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Case Report

Pregnancy in case of exstrophy of bladder

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ABSTRACT

Bladder exstrophy or ectopia vesica is an unusual congenital anomaly which exists in the spectrum of the exstrophy-epispadias complex. It most commonly involves protrusion of the urinary bladder due to defect in the lower abdominal wall. It has variable presentation and often includes abnormalities of the pelvic floor, bony pelvis and genitalia. Patients undertaking pregnancy after surgical repair of such an anomaly are rare. Planned Cesarean section at term is considered the appropriate mode of delivery. We present a case of 21-year-old patient who had uneventful preterm vaginal delivery following surgical repair of bladder exstrophy in childhood.

Keywords: Bladder exstrophy, Pregnancy

INTRODUCTION

Bladder exstrophy is a rare anterior midline defect with an incidence of 1:30,000- 50,000. It is associated with complex genitourinary malformations, requiring difficult surgical treatment.[1] Because of multi-staged bladder and pelvic reconstructive surgeries in these patients, obstetric management presents a challenge and needs multidisciplinary approach by obstetrician and urosurgeon. Preconceptional renal evaluation and regular follow up in antenatal period should be done. Pregnancy may be complicated by recurrent urinary tract infections, malpresentation, preterm labour and genital prolapse. Due to the rarity of this condition, there is limited literature regarding management during pregnancy.

CASE REPORT

A 21 year old primigravida with corrected exstrophy of bladder was registered at our tertiary care center at 20 weeks of gestation. She was married since one year, conceived spontaneously and had regular antenatal follow up. She had multiple surgeries in childhood for correction of exstrophy of bladder, as follows: on day 16 of life, bilateral transverse iliac osteotomy with functional closure of bladder neck was done. Bladder neck reconstruction and bilateral ureteric reimplantation with ileal bladder augmentation with Mitrofinoff's procedure for urinary diversion was done at the age of 15 years. One year later when she was 16, urethral lengthening with rectus sling with urethral repair was done. At 18 years of age, bladder neck closure with colposuspension was done in view of persistent incontinence and genital prolapse. After the procedure, she was unable to pass urine and had to drain urine through Mitrofinoff's opening in right lumbar region with the help of catheter.

She was admitted at 34 weeks of gestation for safe confinement. Her general examination findings were normal. On abdominal examination, uterus was 32 weeks with single live fetus in longitudinal lie, cephalic presentation, and no uterine activity. Multiple vertical scars of previous surgeries were present. Mitrofinoff's stoma was present in right lumbar region. On speculum examination, second degree uterovaginal descent was present. Clitoris was bifid with widely separated labia. Cervical os was 1 centimeter dilated with poor effacement. Her hematological investigations including renal function tests were within normal limits. Her obstetric ultrasonography was suggestive of single live

gestation of 30 weeks with adequate liquor and without any fetal anomalies. Corticosteroids (injection betnosol 12 mg intramuscularly 2 doses at 12 hourly interval) for fetal lung maturity were given. Urology consultation was sought to decide mode of delivery and further management. The proposed plan of action was to conserve pregnancy and perform elective cesarean section with urology standby at term.

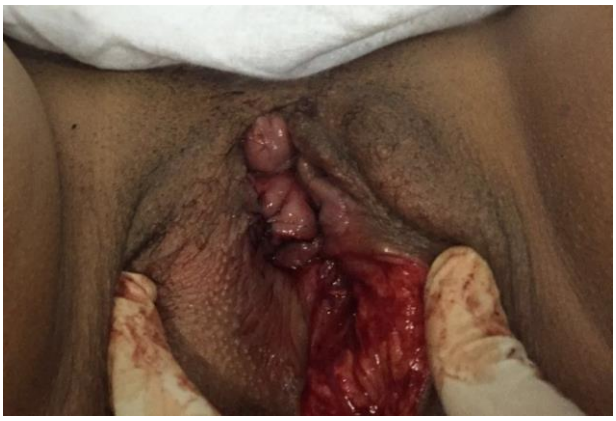

Figure 1: Immediate post-delivery image showing bifid clitoris and mediolateral episiotomy.

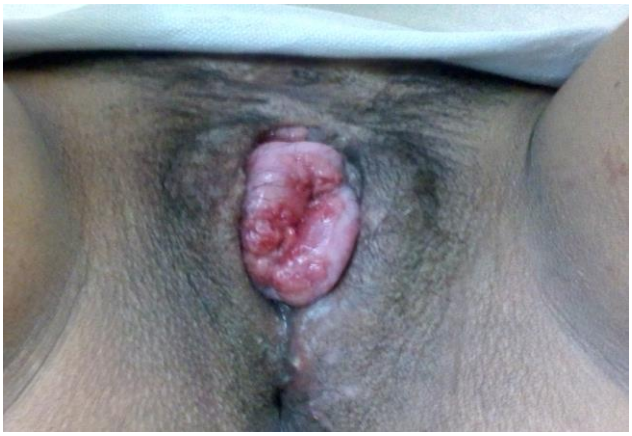

Figure 2: Post delivery after 6 weeks showing second degree uterovaginal descent.

