International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Malik N et al. Int J Reprod Contracept Obstet Gynecol. 2022 Oct;11(10):2864-2865
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20222493

Case Report

Sirenomelia-the mermaid syndrome: a rare invariably fatal congenital anomaly in a term unsupervised pregnancy

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Received: 27 July 2022
Accepted: 26 August 2022

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ABSTRACT

Sirenomelia is a rare congenital anomaly with an incidence of 0.8 to 1 case per 1,00,000 births. The prognosis is grim due to associated genitourinary and gastrointestinal anomalies. Antenatal registration in the first trimester and timely ultrasound go a long way in detection of the anamoly when termination can be still be offered and the mental agony of giving birth to a term neonate with a fatal congenital anomaly can be avoided.

Keywords: Sirenomelia, Caudal regression syndrome, Mermaid syndrome, Potter's facies

INTRODUCTION

Sirenomelia is a rare and invariably fatal congenital anomaly with an incidence of 0.8 to 1 case per 1,00,000 births.[1] It invariably presents with lower limb fusion, sacral and pelvic bony anomalies, absent external genitalia, imperforate anus, and renal agenesis or dysgenesis. Sirenomelia is the most severe and devastating end of the caudal regression syndrome spectrum. Although etiology is not clear, maternal diabetes, genetic predisposition and vascular hypoperfusion have been postulated to have a causal association with caudal regression syndrome.[2,3] It is sometimes associated with twin pregnancy, most often monozygotic. The syndrome of caudal regression is postulated to be the result of injury to the caudal mesoderm at 28 to 32 days of gestation leading to vascular hypoperfusion to the lower limbs.

CASE REPORT

A 26-year-old unbooked gravida 2 para 1 with 35 weeks pregnancy in late first stage of labor came to the obstetric emergency of our hospital. She had no antenatal supervision. An ultrasound scan done a week back in the peripheral health facility showed IUGR and oligohydramnios. No comment was made on any gross congenital anomaly. There was no history of maternal diabetes. Her marriage was non-consanguineous and there was no history of exposure to any known teratogenic drug or radiation during the first trimester. On examination her vital signs and cardiorespiratory system was normal. On obstetric examination fundal height was 32 weeks with a single live fetus in cephalic presentation. Liquor appeared decreased. She was 6-7 cm dilated and membranes were present. Two hours after admission she delivered a baby with birth weight 1.6 kg. On examination, the neonate had features of Potter's facies including low set large ear and a receding chin. There was complete fusion of the lower limbs from the perineum to the ankles and one foot was present. The external genitalia and anal orifice were absent. A small phallus like structure was seen in the back with an orifice from which urine was being passed. The baby died an hour after birth.

Figure 1: The neonate.

Figure 2: Baby died an hour after birth.

DISCUSSION

In the early second trimesters, amniotic fluid is sufficient to allow detection of abnormal lower limbs, undetermined external genitalia, anorectal atresia and lumbosacral agenesis.[5] Therefore, the emphasis on early registration during pregnancy with antenatal supervision and timely level 2 scanning can help in alleviating the suffering associated with giving birth to a non-viable congenitally anomalous baby at term. The reported percentage of elective termination of pregnancy for the fetal anomaly is about 49.5%.[5] Our patient had an unsupervised pregnancy and ultrasound done at 8 months in the periphery was sub optimum probably due to oligohydramnios. Maternal diabetes, retinoic acid, heavy metals and teratogen exposure, genetic variables, monozygotic twins, male gender and the mother's age of 20 years or >40 years were all risk factors for sirenomelia malformation.[3-5] However, none of these conditions were there in the present case.

CONCLUSION

Mermaid syndrome or sirenomelia is an invariably lethal congenital anomaly with a grim prognosis due to associated vertebral, urogenital and gastrointestinal abnormalities. With antenatal supervision and timely diagnosis using ultrasonography termination of pregnancy may be considered.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Malik N, Jain S, Chaudhary S, Kumari B, Madan N, Verma R. Sirenomelia-the mermaid syndrome: a rare invariably fatal congenital anomaly in a term unsupervised pregnancy. Int J Reprod Contracept Obstet Gynecol 2022;11:2864-5.