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Case report

MEGALOURETHRA: A CASE REPORT

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ABSTRACT

Congenital megalourethra is a rare mesenchymal anamoly of the male urethra. It is defined as dilation of the anterior urethra due to absence of development or deficiency of erectile tissue of the penis. It leads to deformity of the penis (scaphoid megalourethra) or impotence (fusiform megalourethra) and also renal insufficiency and pulmonary hypoplasia. The associated anomalies are often life threatening and influence the management and prognosis. We present here a report of a case of megalourethra.

Keywords: Congenital, Fusiform, Megalourethra, Scaphoid.

INTRODUCTION

Megalourethra is a congenital disorder, characterized by dilatation as a whole or a part of urethra with a specific penile ventral deformity known as the pelican bag. It is a rare form of functional lower urinary tract obstruction.[1] Maternal diabetes and associated VACTERAL syndrome are implicated.[2,3] It is of two type scaphoid and fusiform. A meticulous surgery is needed to restore the original shape and function.

CASE REPORT

An eight month male child parent complaining of penile deformity without derangement of stream admitted in our institute. On local examination the ventral penile surface was markedly dilated and redundant. Penoscrotal angle was reduced. Urethral meatus was normal. It was fusiform megalourethra. (fig 1) Examination revealed normal genitalia with normally descended testis and scrotum. The urinary stream was normal except for a terminal dribbling.

There was no relevant family history or antenatal history. Patient was screened for other congenital anomalies and proved devoid of any association and syndrome. Routine investigation was done, those were in normal level except renal function test which were deranged. Blood urea 87mg/dl and serum creatinine 1.67mg/dl and potassium 5.7 mEq/lit.

USG abdomen suggestive of bilateral hydroureteronephrosis, dilated PCS, cystitis with megalourethra. (fig 2).

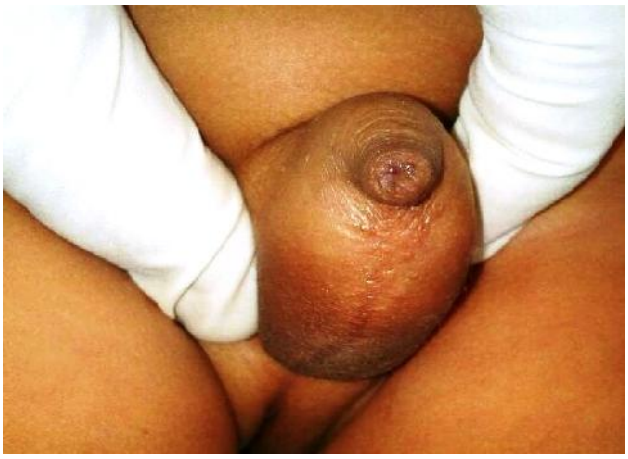

Fig 1: Pre Operative megalourethra photo

MCU revealed urethra dilated with no reflux seen. (fig 3) Because of deranged renal function so planned to open the urethra. (fig 4) A urethral fistula that resulted was later planned for repair.(fig 5)