However, patient went in spontaneous preterm labor and had a preterm vaginal delivery at 35 weeks of gestation; male child of 1.8 kilograms was delivered. Mediolateral episiotomy was given. There were no perineal or paraurethral tears. Baby was kept in neonatal intensive care unit in view of low birth weight and prematurity. Patient had uneventful postnatal recovery and she was discharged on day 4.

DISCUSSION

Exstrophy of bladder is a very rare congenital anomaly in which there is a defect in the closure of the lower abdominal wall and exposure of the bladder mucosa, ureteral orifices, bladder neck, and urethra to outer abdominal wall.[1] It is often associated with public bone anomalies. It was first described in 1595. It is one of eight very rare defects which occurs very few times per 100,000 births; the others are acardia, amelia, conjoined twins, phocomelia, cyclopia, cloacal exstrophy and sirenomelia.[2] It is 2.8 times more common in males as compared to females and 1.7 times more common in Caucasians as compared to Blacks, Hispanics and other races.[3]

There is 0.5-3 % risk of recurrence in families with one affected member.[4] It can occur as an isolated syndrome or may be associated with multiple congenital anomalies which include epispadias–exstrophy complex, anal defects, omphalocele, neural tube defects, and skeletal defects such as omphalocele, exstrophy of bladder, imperforate anus, spinal defects (OEIS) complex.[2] Pathogenesis of bladder exstrophy remains unknown.[2] Most probably it occurs due to failure of migration of infraumbilical mesenchyme between the ectodermal and endodermal layers of the cloacal membrane.[2] It is not associated with any specific genetic or non-genetic factors but some chromosomal factors are starting to be identified.[5] The clinical approach to patients with bladder exstrophy is usually challenging for the pediatric urologist.

The aim should be satisfactory closure of the abdomen with preservation of kidneys, urinary continence, and avoidance of complications.[6] Staged reconstruction and complete primary repair are the two basic treatment options available. The staged reconstruction consists of initial closure of the bladder plate and posterior part of urethra, with subsequent steps of repair of epispadias and bladder neck reconstruction. In complete primary repair, closure of the bladder plate and reconstruction of urinary tract through colocystoplasty or ureterosigmoidostomy along with epispadias repair is done simultaneously.[6] Malignant bladder and kidney tumors are more common if reconstructive procedures are not done and they usually occur in third and fifth decades of life.[2] Pelvic organ prolapse is seen in 18% of cases.[7]

The greatest inconvenience in the life of these patients is the need to wear diapers for urinary incontinence and recurrent urinary infections. Irrespective of surgical procedures and gender, these patients have a poor quality of life as compared to the normal population in terms of general health.[8] As per multicentric study based on long-term follow-up of patient with exstrophy of bladder (which included 9 women and 16 men), 5 of 9 women were married; 6 women were sexually active; 2 women had dyspareunia; and one woman denied sexual intercourse because of genital prolapse. 2 women conceived spontaneously, while 1 conceived from artificial insemination due to male infertility.[8] 7 women were active professionals with their education level and employment rates similar to those of the general population.[8] In 1922, the first pregnancy in a patient with ureterosigmoidostomy was documented and since then only 252 cases of pregnancy with bladder exstrophy have

been reported.[9] These patients have higher rate of infertility, ranging from 8 to 32%, most likely due to tubal factors owing to multiple adhesions of previous surgeries. They also have higher chances of preeclampsia, miscarriage and preterm labor.[6] They are also prone to pelvic floor prolapse, recurrent urinary tract infections and hydronephrosis.[10]

Because of these multiple factors, pregnancy is very high risk as well as precious in these cases and requires multidisciplinary approach involving obstetricians and urosurgeons. Preconceptional renal evaluation should be done. These cases should be managed at tertiary care centers. The appropriate mode of delivery is planned Cesarean section at term by an experienced obstetrician with the help of urosurgeon.[11] Because of distorted pelvic anatomy, multiple pelvic adhesions are usually seen during Cesarean section. Complications like fistula formations and ureteric transaction are known to occur. However, a Cesarean section can minimize chances of uterine prolapse, vascular problems involving the augmented ileum and urinary tract complications.[10] Prematurity is a major neonatal risk.

The rate of preterm delivery is as high as 29%.[6] According to Greenwell et al, vaginal delivery should only be considered in uncomplicated cases in presence of senior obstetrician.[11] Deans R et al conducted a study in 2012 to find the reproductive outcomes in patients with bladder exstrophy. As per study results, 66% of cases conceived, 35% had spontaneous abortion, 7% were stillborn, 8% of cases had postpartum hemorrhage. Major complications like fistula formation was seen in 4% of cases and transaction of ureter in 4% of cases.[12]

CONCLUSION

Though pregnancy is high risk for both mother and baby, patients with bladder exstrophy should not be discouraged to have their own children. For better pregnancy outcome, interdisciplinary team work and operative delivery should be planned; emergency situations should be avoided.

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