IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 24, Issue 3 Ser. 2 (March. 2025), PP 45-47
www.iosrjournals.org

A Rare Pelvic Cyst Associated With Posterior Urethral Valves: Long Term Follow Up In A Neonate.

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Abstract:
This is a case report of a fifteen-year-old boy on long term follow-up for treatment of posterior urethral valves with left renal dysgenesis treated in neonatal period. He presented to our outpatient department with an abnormal pelvic cyst shown on routine ultrasound of kidneys and bladder. In view of the rarity, he underwent detailed evaluation with micturating cystourethrogram, magnetic resonance urography, Diethylenetriamine pentaacetate (DTPA) scan, cystoscopy and retrograde pyelogram.
The investigations led to the diagnosis of Zinner syndrome (a triad of seminal vesicle cyst, obstruction of ejaculatory ducts and ipsilateral renal agenesis/ dysgenesis). The association of posterior urethral valves with Zinner syndrome is rarely reported. We report the first case of Zinner syndrome with posterior urethral valves in a child from India.

Date of Submission: 27-02-2025 Date of Acceptance: 07-03-2025

I. Introduction:

PUV (Posterior urethral valve) is the most common cause of bladder outlet obstruction in males in paediatric and neonatal age group. After the initial neonatal management of incision of PUV with or without diversion, the next few months are left for bladder function to improve with supportive care. These patients need long-term follow-up even after puberty and adulthood(1). We report an association with PUV which has been rarely reported, diagnosed in on long term follow up.

II. Case report:

The child initially presented at three months of life with complaints of passing urine in a thin stream with straining. His ultrasound (USG KUB) showed right hydronephrosis and left absent kidney. Micturating cystourethrogram (MCU) was suggestive of PUV (Fig: 1 A). Dimercaptosuccinic acid (DMSA) scan showed a right enlarged adequately functioning kidney with no scars and a very small, practically non visualised left kidney with differential functions of 95% and 5% respectively. Cystoscopy confirmed PUV with trabeculated bladder and incision of valves was done. He underwent repeat MCU at 6 months of life which showed residual valves and incision was done. Following this, child remained symptom free and was on regular follow up.

DMSA scan at one year of life showed a single enlarged adequately functioning right kidney with no scars and left kidney was not visualised. Follow up MCU at four years showed no valves, bladder was smooth walled with no evidence reflux (Fig 1 B). He was on follow up till eight years of life and then lost to follow up.

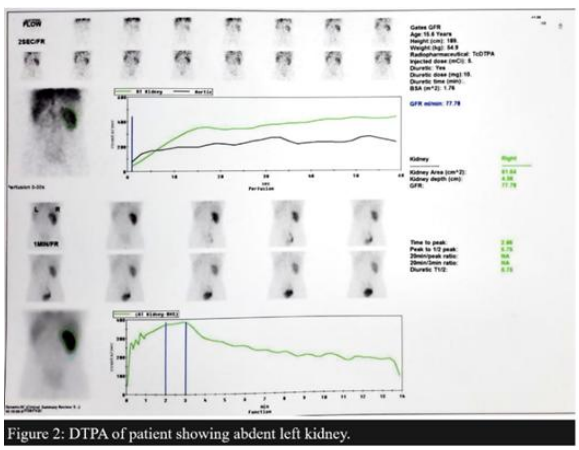

Figure 2: DTPA of patient showing abdent left kidney.

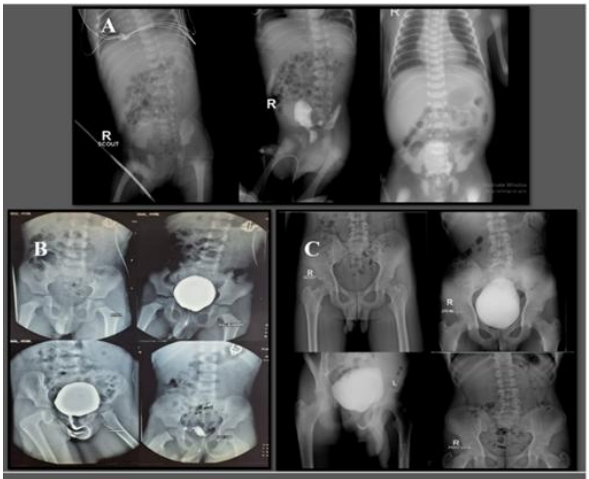

Fig1: MCU done at neonatal period (A), 4 years (B) and at 15 years (C) respectively.

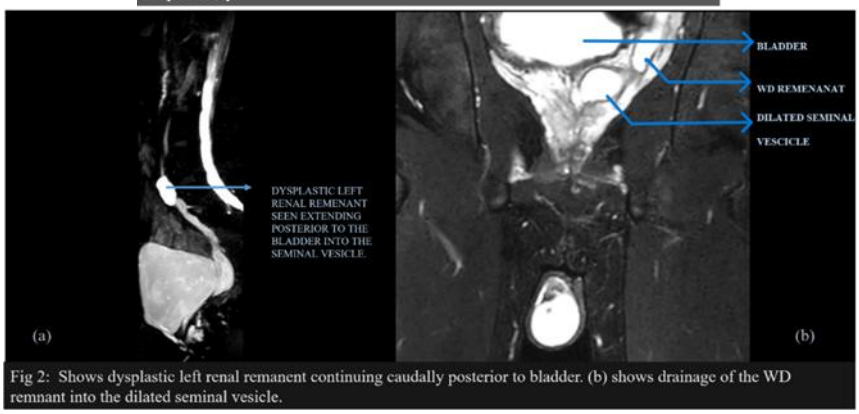

Fig 2: Shows dysplastic left renal remanent continuing caudally posterior to bladder. (b) shows drainage of the WD remnant into the dilated seminal vesicle.

