Case Report

Potter's Syndrome: A Case Report

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Abstract

Potter's syndrome is a rare congenital disorder characterized by characteristic atypical appearance of a newborn due to the oligohydroamnios experienced during the intrauterine life. Its incidence varies from 1 in 2000 to 1 in 5000. A 24-yr-old primigravida at 39 weeks of gestation was admitted at Monowara Sikder Medical College & Hospital on 30th May 2019 with labour pain. Her ultrasonography revealed amniotic fluid index was zero. On per-abdominal examination, fetal parts were easily palpable but fetal heart sounds were not heard. Per-vaginal examination revealed footling breech. Then she underwent emergency lower segment caesarean section (LSCS) and a still born baby was born with potter syndrome. The fetus had typical facial appearance of "potter facies". Apart from typical Potter's facies, pulmonary hypoplasia, skeletal deformities are associated with this condition, severity depends on the degree and duration of oligohydramnios. Therefore, it is very important to pick up this condition during antenatal ultrasound at earlier gestation so that timely decision regarding further workup and pregnancy termination can be undertaken. This case report emphasizes upon the importance of regular antenatal check ups and examination in each and every patient as it picks up the suspicious cases which can lead to further workup, definite diagnosis of the condition and timely decision regarding management.

Keywords: *Congenital disorder, Potter facies, Potter's syndrome, Footling Breech.*

Introduction

Potter's syndrome is a rare congenital disorder characterized by characteristic atypical appearance of a newborn due to the oligohydroamnios experienced during the intrauterine life. This condition was first described by pathologist Edith Potter in 1946.[1] Its incidence varies from 1 in 2000 to 1 in 5000.[2] Male predilection is seen due to increased incidence of Prune Belly Syndrome and obstructive uropathy due to posterior urethral valve in them.[3] It is accompanied by oligohydramnios which may have different etiologies: Bilateral renal agenesis, obstructive uropathy, renal hypoplasia, polycystic or multi-cystic kidney disease, premature rupture of membranes which results into characteristic physical deformities and lung hypoplasia.[4,5] Main cause of increased morbidity and poor prognosis of fetus is lung hypoplasia. There are increased chances of recurrence in future pregnancies (3-6%).[6]

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Case Report

A 24-year-old primignavida at 39 weeks of gestation who was admitted at Monowara Sikder Medical College & Hospital on 30th May, 2019 with labour pain. Her ultrasonography revealed amniotic fluid index was zero. On per-abdominal examination, fetal parts were easily palpable but fetal heart sounds were not heard. Per-vaginal examination revealed footling breech. Then she underwent emergency LSCS and a still born baby was born with potter syndrome.

Figure 1 Malformed limbs and ambiguous genitalia in Potter's syndrome

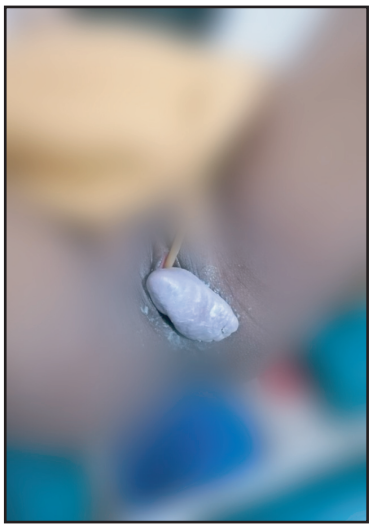

Figure 2 Footling breech presentation

Discussion

Potter's syndrome or sequence is a rare fatal disorder. Some authors believe that Potter's sequence rather than Potter's syndrome is a better term to describe this entity because there are different etiologies which lead to the common result of oligohydramnios, which in turn leads to characteristic physical appearance and lung hypoplasia and its sequelae. Potter's facies is described as flat nose, recessed chin, prominent epicanthal folds and low set abnormal ears. Degree of pulmonary hypoplasia in such cases depends on the severity and duration of oligohydramnios. The stage of lungs development is important at which oligohydramnios occurred. Skeletal anomalies frequently associated with this condition are hemivertebra and sacral agenesis.

Oligohydramnios is defined as decreased in liquor amnii for the period of gestation. In intrauterine life, prior to 16 weeks of gestation, proportion of liquor amnii production depends on transmembrane flow, after that it is mainly by fetal urine production. Its exact genetic basis is still not known, studies are going on. Evidence has been found regarding role of transcription factors Lim1 and Pax 2 during nephrogenesis and deficiency leading to renal agenesis in mice.[7,8] Potter's syndrome can be divided into four subgroups depending on the underlying etiology.[9] Classic Potter's syndrome is always fatal.[3] Therefore, it is very important to pick up this condition during antenatal ultrasound at earlier gestation so that timely pregnancy termination can be done.

Workup of patients with Potter's syndrome includes search for other nonrenal defects, autopsy, chromosomal analysis, urologic examination or renal ultrasound of parents. Ultrasonographic prenatal monitoring of further pregnancies should be done in such cases because of chances of recurrence in such families. In present case, age factor of parents, history of early neonatal deaths, history of intake of some medications affecting chromosomal division in first trimester leads to possibility of some genetic causes for bilateral renal agenesis leading to oligohydroamnios leading to potter sequence.

Conclusion

This case report emphasizes upon the importance of regular antenatal check ups and examination in each and every patient as it picks up the suspicious cases which can lead to further workup and definite diagnosis of the condition and timely decision regarding management.

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