ORIGINAL RESEARCH PAPER

Paediatrics

KEY WORDS:

CONGENITAL ISOLATED PRIMARY OBSTRUCTIVE MEGALOURETHRA, EMBRYOLOGY AND MANAGEMENT - RARE CASE REPORT

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ABSTRACT

Male urethra may presents with mesenchymal anomalies in form of dilated anterior urethra or deficient erectile tissue of the penis. It may present with deformity of the penis (Scaphoid megalourthra) or Impotence (Fusiform megalourthra). Megalourethra is a non-obstructive dilatation of the penile urethra .Herewith reporting a case of 2 yrs old child presented with obsrtuctive megalourethra immediately after birth with signs of lower urinary tract obstruction and severe uro-sepsis. The diagnosis of megalourethra was established on the basis of clinical and radiological findings. Patient was successfully managed by reduction urethroplasty with staged procedures.

INTRODUCTION :-

Megalourethra is rare anomaly presenting with non-obstructive dilatation of penile urethra (1,2) It is a diffuse dilatation of anterior urethra due to absence of development of erectile tissues of the penis which affects anterior part of urethra and usually cause abnormal shape of penile shaft while voiding. The first case of congenital megalourthra was reported by Obrinsky who also mentioned about association with prune belly syndrome. (3,4)

In 1955 it was defined as " a congenital dilatation of penile urethra without distal obstruction " by Nesbitt (5). Uptill now less than 100 cases have been reported in literature but obstructive megalourethra is not been reported yet (6). Classification of congenital megalourthra based on findings on urethrography as more common schaphoid type and fusiform type was done by Dorairajan. Schaphoid type presents as bulging of ventral urethra and fusiform type presents as a circumferential dilatation of urethra (7).

Herewith presenting with a case of congenital isolated obstructive megalourethra managed with staged approach. The aim is to provide a clear picture about embryology, manifestations and management which are of paramount importance in treatment of this often misdiagnosed rare anomaly.

CASE REPORT

A newborn baby with full term normal delivery referred with complaints of not passed urine for 72 hours post delivery and deformed penis (Fig.1). On per abdomen examination urinary bladder was distended with gross distension of abdomen with signs of uro-sepsis on blood profile. Ultra-sonography showed over-distended urinary bladder with bilateral gross hydro-ureter and hydro-nephrosis, grossly dilated anterior urethra. Blood investigations showed signs of urosepsis with serum creatinine upto 4 . It was very difficult catheterisation with smaller caliber of infant feeding tube also. On retrograde urethrography there was evidence of grossly dilated anterior urethra with a tortuous fold seen which was causing obstruction to outflow due to a kink (Fig.2). Small urinary bladder with grade 2 reflux noted on left side.

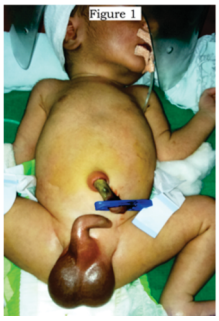


Figure.1

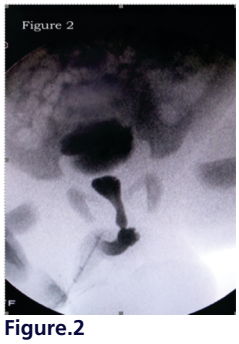


Figure.2

Accepting high risk factors supra-pubic vesicostomy was perfomed. Baby got recovered from uro-sepsis and discharged. On follow up serial sonography studies showed gradual decrease in hydroureter and hydronephrosis with normal serum creatinine values upto 0.8. At 2 years of age , As a preoperative work up along with all routine blood investigations, again retrograde urethrograpy was done presenting with similar findings as in newborn period. On Diagnostic Cystoscposy , scope could not be negotiated beyond urethral kink. Reduction Urethroplasty was planned.

Patient was given regional anaesthesia in the form of caudal block along with sedation. After adequate preparation, glans stitch was taken and penis degloved by circumferential incision. After complete degloving of the penis the urethra was opened longitudinally over the swelling taking care not to open urethra beneath. The excess redundant urethral tissue was excised and urethroplasty performed using vicryl 6-0 continuous sutures over infant feeding tube no 7. Urethroplasty was performed in such way that surture line will come in spiral fashion and not exactly in midline. The urethroplasty was strengthened by a second layer using adjacent spongiosum tissue (Fig 3 to Fig 7). Subsequently skin flaps were sutured using vicryl 5-0 sutures. Adequate haemostasis was achieved. Light dressing was given so as to avoid any excessive pressure over the urethroplasty and skin flaps and at the same time avoid any hematoma formation. The catheter was removed on the 14th POD. Supra pubic foley's catheter inserted. On follow up of 1 month baby started passing urine per urethra with good stream and no dilatation of penis, no fistula. Suprapubic Foleys catheter was removed at 3 months with gradual trials given by blocking the suprapubic catheter intermittently