Presently, at fifteen years, he revisited us as he was incidentally diagnosed with a pelvic cyst elsewhere on routine USG. We evaluated him further. His with urine routine was unremarkable; Sr.creatinine was normal (1 mg/dl). USG KUB showed right mild hydroureteronephrosis with few non obstructive calculi. Left kidney was not visualised. There was bulky left seminal vesical with a dilated tubular structure cranial to it (? left ejaculatory duct obstruction) with a cystic lesion in pelvis along the left iliac vessels measuring 2.5x 1.5 cms. MCU showed no evidence of reflux, the bladder was smooth walled with adequate capacity (Fig 1 C). Voiding cystometry showed normal pressure bladder. Right retrograde pyelogram (RGP) showed the ureter was mildly dilated and contrast was freely entering the pelvicalyceal system which cleared in 3 hours delayed imaging. The left ureteric orifice was not identified. His DTPA and DMSA showed a right normal functioning kidney with unobstructed drainage and absent left kidney (Fig 2) . Magnetic resonant (MR) urography showed right normal kidney. Prostrate and right seminal vesicle were normal. Left seminal vesicle was dilated. A dilated tubular structure was seen extending from the left seminal vesicle cranially along the iliac vessels upto the level of pelvic inlet ? dilated mesonephric remnant which could represent an ectopically inserted left ureter. A loculated fluid intensity lesion measuring 2.5 cm was seen in the left common iliac region, likely representing the dysplastic renal remanent (Fig 3). Child was diagnosed with Zinner's syndrome with ectopic insertion of the ureter. As he remains asymptomatic, he is advised to be on follow up.

III. Discussion:

ZS is rare condition with approximately sixty cases reported in literature till date in paediatric age group. It is usually incidentally discovered between the second and fourth decades of life. However, in this era of advancement in diagnostic technology, it is being increasingly detected at younger ages (2)(3).

The Wolfian duct (WD) develops into ductus deferens, seminal vesicle, and epididymis owing to testosterone and anti-mullerian hormone. An abnormality of development of the distal part of the WD can cause obstruction of the ejaculatory duct, leading to dilatation and cysts of the ipsilateral seminal vesicle.(4) The common embryological origin of the WD and the ureteric bud can be the cause of combined alteration in the development of both systems.(5) However, the development of bladder is endodermal in origin. The urogenital septum divides the cloaca into rectum posteriorly and urogenital sinus anteriorly. The urogenital sinus continues

to form the bladder, with the inferior end forming the urethra. As the WD fuses with the cloaca, part of the duct gets incorporated into the posterior wall of the bladder.

Children with ZS are mostly asymptomatic. In adults it causes lower urinary tract symptoms, pelvic pain, and infertility (6). The diagnosis in paediatric age is mainly beneficial in adult age, during the period of the sexual activity, if patient presents with infertility caused by low ejaculatory volume or a possible anti-sperm antibodies production due to unilateral testicular obstruction.(3)(7)

The first-line investigation is ultrasound. The seminal cysts are seen as a cystic pelvic mass that can demonstrate thick or irregular wall and calcifications with internal echoes depending on haemorrhage or infection. Scintigraphy identifies ipsilateral kidney agenesis/ dysgenesis. Pelvic MRI is the preferred modality for diagnosis. Diagnostic cystoscopy can be done to see for hemitrigone, ectopic ureteric orifice, the anatomy of the urethra and to analyse the connection of the cysts with the urethra (2)(8). In this child, the renal dysplasia was initially thought to be secondary to the 'pop off' mechanism in view of the PUV (9). The serial ultrasounds failed to detect the seminal vesicle cyst till he was 15 years.

Asymptomatic patients are managed conservatively. Surgical management is for symptomatic patients with persisting pelvic pain during ejaculation, recurrent orchiepidydimitis, bladder outlet obstruction and lower urinary tract symptoms due to enlarged cysts causing irritation at the bladder outlet. Surgeries proposed are unroofing of the cysts, vesiculectomy with the resection of renal remnants, with or without vasoligation. Minimal access surgeries allow more precise dissection in the depth of the pelvis, with minimal damages to neighbouring structures.(3)

IV. Conclusion:

The association of PUV with ZS is very rare. While a child with unilateral renal agenesis associated with PUV presents with a pelvic cyst, ZS can be considered along with other differential diagnosis. Pelvic MRI helps in establishing the diagnosis. The management options will depend on the symptomatology of the patient. This case is discussed as it highlights this rare association and need for life long follow up of patients treated for posterior urethral valves.

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