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CONGENITAL ANTERIOR URETHRAL VALVE : A RARE CASE REPORT

Paediatric Surgery

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KEYWORDS

INTRODUCTION:

Posterior urethral valves are one of the most common causes of urinary tract obstruction in the pediatric population. They are obstructing membranous folds in the lumen of the posterior part of the urethra and are mostly exclusive to male patients. Posterior urethral valves were first described by Morgagni in 1717 and then by Langenbeck in 1802, who reported valve-like folds in dissected cadavers.

Posterior urethral valves are classically split into three subtypes by Young's criteria according to the orientation of the valves within the urethra.[1]

Type I: 95% - Posterior urethral folds (Plicae colliculi), which arise from the caudal verumontanum along the lateral margins of the urethra fuse anteriorly causing an obstruction.

Type II: Membranes cranially attached to the bladder neck originating from the verumontanum. These are now thought to be due to hypertrophy of the plicae colliculi and not obstructive valves.

Type III: 5% - Round membrane at the caudal verumontanum with a hole in the middle that is either above (type IIIa) the verumontanum or below it (type IIIb). Neither subtype's hole communicates directly with the verumontanum.

The anterior urethral valve is a rare congenital anomaly that causes lower urinary tract obstruction in children. It can occur as an isolated entity or in association with a proximal diverticulum; probably representing a spectrum of the disease [2].

Various proposed hypotheses include an abortive attempt at urethral duplication, failure of alignment between the proximal and distal urethra, imbalanced tissue growth in the developing urethra leading to a remnant of excess tissue acting as a valve, and congenital cystic dilatation of periurethral glands leading to a flap-like valve [3]. Forty percent of the AUVs are located in the bulbar urethra, 30% at the penoscrotal junction, and 30% in the pendulous urethra [4]. The clinical manifestation is highly variable and depends on age of the patient and degree of obstruction. It may present with severe obstruction and bilateral severe hydrouretero-nephrosis, end-stage renal disease, and even bladder rupture [5]. It is a posteriorly directed semilunar fold and arises from the anterior urethra. It can mimic an anterior urethral diverticulum, but the posterior lip is absent in the valve [6]. It can very rarely be associated with posterior urethral valve [7].

VCUG is the diagnostic modality of choice in the diagnosis of anterior urethral valve. It can reveal a dilated or elongated posterior urethra, a dilatation of the anterior urethra, a thickened trabeculated bladder, a hypertrophied bladder neck, VUR, and urethral diverticula. The urethra appears dilated proximal to the valve and narrowed distal to it on VCUG. A valve may be demonstrated on RGU as a linear filling defect along the ventral wall, or it may show a dilated urethra ending in a smooth bulge or an abrupt change in the caliber of the dilated urethra on VCUG [8]. VCUG may also reveal an associated anomaly in addition to demonstrating the valve. One-third of cases have shown VUR and upper tract deterioration was present in one-half of cases [9]. Urethroscopy usually helps in confirming the diagnosis. The treatment includes the destruction of the valve by electrocautery or by a resecting hook [9].

Case Summary -

A 5-year-old boy presented with chief complaints of dribbling of urine, straining and narrow passage of stream since birth.

The child does not have any history of fever, trauma or any mass in the abdomen.

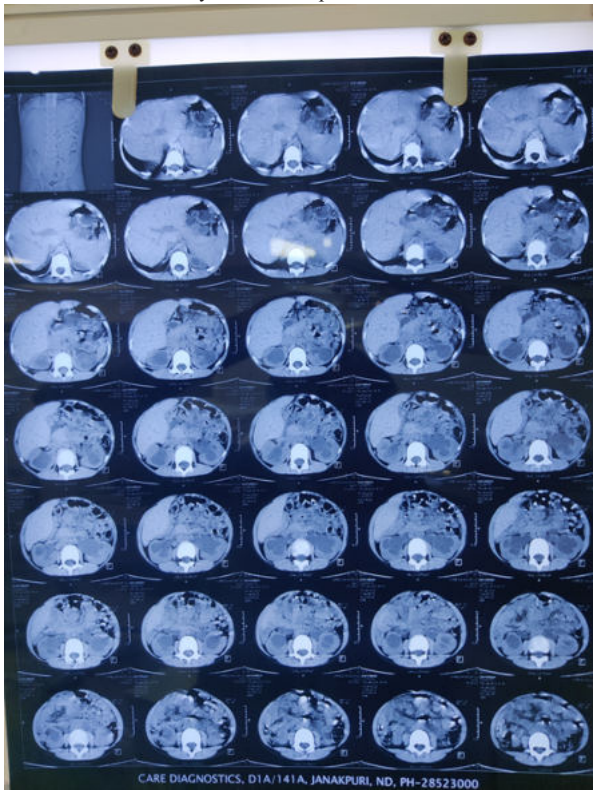
INVESTIGATIONS-

HAEMATOLOGICAL INVESTIGATIONS WERE

Hb-7.5 g/dl
Tlc -16500 cells/liter
Platelets -3.6lakh/ml

RADIOLOGICAL INVESTIGATION

SONOGRAPHY AND CT KUB revealed diffusely thickened bladder with Bilateral hydroureteronephrosis.



MCU -Micturating cysto-urethrogram showed an irregularly shaped bladder with diverticulum. The urethra was unremarkable. There was no reflux seen on MCU. The boy was planned for cystoscopy after ensuring that he did not have any urinary infection. Cysto Scopy revealed an anterior urethral web with no posterior urethral valves. The

bladder had trabeculations with bilaterally dilated ureteral orifices.

PER OP FINDINGS

PIC 1 - BLADDER STATUS

PIC 2 ANTERIOR URETHRAL WEB CAUSING OBSTRUCTION

PIC 2-POST FULGURATION STATUS

Obstruction is at the level of bulbar urethra with unique presentation. no puv were found and there was no significant bladder neck hypertrophy.

Post operatively child passed urine in good stream after 5 days removal of foleys catheter and is kept on follow up .

CONCLUSION-

There is lacunae in literature in cases of anterior urethral web, hence this case has been reported. The differential diagnosis could be congenital urethral sticture,cobbs collar and anterior urethral valves or traumatic strictures but they have been reported in numbers in literature.

Although most urethral strictures in males are post-traumatic, there are rare reports of congenital urethral strictures of the bulbous urethra in neonates and older children [10]. These might be secondary to a failure of canalization of the cloacal membrane during fetal development [11, 12] and have also been referred to as Cobb's collar, Moormann's ring and Young's type III valve [13, 14].

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