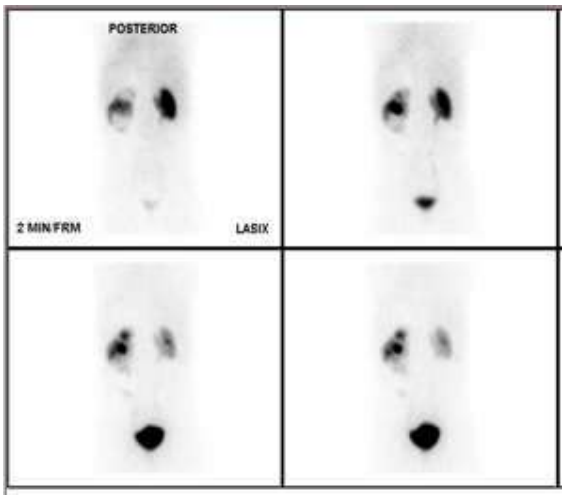

Fig. 2. Diuretic renal scintigraphy showing the left kidney with good function and partially obstructed drainage.

Cystoscopy revealed both ureteric orifices to be of normal shape and position. The left ureter was cannulated with 3Fr ureteric catheter and a retrograde pyelogram (RGP) was done. RGP showed an incomplete duplication of the left ureter with contrast reaching the upper moiety pelvis while the lower ureter was blind ending. At surgery, the lower moiety renal pelvis was grossly dilated with ureteric atresia and a gap of at least 3 cm between the two blind ends. (Fig. 3). The upper moiety pelvis and ureter was normal. A lower moiety ureteropelvic anastomosis was done. The post-operative period was uneventful and renal cortical scan performed after six weeks showed normal function of left kidney with 49 % differential function (Fig. 4). Child is doing well on follow up.

Discussion

Ureteral bud arises from the mesonephric duct at 28 days of gestation. Cephalad growth and contact of the ureteral bud with the metanephric mesoderm induces the development of the entire renal collecting system. Caudal growth into the urogenital sinus marks its connection with the future bladder. Ureteric atresia is a rare malformation occurring either due to failure of ureteric recanalization as a result of relative ischemia, or failure of resorption of Chwalla's membrane (a two layered membrane transiently separating the ureteric bud from the bladder and which gets resorbed later during development) [2,3]. Although atresia occur anywhere along the course of ureter, distal atresia's are more common. Proximal ureteral atresia is extremely rare with only two cases reported in literature till date [1,4].

Ureteral atresia is usually associated with a dysplastic kidney. Dysplasia is attributed to

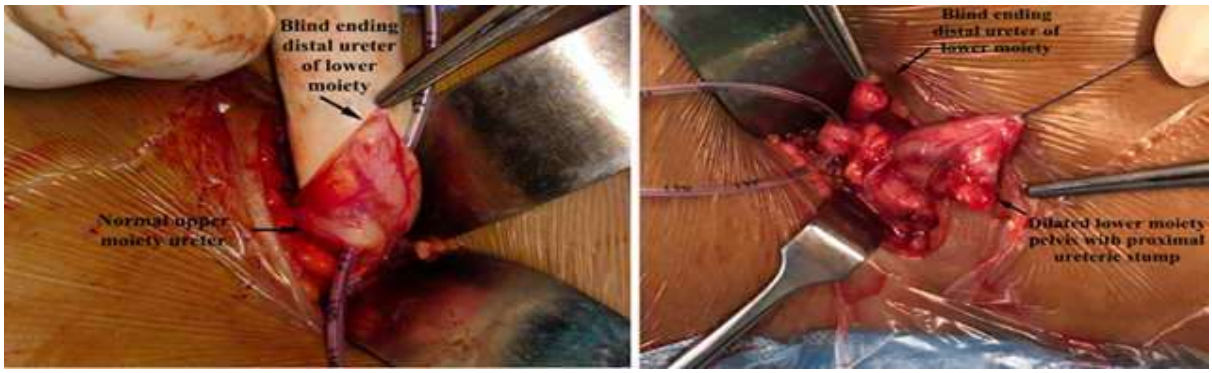

Fig. 3. Intra-operative images showing dilated lower moiety renal pelvis with proximal ureteric stump and distal blind ending ureter.