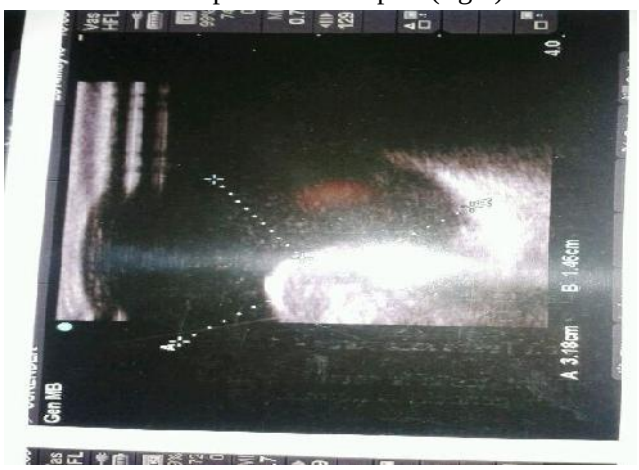

Fig 2: USG shows megalourethra

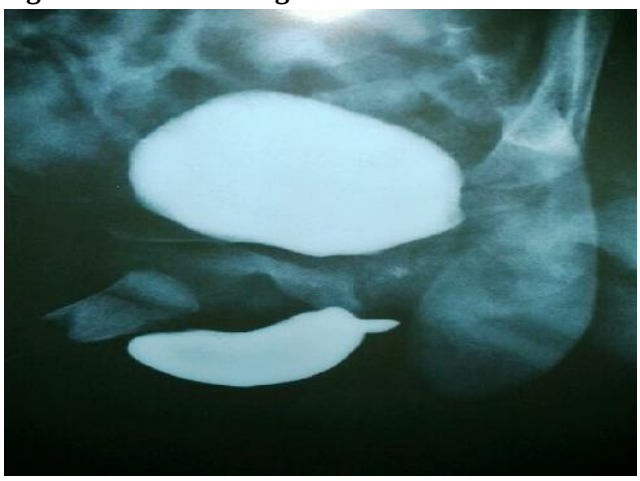

Fig 3: Micturiting cystourethrography showing dilated urethra

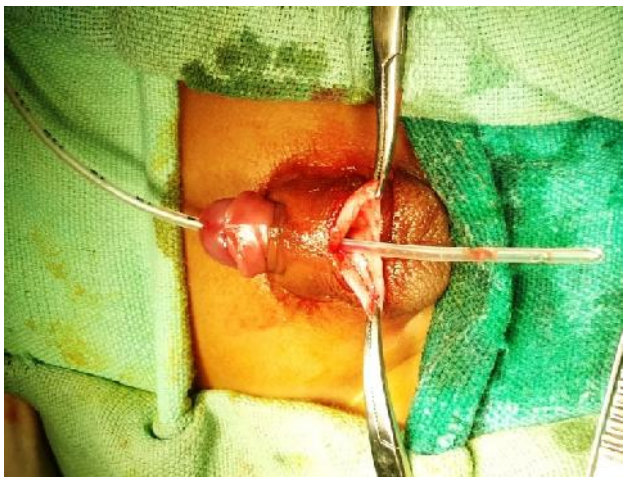

Fig 4: Intraoperative photo showing ventral created fistula and anterior opening of urethra

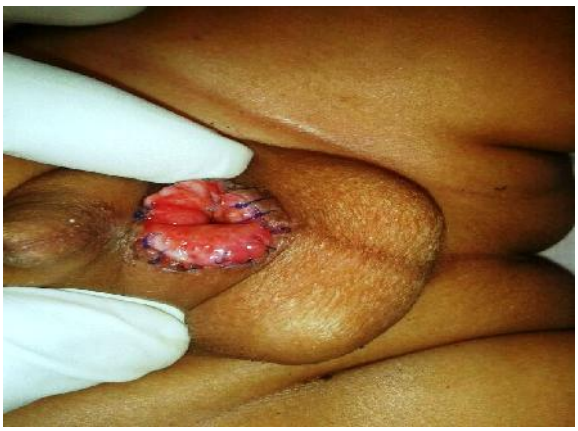

Fig 5: Photo shows ventral urethral fistula

DISCUSSION

Megalourethra results from deficiency of the mesodermal tissues of the phallus due to defective migration, differentiation or development of the erectile tissue.[3]

There are two varieties of congenital megalourethra-scaphoid and fusiform. In the fusiform the corpus spongiosum is maldeveloped in the anterior urethra leads bulging of ventral urethra where as in scaphoid both spongiosum and cavernosum maldeveloped or deficient causing circumferential expansion of urethra. Intermediate form has been reported.[4] Fusiform type is more common. The fusiform variety is more serious than scaffold type and is associated with more serious anomalies and carries a poor prognosis. [5]

This anomaly can be associated with other urinary tract anomalies including dysplastic kidneys, vesicoureteric reflux, undescended testis, Prune belly syndrome, megaureter, hypospadias, posterior urethral valve and other system abnormalities[5] including Vacteral.

The fusiform variety has a higher incidence of oligohydromnios, pulmonary hypoplasia. As a result of oligohydromnios which can result in neonatal death and chronic progressive renal failure that can result in end stage renal disease.[2]

The management is classically the Nesbit repair. The timing and method of reconstruction in patients with congenital megalourethra are dictated by the anatomical construction of the phallus and the health of the child. In those patients who survive the immediate neonatal period, reconstruction of the phallus can be undertaken. Nesbit described after a

circumcising incision and degloving of the penis. He performed a longitudinal reduction urethroplasty on a catheter. Some have advocated urethral placation, However the rarity of the defect precludes any generalization with regards to surgical management. Each case must be considered individually.[4]

It has been suggested that sac like dilatation of distal urethra cause proximal obstruction with resultant dilatation of upper urinary tract. The urinary tract return to normal after correction of megalourthra. Detailed investigation of the upper and lower urinary tract should be carried out at the earliest to detect anomalies and to treat them early to reduce morbidity and mortality.

CONCLUSION

The prognosis of isolated scaphoid megalourethra is very good. The operation is simple and successful. On the other hand, the fusiform type is difficult to treat. Early diagnosis and management affords a long lasting cure to the patients.

Conflict of interest: Nil

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