Case Report

Congenital Uterovaginal Prolapse – A Rare Case Report

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ABSTRACT

Background: Congenital Utero-vaginal prolapse is rare disorder commonly associated with meningomyelocele and rarely with hydrocephalus. Case Characteristics: Unbooked pregnancy with breech presentation delivered new born child with 3\textsuperscript{rd} degree primary Congenital Utero-vaginal prolapse. Observation: Child had severe birth asphyxia, lumbo sacral meningomyelocele, hydrocephalus and CTEV along with genital prolapse. Outcome: The mass was reduced and both lower limbs were strapped together using crepe bandage which was applied in a mermaid fashion to prevent re-protrusion. Message: Maternal supplementation of folic acid at time of conceiving child could prevent congenital genital prolapsed.

Keywords: Congenital uterovaginal prolapse, meningomyelocele, hydrocephalus, folic acid, breech, birth asphyxia.

INTRODUCTION

Uterovaginal prolapse (UVP) presenting at birth is very rare. Very few cases of neonatal UVP have been reported in literature.[8,10,3]. Uterovaginal prolapse is the downward descent and protrusion of the uterus and vagina to the exterior via the introitus. It results from weakness of the cardinal ligaments and uterosacral ligaments which provide support to the uterus. Neonatal genital prolapse is a condition manifesting at birth or within the first few days of life. Defects of innervation to the pelvic floor musculature and ligamentous support can cause a flaccid paralysis, allowing downward protrusion of the abdominal and pelvic organs. Most reported neonatal UVP were either 3\textsuperscript{rd} degree or procidentia.[8,10,3]. Congenital UV prolapse is commonly associated with neural tube defects seen in 80-86\% cases.

CASE REPORT

A 22 yr old lady, unbooked prima gravida, presented in obstetrics and gynaecology department, in labour pain with leaking per vaginum. USG scan revealed oligohydramnios with live fetus at 40 wk of gestation in breech presentation with congenital malformations. Her random blood plasma value was within normal limit. As the baby was congenitally malformed,
she was given trial of labour after patient’s consent. Following non progression of labour emergency caesarian was done under spinal anaesthesia and a live, term, congenitally malformed severely asphyxiated female baby was born.

The mother had single episode of fever during the pregnancy for which she took over-the-counter medications. She neither ingested herbal medications nor any un-prescribed drugs during pregnancy. There was no history of congenital abnormality and diabetes in her family.

On examination, the child weighed 2.3 kg. There was a fleshy, reddish, and edematous mass protruding from the vulva [Figure 1]. The wall was thrown into folds. At the apex there was a blind-ended opening. The urethral opening was located separately above the mass. The anal opening was not present. She had a meningo-myelocele protruding out from her spine in lumbosacral region.[Figure 2].

Lower limb showed CTEV foot deformity. Head circumference was 38 cm which was more than 3SD for age. Anterior fontanelle was wide open with size of 5 by 4.5 cm. Rest of systems were within normal limits. No masses were felt on abdominal examination. The skin was neither wrinkled nor thrown into folds. Skin elasticity was normal. Spinal X-ray and abdominal ultrasound scan could not be performed to exclude VACTER association.

After passing urethral catheter into bladder, the mass was reduced and pushed inwards[4]. To prevent re-protrusion on straining, both lower limbs were strapped together using crepe bandage which was applied in a mermaid fashion extending from the buttocks to the lower legs sparing the anus for defecation[9]. The child was put on oxygen and vasopressor support. Further investigation could not be carried as baby was sick but later child succumbed to her illness at age of 96 hr.

DISCUSSION

Genital prolapse was first documented in the Egyptian medical papyrus, the Ebers papyrus, dated 1550 BC, neonatal prolapse was not reported until 1723. Only few cases have been documented in literature.[8,10,3]. The extent of prolapse may vary, ranging from isolated vaginal prolapse to include the cervix or uterine corpus as well. Most of the cases are are 3rd degree as seen in our case.Differential diagnoses include vaginal polyps, urethral prolapse, paraurethral cysts, and rhabdomyosarcoma. Diagnosis however be readily confirmed with restoration of normal anatomy upon reduction of the prolapse and recognition of associated spinal cord abnormalities.

The etiology of congenital UVP is not well-established. Spina bifida, especially myelomeningocele is the most common identifiable risk factor [8,10,3,4]. In 1955, Malpas separated these two etiologies into primary and secondary causes of neonatal genital prolapse, respectively [7].Our patient would be classified as a case of primary genital prolapse with a neurologic etiology. In the fetus and the neonate there is minimal or no angulation and the orientation is almost vertical, as the true pelvis is not well-formed yet and the pelvic organs are essentially abdominal. Hence, raised intra-abdominal pressure as occurs during prolonged breech delivery is transmitted straight down the uterus and pushing it out through the vagina and introitus.

Our study subject had meningomyelocele which along with breech presentation could result into UVP. Congenital UVP is also seen in neonates with congenital cutis laxa.[9] which is a genetic disorder characterized by loose and redundant skin with reduced elasticity.Hernias and rectal prolapse are common associations in this condition. The index patient did not clinically manifest any such abnormality. Approximately 90% of fetuses with prolapse are born to women with no predisposing factors. However maternal factors such as exposure to valproic acid, thalidomide, or aminopterin and presence of maternal diabetes are etiologically related to this disorder.

Termination of the pregnancy is an option when the diagnosis of spina bifida is made in early in gestation.
These fetuses should be delivered at term, with the exception of those developing a rapidly progressive hydrocephalus. Mode of delivery is controversial. Delivery should take place in a tertiary center, where a team of neurosurgeons, paediatricians, and rehabilitation therapists is available for immediate evaluation.

Such cases can be prevented if an early second trimester level II ultrasound can diagnose a case of neural tube defect (NTD). The present recommendation of the US Public Health Service and the ACOG that all women of reproductive age consume 400 microgram of folic acid to reduce the risk of having a fetus with NTDs. But these preventive measures were not performed in our index case. Successful reduction can be achieved by digital reduction and strapping the buttocks together with crepe bandage in a “mermaid” fashion as was done in our case.

CONCLUSION
To conclude UVP presenting at birth is a rare condition. The possible risk factors present in this patient include breech position and meningomyelocele. Successful reduction was achieved by digital reduction and strapping the buttocks together with crepe bandage in a “mermaid” fashion. Folate prophylaxis at time of conceiving along with early intranatal diagnosis using USG is mainstay of preventing this condition.

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REFERENCES
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Figure 1: Uterovaginal prolapse at birth

Figure 2: Meningomyelocele