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LETTER TO EDITOR (VIEWERS CHOICE)

SIRENOMELIA OF POSTNATAL DIAGNOSIS ABOUT A FRESH STILLBORN

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Sirenomelia, defined by Stevenson as "a limb deformity in which the normally paired lower limbs are replaced by a single midline limb".[1] This rare and frequently fatal birth defect occurs with a frequency rate of 0.8-4 per 100,000 pregnancies.[2,3,4] The correct causes are obscure. Still, a few chance variables have been recognized, including maternal diabetes mellitus, teratogenic drugs, genetic susceptibility, vascular hypoperfusion, cocaine use, exposure to landfill water and maternal age being less than 20 years or greater than 40 years.[5,6,7,8] It is more common in monozygotic twins and males.[3,9] Associated anomalies include absent or atypical external genitalia, imperforate anus, rectal atresia, absent urinary bladder, single umbilical artery, renal agenesis, esophageal atresia, omphalocele, pulmonary hypoplasia, cardiac defects, diaphragmatic hernia, lumbosacral/pelvic bone abnormalities and spina bifida.[10] Prenatal diagnosis can be achieved with sonography in the first trimester, identifying symptoms such as nuchal translucency, fused or single lower limb, renal agenesis, a single umbilical artery and oligohydramnios.[11]

A 30-year-old woman, gravida three, para three, with two healthy children aged eight and six years, was admitted for a cesarean section at 35 weeks gestation due to severe oligohydramnios and fetal distress. She had no personal or family history of diabetes and had only taken iron and folic acid supplements during pregnancy. The parents were non-consanguineous and reported no conditions or birth defects in their family history, nor was there any history of radiation exposure. The pregnancy was poorly monitored. The baby did not cry within the golden minute after birth and exhibited severe birth defects, necessitating transfer to the SCANU of a secondary healthcare facility for better management. APGAR score was 4. The newborn was resuscitated for 20 minutes but unfortunately passed away due to cardiac and respiratory arrest. The newborn exhibited severe birth defects, resembling the mermaid syndrome. Weight of the baby was 2300 gm, length was 50 cm and OFC was 35 cm. Authorization for post-mortem examination was declined by the parents. The newborn was declared stillborn with third gender. The genetic study like karyotyping, SRY and WES genes was not possible in our setting. The newborn was categorized as type 1 according to the Stocker and Heifetz classification of sirenomelia by infantogram. Ultrasonography revealed bilateral renal agenesis with absent urinary bladder.

The vitelline artery steal theory suggests that the abnormal presence of a large umbilical artery, causes ischemia to the caudal region of the embryo. The inadequate blood flow disrupts the normal development of the inferior region of the body structures. The defective blastogenesis may cause the fusion of the lower part of the body, malrotation, or dysgenesis. As sirenomelia is a serious birth defect and incompatible with life due to pulmonary hypoplasia and renal failure resulting from renal agenesis, medical termination of pregnancy is admissible.

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