Congenital megalourethra with left ectopic megaureter and bulbar urethral stricture: A rare entity

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ABSTRACT

Congenital megalourethra is a rare disorder of anterior urethra. This disorder is characterized by the congenital absence of corpora spongiosum or cavernosa or both which leads to dilatation of the urethra. We present a case of congenital megalourethra in a 9-month-old child, which was associated with left ectopic megaureter and bulbar urethral stricture. Most of reported cases were treated by Nesbit urethroplasty. This case was initially treated by emergency trocar cystostomy and the left ureteroneocystostomy, and finally, reduction urethroplasty was carried out as a definitive treatment.

Key words: *Congenital, Cystostomy, Megalourethra, Urethroplasty*

Congenital megalourethra is a rare disorder of anterior urethra due to congenital absence of corpora spongiosum and/or cavernosa which leads to extensive dilatation of penile urethra. This condition may be of scaphoid type where dysgenesis of corpora spongiosum occurs or fusiform type where cavernosa and spongiosum, both are absent or ill developed. The first case of megalourethra was reported by Obrinsky; and Nesbit was the first to describe its treatment as urethroplasty [1]. Most of cases are associated with other congenital abnormality. We report a case of congenital megalourethra associated with left ectopic megaureter and bulbar urethral stricture in a 9-month old child. This case was managed initially by trocar cystostomy and ureteroneocystostomy, and then, urethroplasty was done as definitive treatment.

CASE REPORT

A 9-month-old child presented in outpatient department with complaint of voided with poor stream and associated swelling of the penis. The swelling increased during the act of micturition. On examination, a swelling of 3 cm × 3 cm was found on the right ventral surface of the penis which became more prominent like a balloon duringtx micturition. Both testes were palpable in the scrotum (Fig. 1). The weight of child was 6 kg. All laboratory tests including hemogram, renal function test, routine examination of urine, and culture and sensitivity of urine were within normal limit. High-frequency ultrasonography showed the left hydro-nephrotic kidney and the left ectopic megaureter open into prostatic urethra and trabeculated bladder (Fig. 2). Retrograde urethrogram showed the stricture of bulbar urethra and dilatation of distal urethra in this patient (Fig. 3). Micturating cystourethrogram showed normal capacity of bladder with diverticula on left side and without vesicoureteric reflex. Initially, trocar cystostomy and the left ureteroneocystostomy were carried out, and later on, reduction urethroplasty was done. The patient is doing well post operatively and voided a normal stream without any abnormality of the penile shaft.

DISCUSSION

Congenital megalourethra is characterized by elongation and dilation of the penile urethra along with ill developed corpora

