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Case Report

Bladder exstrophy with inguinal hernia: a case report

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ABSTRACT

Exstrophy of urinary bladder with epispadias involves protrusion of the urinary bladder through a defect in the lower abdominal wall accompanied by separation of pubic symphysis. It is a rare but challenging condition that causes significant physical, functional, social, sexual and psychological problems later in life. Bladder exstrophy commonly involves males and most cases are sporadic. Inguinal hernia is a complication associated with bladder exstrophy and it occurs due to lack of obliquity of the inguinal canal secondary to pubic diastasis. Authors report here, a case of antenatally diagnosed case of classic bladder exstrophy associated with left sided inguinal hernia which was incidentally diagnosed on tenth day of life. Our neonate underwent primary bladder closure with herniotomy. Staged reconstruction of epispadias and bladder neck has been planned at a later date. Recurrence of inguinal hernia after repair is common and bilateral inguinal exploration while performing herniotomy is advised to prevent its recurrence. Prognosis of such cases depends on the degree of continence achieved. With timely reconstructive surgery, continence rates can be as high as 60-70 percent.

Keywords: Exstrophy, Epispadias, Herniotomy, Inguinal hernia

INTRODUCTION

Urinary bladder exstrophy constitutes a rare spectrum of genitourinary malformations in which there is incomplete closure of lower abdominal wall leading to bladder protrusion. The spectrum ranges from bladder exstrophy with epispadias (in boys) where it is termed classic bladder exstrophy to more complex cloacal exstrophy where entire hindgut along with the bladder is exposed. Incidence is 3 to 5 per 100000 births.[1-3]

It occurs as an isolated sporadic defect but association with other anomalies such as omphalocele, anal defect, neural tube defect, cleft lip/palate, preterm birth etc have also been reported.[4,5] Prenatal diagnosis is made by ultrasound examination which shows non-visualisation of bladder filling, diminutive genitalia and a lower abdominal mass.[4] Fetal MRI can provide more detailed anatomy of the defect with accurate sexual differentiation.[6,7] Inguinal hernia is a common association with this entity which can present anytime between birth and adulthood.[8] Hernia is more often bilateral, more common in male and has a tendency to recur even after surgical correction. Modern staged reconstruction is the surgery of choice for bladder exstrophy.[9,10] However, in selected cases, cystoscopic injections of dextranomer (DX) or polydimethylsiloxane (PDMS) microspheres provide sufficient bladder neck control to achieve continence.[11]

CASE REPORT

A 24 years old primigravida was registered to our centre with her antenatal scan (3rd trimester) suggestive of non-

visualisation of urine filled urinary bladder of the fetus with apparently normal upper urinary tract. Antenatal history was uneventful. Mother was immunised and had adequate weight gain during pregnancy. A 3 kg full-term male child was delivered vaginally. On examination,the baby was hemodynamically stable, however per-abdomen examination revealed a mass protruding from the lower abdomen along with epispadias and a low lying umbilicus (Figure 1). Rest of the systems were normal. A clean wrap was put over the mass to reduce the fluid loss. Abdominal ultrasound revealed bilateral undescended testis with normal kidneys. On the 10th day of life, neonate was noticed to have left sided inguinal hernia. Primary closure of bladder mucosa along with herniotomy was done and child was discharged. Epispadias and bladder neck reconstruction is planned for the baby after attaining adequate weight gain. On follow-up the child has been thriving well and has gained adequate weight.

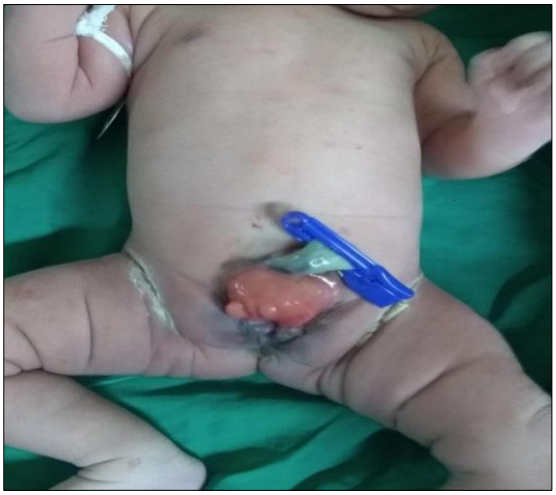

Figure 1: The mass protruding from the lower abdomen (bladder exstrophy), a low-lying umbilicus and epispadias.

DISCUSSION

Bladder exstrophy represents a spectrum of disorders where some part or all of the lower urinary tract fails to close and gets exposed through the lower abdominal wall. Spectrum varies from simple epispadias to more severe cloacal exstrophy where hindgut along with the bladder mucosa is exposed through a defect in the lower abdominal wall. Most cases are sporadic with male preponderance, male to female ratio is 2:1 in classic exstrophy, however it is observed to be 1:1 in cloacal exstrophy.[12] It is reported more commonly in Caucasian population.[12] No particular risk factor has been identified, nonetheless, some studies have documented advanced maternal age, maternal smoking and alcohol use as risk factors.[12,13] This entity can be identified antenatally with ultrasound but for a more detailed evaluation, a fetal MRI may be done, hence providing a scope of valid counselling for parents to continue the pregnancy.[14] One study has reported elevated maternal serum Alpha-feto protein levels as a marker for prenatal diagnosis but it has very low specificity as it can be raised in a number of other conditions as well.[4]

Medical management of bladder exstrophy is limited to supportive care only. Surgical management is the mainstay. Many changes have occurred in surgical management over the last few years with staged closure of the defect being the recent one.[15] In this approach, primary closure of the bladder mucosa is done in the neonatal period, epispadias closure by 1-2 years of age, and bladder neck reconstruction at the age of 5-6 years. Inguinal hernia being a common association, children with bladder exstrophy should be carefully examined for the same before bladder closure.[8] There is decreased likelihood of primary or recurrent inguinal hernia if pelvic osteotomy with tension free approximation of the pubis is performed as this maintains the obliquity of the inguinal ligament.[16] Children with bladder exstrophy, have impaired overall health related quality of life.

CONCLUSION

Exstrophy - epispadias complex can be diagnosed antenatally on ultrasonography. Clinical examination at birth reveals exposed bladder mucosa through a defect in the abdominal wall, along with epispadias and low lying umbilicus. Baseline renal function tests and imaging of the upper urinary tract should be performed before complex reconstruction of the urinary tract. Inguinal hernia is commonly associated with this anomaly and has a high rate of recurrence even after the corrective surgery. Cloacal exstrophy in past had a high mortality but advanced surgical skills with parenteral nutritional support has led to successful reconstruction and improved survival. However, as there are only few population based epidemiological data available for this rare anomaly, hence standardised protocols for management cannot be drawn.

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