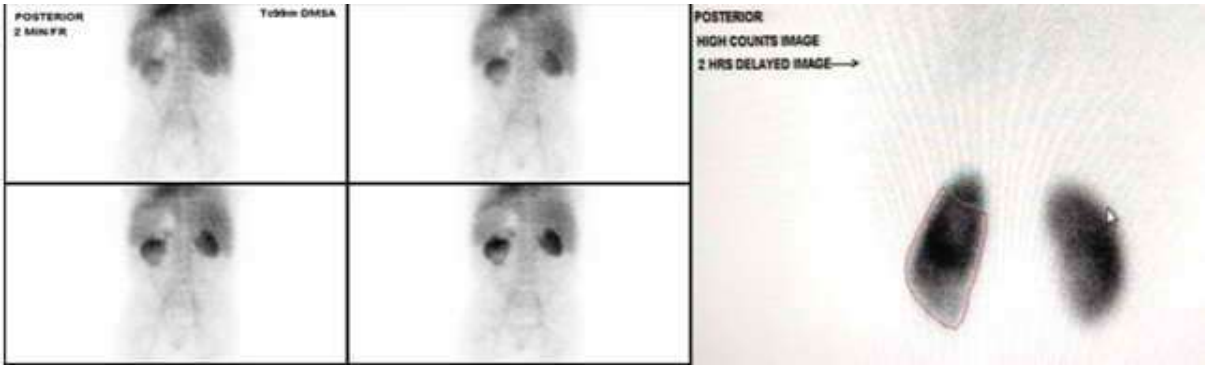

Fig. 4. Postoperative DMSA scan showing the left kidney with differential function of 49 %.

both abnormal development of the ureteric bud and lack of drainage from the evolving kidney. However there have been at least 7 instances where renal function was seen preserved in cases of ureteral atresia. Macpherson et al. [4], Zundel et al. [5] and Hedden et al. [6] have reported cases where children with ureteral atresia presented with urinoma at birth and early surgical intervention helped in successful preservation of function. It was postulated in these cases that decompression secondary to rupture helped the growing kidney escape dysplasia.

Bagnara et al. [3] and Ashimine et al. [7] reported two cases of antenatally detected hydronephrosis which was diagnosed as ureteral atresia after birth. Reconstruction procedures were done and good renal function was noted postoperatively. Wu Shuiquing et al. [8] reported an older child with pelviureteric junction obstruction and distal ureteral atresia. Pyeloplasty and Boari flap reconstruction was done and renal function was found to be preserved [8]. How renal parenchymal function was preserved in these three cases still remains an enigma.

The oldest patient with preserved renal function was a ten year old boy reported by Morozumi et al [9]. This child had crossed fused ectopia and it was postulated that urinary leakage across the crossed moiety might have helped preserve function [9]. A similar mechanism with urine leaking across the duplex moiety could help explain the preserved renal function in our patient.

The common clinical presentations include antenatally detected hydronephrosis, palpable mass and pain. Preoperative diagnosis is

difficult and atresias are usually recognized intra-operatively. The gold standard investigation for diagnosing ureteral atresia is an antegrade or retrograde pyelogram [1]. Dysplastic kidneys usually involute and in asymptomatic incidentally detected patients, no further treatment is necessary. In symptomatic patients with non-functioning kidneys, nephroureterectomy should be done. When function is preserved, pyeloplasty, uretero-ureterostomy or ureteric reimplantation are the surgical options that can be considered [3].

Conclusion

Ureteral atresia is a rare anomaly with proximal atresia being extremely rare. Location, length of atretic segment and functional status of the kidney determines the best surgical option. Outcome depends on the degree of renal dysplasia and timely intervention can help preserve any remaining renal function.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: All photos were taken with parental consent.

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