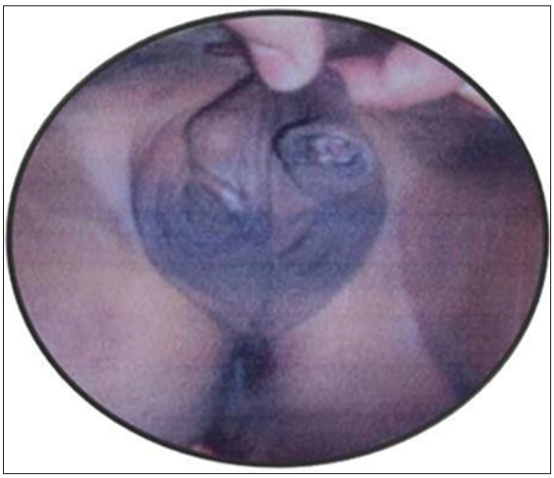

Figure 1: Swelling on ventral surface of penis

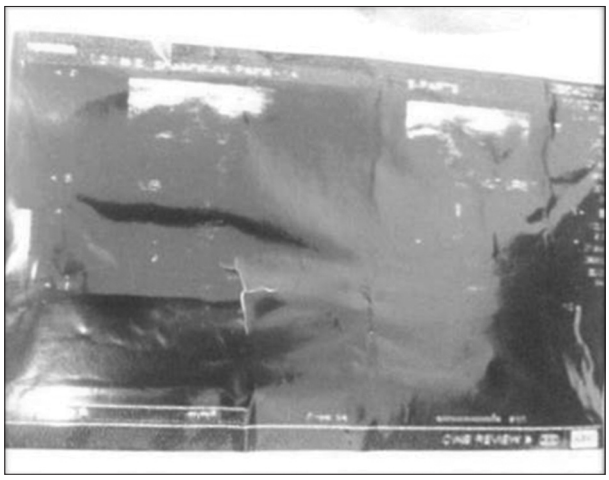

Figure 2: USG: Showing Megalourethra

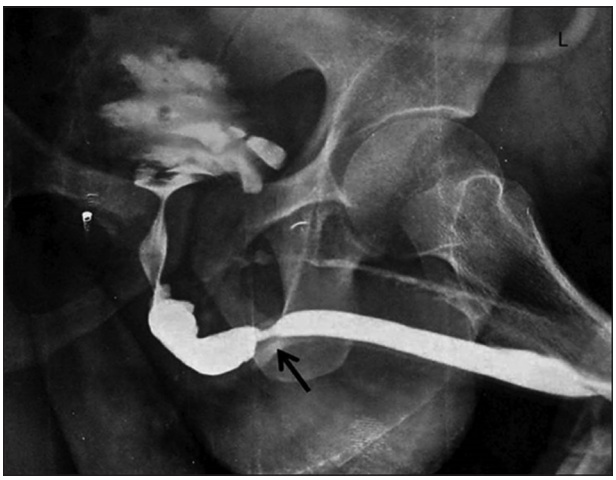

Figure 3: RUG: Showing bulbar uethral stricture

spongiosum and cavernosa. This is a rare abnormality of the penile urethra caused by mesenchymal defect [2]. It usually involves anterior urethra and subdivided into two types: Fusiform and scaphoid [3]. The scaphoid type is more common. If the corpora cavernosa are affected, then fusiform type of megalourethra develops. On the other hand, the scaphoid type is because of ill development of the corpus spongiosum. The fusiform type is more severe than scaphoid type [4]. The fusiform type is always associated with other congenital disorders with poor prognosis as compared to scaphoid type. In one study, associated congenital anomalies are seen in 80% of scaphoid type and 100% of fusiform type [5]. The most common congenital abnormality associated with megalourethra is urogenital abnormality. The congenital abnormality may be imperforated anus, posterior urethral valve, prune-belly syndrome, esophageal atresia, VATER Complex, trisomy 21, and malformation of the cardiovascular and musculoskeletal system [6]. The prognosis is worse in case of these associated malformations. In the present described case, patient has megalourethra with the left ectopic megaureter and bulbar urethral stricture. Considering all these associated congenital abnormalities, Hamzaoui et al. recommended the renal function test and imaging of the upper and lower urinary tract [7]. The chance of survival for all infants with megalourethra is possible if the amniotic fluid is normal, but most of live born infants have voiding, renal, and sexual dysfunction [8]. The prenatal diagnosis should be carried out in view to detect such associated congenital anomaly and take decision about termination of pregnancy. The milder form of megalourethra is spontaneously resolved at 19-34 weeks of gestation [6]. Nesbit described the treatment for megalourethra as longitudinal reduction urethroplasty [1]. The management of fusiform is complex and staged procedure that includes sex reassignment, major penile reconstruction, and the placement of penile prosthesis [5]. Long-term follow-up is required in these patients for the assessment of erectile function and fertility potential [6].

CONCLUSION

Congenital megalourethra is a rare disease. A good outcome can be anticipated if diagnosis is done in prenatal stage. Trocar cystostomy and ureteroneocystostomy are emergency option, but urethroplasty and phallic reconstruction are the definitive treatment in such patient.

REFERENCES

1. Nesbitt TE. Congenital megalourethra. J Urol. 1955;73(5):839-42.
2. Sreetharan V, Sommerlad BC, Kangesu L. A case of congenital megalourethra. J Plast Reconstr Aesthet Surg. 2006;59:550-2.
3. Centinkursun S, Ozturkh H, Sakarya MT, Surer L. Congenital megalourethra. Indian J Pediatr. 1996;63(4):566-8.
4. Berrocal T, Lopez-Pereira P, Arjonilla A, Gutierrez J. Anomalies of the distal ureter, bladder, and urethra in children: Embryologic, radiologic, and pathologic features. Radiographics. 2002;22(5). DOI: 10.1148/radiographics.22.5.q 02se 101139.
5. Jones EA, Freedman AL, Ehrlich RM. Megalourethra and urethral diverticula. Urol Clin North Am. 2002;29:341-8, vi.
6. Wax JR, Pinette MG, Landes A, Cartin A, Blackstone J. Prenatal sonographic diagnosis of congenital megalourethra with in utero spontaneous resolution. J Ultrasound Med. 2009;28:1385-8.
7. Hamzaoui M, Essid A, Laabidi B, Ben Attia M, Houissa T. Neonatal congenital mega urethra. Prog Urol. 2002;12(4):680-3.
8. Amsalem H, Fitzgerald B, Keating S, Ryan G, Keunen J, Salle JL, et al. Congenital megalourethra: Prenatal diagnosis and postnatal autopsy findings in 10 cases. Ultrasound Obstet Gynecol. 2011;37(6):678-83.

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