Both babies with sirenomelia’ in twin pregnancy: a case report and review of literature

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ABSTRACT

Sirenomelia, also called as ‘Mermaid syndrome’, is a rare congenital malformation of uncertain aetiology. It is characterised by fusion of the lower limbs and commonly associated with severe urogenital and gastrointestinal malformation. We report a case of twin pregnancy with both babies had sirenomelia. No previous case of ‘both babies with sirenomelia’ in twin pregnancy has been reported till date.

Keywords: Twin, Sirenomelia, Mermaid syndrome, Oligohydramnios

INTRODUCTION

Sirenomelia, a very rare congenital anomaly, in which fused lower limbs, rudimentary or absent genitalia and absent excretory pores result in fish-tail like external appearances of lower extremity of the foetus resembling the ‘mermaids’ of Greek mythology, hence commonly known as ‘mermaid syndrome’. It is equally been referred as symmelia, sympodia monopodia, sympus. The reported incidence of sirenomelia is approximately 1 in 100000. It is a lethal congenital condition having internal anomalies. Many of these cases reported worldwide. We are presenting a unique case where both babies born out of twin pregnancy had sirenomelia. After a thorough search of literatures, to best of our knowledge, no previous case of ‘both babies with sirenomelia’ in twin pregnancy has been reported till date.

CASE REPORT

Mrs. LP, 19 years, G1P0+0, married for 1yr, from Balurghat, of poor socioeconomic status, conceived spontaneously, admitted at Malda Medical College and Hospital, West Bengal at 32 weeks of gestational age with complains of diminished foetal movement being referred from local health facility. Her antenatal care was inadequate. USG was not done. It was not a case of consanguineous marriage. There was neither any history of twining or congenital anomaly in the family nor any family history of diabetes mellitus. There was no history of exposure to toxins or drugs or any significant febrile episode in antenatal period. Her antenatal blood glucose and Hb% were normal. On admission USG revealed twin live foetal gestations with both foetuses on transverse lie, 2nd foetus having an encephalocoele. USG revealed corroborated gestational age but gross oligohydramnios and moderate IUGR. Unfortunately no other anomaly was commented. Decision of LUCS was taken as both foetuses were in transverse lie with severe oligohydramnios. LUCS was done with Inj. Betamethasone given, considering at least one salvageable baby. But both babies delivered out of single amniotic sac (monochorionic monoamniotic). Both had fused lower extremities, absent genitalia and absent anal and urethral orifices. The lower extremity of one baby was simply tail like (simpus apus). The other baby also
had single fused tapering lower extremity, having two legs fused together side to side, with a single foot directed posteriorly (simpus unipus) with few digits. Both babies had prominent infraorbital folds, hypertelorism, abnormal ear lobe ation, small receding chin and flattened nose tip suggestive of ‘Potter’s facies’. One baby had an occipital encephalocele. They were respectively weighed 1.2 kg and 1 kg., non-vigorous with very poor APGAR scores. Both were connected to a single placenta with two different cords. Both cords were 2 vessels cords suggestive possibility of single umbilical artery in each cord. They were quickly shifted to SNCU but died within hour. Patient party removed the bodies and refused further post-mortem investigation (autopsy and radiological) as such congenital malformations are considered as effect of evil-spirit from their local belief. Maternal diabetes often associated with renal agenesis, absent or hypoplasia of the vasculature distal to the artery leads to poor perfusion of the caudal region and excess RA (RA), causes reduced Bmp signalling in the caudal region and limb abnormalities. Some researchers suggest sirenomelia is a severe form of caudal regression syndrome and VACTERL (‘vertebral defects, anorectal atresia, cardiac abnormalities, tracheo-oesophageal fistula, renal and limb abnormalities’) association due to overlapping features. Hypothesis suggestive of defective blastogenesis in 3rd week of intrauterine life at the phases of gastrulation results in dysgenesis of caudal region of the fetus. There is genetic hypothesis also. Sirenomelia occurs in mice lacking Cyp26a1, an enzyme that degrades Retinoic Acid (RA), causes reduced Bmp signalling in the caudal embryonic region and excess RA. But sporadic nature of human sirenomelia is a contrast of genetic-model for mice. Duhamel proposed the lower limb fusion theory and fixed criteria for “mermaid syndrome”. Some authors describes three simple types, simpus apus (no feet, one tibia, one femur), simpus unipus (one foot, two femur, two tibia, two fibula), simpus dipus (two feet and two fused legs (flipper like)). Sirenomelia is classified on the basis of skeletal elements present in lower extremity, given in Table 1.
Table 1: Classification of sirenomelia.

<table>
<thead>
<tr>
<th>Type</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>All thigh and leg bones are present</td>
</tr>
<tr>
<td>II</td>
<td>Single fibula</td>
</tr>
<tr>
<td>III</td>
<td>Absent fibula</td>
</tr>
<tr>
<td>IV</td>
<td>Partially fused femurs, fused fibulae</td>
</tr>
<tr>
<td>V</td>
<td>Partially fused femurs</td>
</tr>
<tr>
<td>VI</td>
<td>Single femur, single tibia</td>
</tr>
<tr>
<td>VII</td>
<td>Single femur, absent tibia</td>
</tr>
</tbody>
</table>

**CONCLUSION**

Sirenomelia is a rare congenital anomaly, usually incompatible with life. Severe oligohydramnios should raise the suspicion of sirenomelia. A proper antenatal check-up with ultrasound examination should diagnose the cases in antenatal period. If detected early, voluntary termination of pregnancy can be advised in this near-lethal congenital malformation. Though there are few reports of infant wellbeing, newborn survival in such malformed babies is still a far-reached goal in our resource-poor set-up. There is scope of further research in such rare congenital anomalies.

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**REFERENCES**


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