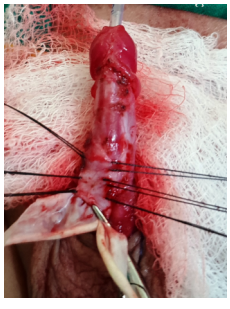


Figure.3

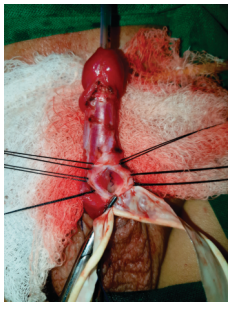


Figure.4

Figure.5

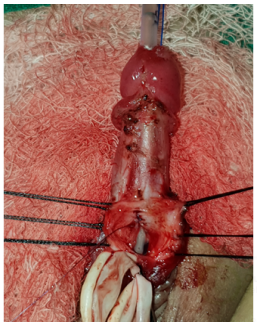
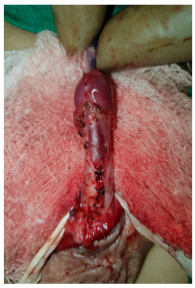


Figure.6



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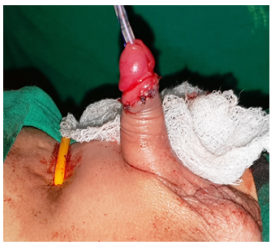

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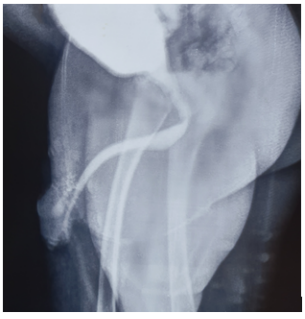

Figure.8

Regular follow-up up to 1 year showed no recurrence of penile swelling with normal voiding pattern confirmed with micturating cysto-urethrogram (Fig.8). There were no fistulas, infections or any other complications. Baby is under observation on follow up for urinary bladder dynamics, left sided persistent reflux and vesicostomy closure.

DISCUSSION

Functional lower urinary tract obstruction due to congenital megalouretra is very rare form which may caused by primary or secondary agenesis / hypoplasia of penile corporal tissues. The exact embryological cause of congenital megalourthra is not clearly understood. The most widely accepted theories propose a defect in migration or development of mesenchymal tissues of the phallus (2). Mild delays with earlier and more complete canalization may be associated with scaphoid while longer delays with later and less complete canalization and fusiform megalourethra (8). Owing to the poor development of supporting erectile tissue there is stasis of urine causing functional obstruction (9).

Congenital megalourethra has been classified into two types:
(a) Scaphoid type

A milder form that involves the urethra and corpus spongiosum alone where during micturition the urethra dilates in a scaphoid fashion as the dorsal aspect is well supported by the spongiosum.
(b) fusiform type

More severe form as corpus spongiosum and corpora cavernosa both are involved resulting in fusiform dilatation of the phallus during voiding. However, this classification is neither pathological nor etiological and does not help in prognosis.

Recently, Amsalem et al classified the condition into (a) primary (or ex-vacuo), caused by absence or hypoplasia of the corpora spongiosa and cavernosa, associated with normal amniotic fluid volume, usually preserved renal function and better outcome, and (b) obstructive (secondary), which results in oligohydramnios with a higher incidence of renal failure, pulmonary hypoplasia and thus mortality (10). In either types the corpora are hypoplastic might be due to result of an initial absence of the corporal tissue in the primary type or secondary to pressure in the obstructive type. The "primary" type can become obstructive if blocked by debris and can result in oligohydramnios later in gestation. A mortality rate of 13% in the "primary/scaphoid" type and 66% in the "secondary/fusiform" type has been reported in the literature (11).

Various congenital anomalies are associated with megalourethra mostly related to urogenital system and sometimes with other organ systems as well. Jones et al in a review have reported, associated congenital anomalies are seen in 80% of scaphoid type and 100% of fusiform type (2). The commonly associated anomalies along with congenital megalourethra include renal dysplasia-hypoplasia, hydronephrosis, hydroureter, vesicoureteric reflux, prune-belly syndrome, urethral duplication, megacystis, hypospadias, posterior urethral valves, and undescended testes. Other system anomalies including VATER (vertebral, anal atresia, trachea-oesophageal fistula, and renal anomalies) and VACTERL (vertebral, anal atresia, cardiac, tracheaesophageal fistula, renal, and limb deformities) are described (6). The treatment of megalourethra may be one stage or two stage urethroplasty depending on the age of presentation and general condition of the patient. For scaphoid type, Nesbitt first described a longitudinal reduction urethroplasty as was performed in these cases (5). Heaton and colleagues described a technique of urethral plication for some cases of scaphoid megalourethra (12).

The management of fusiform type is complicated ranging from sex reassignment to major phallic reconstruction (2).

CONCLUSION

Congenital megalourethra is a rare and surgically correctible malformation of the anterior urethra. Antenatally it can be picked up by ultrasound. The condition should be specifically looked for in all fetuses with suspected lower obstructive uropathy and oligohydramnios. Detailed examination of the fetal perineum and genitalia will provide clues to the correct diagnosis. . It is essential to consider megalourethra in the differential diagnosis of all children with penile or penoscrotal swelling . Management might be single staged or multi-staged has to be individualized depending on type and the presence of associated congenital anomalies. In isolated scaphoid type of megalourethra reduction urethroplasty gives excellent result. As very few cases has been reported yet , many cases are misdiagnosed or incorrectly treated.

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