

Prenatal Diagnoses of Mermaid Syndrome in a Twin Pregnancy- A Rare Case Report

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Abstract

Sirenomelia which also is known as “mermaid syndrome” affects 1/60 000 - 1/100 000 of all births. Due to fusion of the lower limbs, foetus give the appearance of a mermaid’s tail. In this study a case of sirenomelia is reported and the main criteria for early detection are discussed.

A 25-year-old woman was referred to the radiology department of Royan institute for anomaly scan ultrasound. The Two-dimensional ultrasound revealed a twin pregnancy. First fetus was normal in ultrasound survey. In other fetus lower limb fusion of the fetus was depicted. A fused femur and fused tibia by further Three-dimensional ultrasound imaging revealed the typical view of sirenomelia. There was no evidence of renal agenesis.

She was referred a perinatologist. The reduction procedure was carried out for sirenomelia. A week later, the heart rate of first twin was not detected in ultrasound survey. The woman underwent induction and delivered both of them.

In conclusion two-dimensional and three-dimensional ultrasound in first stage of pregnancy can detect this kind of fatal anomaly for better decision and sooner termination of pregnancy. Magnetic resonance imaging and three-dimensional ultrasound can help earlier exact prenatal diagnoses of this type of abnormality.

Keywords: Sirenomelia; Three-Dimensional Sonography; Two-Dimensional Sonography; MRI

Introduction

Sirenomelia which also is known as “mermaid syndrome” and “Symelia dipus /apus” is a lethal and rare congenital abnormality. Due to fusion of the lower limbs, foetus give the appearance of a mermaid’s tail. It’s estimated the incidence is between 1/60 000 and 1/100 000 of all births [1,2].

First, this syndrome was introduced by Rocheus in 1542 and Palfyn in 1553 [3]. It is the most severe form of caudal regression syndrome and often occurs in diabetic mothers [4]. In more than fifty percent of all cases, stillbirth happens [4,5].

Prenatal diagnosis of the sirenomelia is possible during pregnancy by ultrasound. The diagnosis is easier in first trimester, since amniotic fluid is normal and its source is placenta and membranes. But in second trimester it may be difficult due to oligohydramnios caused by renal agenesis [5]. So early diagnosis is important in pregnancy.

In this study a case of sirenomelia is reported. Three Dimensional Ultrasound (3D-US) after 2- Dimensional Ultrasound (2D-US) was applied for earlier and more exact detection.

Case Presentation

A 25-year-old woman, gravida 1, para 0, was referred to the radiology department of Royan institute for anomaly scan ultrasound the maternal medical, drug, obstetrics and family histories were insignificant.

The 2D-US revealed a monochorionic -diamniotic female twin pregnancy.

Biometric parameters (Bi Parietal Diameter (BPD), Head Circumference (HC), Abdominal Circumference (AC) and Femur Length (FL)) corresponded to 18 weeks of gestational age. A single posterior placenta was reported.

First fetus was normal in ultrasound survey. In other fetus upper extremities and spine were normal. Amniotic fluid index was in the normal range and kidneys and bladder were seen.

Lower limb fusion of the fetus was depicted (Figure 1). A fused femur and fused tibia by further 3D-US imaging revealed the typical view of sirenomelia (Figure 2). There was no evidence of renal agenesis.



Figure 1: Fused lower extremities seen in two dimensional ultrasound scan.

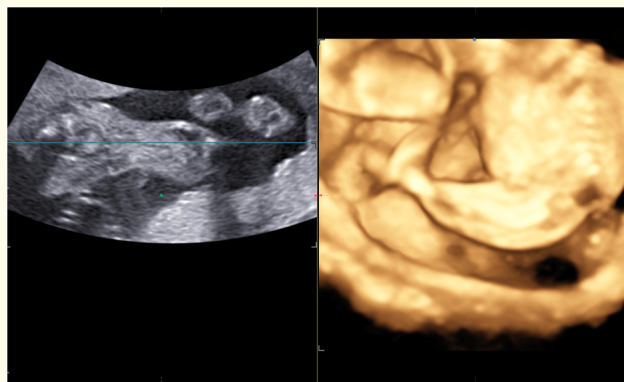


Figure 2A: Three-dimensional image of the fused lower extremities.

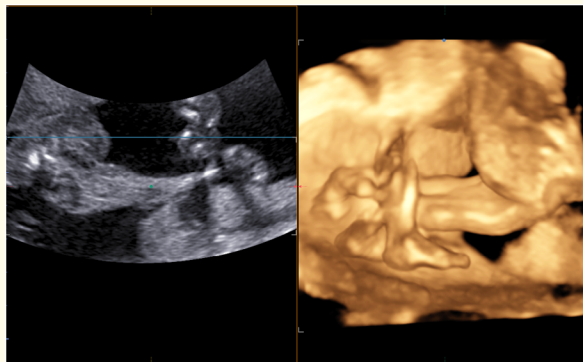


Figure 2B: Three-dimensional image of extremities in twin.

The woman refused doing Magnetic Resonance Imaging (MRI) assessment. She was referred to a perinatologist. The reduction procedure was carried out for sirenomelia. In this method, under the guidance of an ultrasound probe, the fetoscope was guided into the uterus and the location of the umbilical cord on the surface of the fetus’s abdomen was determined. Then bipolar forceps were guided to the desired location with the help of a fetoscope and the blood supply to the sirenomelia was completely cut off by coagulation of the umbilical cord and the sirenomelia was reduced.

A week later, the heart rate of the normal twin was not detected in ultrasound survey. The woman underwent induction and delivered both of them. All procedures performed in the study were in accordance with the ethical standards of the Medical Ethics Committee of Royan Institute, Iran. The written informed consent form was obtained from patient.

Discussion

Sirenomelia is more common in one of two monozygotic twins and shows a male to female ratio of 2.7 to 1. It is remarkable issue that although 10%-15% of cases occur in twins, conjoin twin with Sirenomelia feature have not been reported [1]. In spite of criteria for diagnostic of sirenomelia in pregnancy, yet such cases are reported at delivery time. Early detection is important and the risk factors should be considered [6].

The main risk factor of Sirenomelia is diabetes. Considering the fact that our case was normal in terms of medical history and there were no history of such a rare anomaly in her family, exposure to other risk factors was suspected. Other risk factors are exposure to air pollution, cocaine and tobacco and alcohol, cadmium, lithium, phenytoin, sodium valproate, carbamazepine, warfarin, methylergonovine, diethylpropion, trimethoprim, and ochratoxin. Moreover, she was not drug or substance user; therefore, unknown exposure to the other materials was presented [7].

Prenatal diagnosis in first stages is determinant since diagnosis is difficult at late trimester due to oligohydramnioses induced by renal agenesis [8]. It depends on sufficient amniotic fluid for better detection [2]. Sonography and MRI are both diagnoses and treatment aides tools in such cases [9]. 3D-US is useful in first and early second trimester [2]. Skeletal and its reconstruction feature and soft tissue approach in Three- Dimensional sonography is strategic application in suspected cases in first stages [1].

Single lower extremity with shortened spine and renal or bladder agenesis is the principle clue in ultrasound assessment. Other findings are listed below:

- Anhydramnios induced by bilateral renal agenesis.
- Absence of normally tapered lumbosacral spine.
- Single umbilical artery [2].

According to Margaret Harry (2017) sirenomelia has 7 types as follows:

I: All thigh and leg bones are present

II: Single fibula

III: Absent fibula

IV: Partially fused femurs and fused fibula

V: Partially fused femurs

VI: Single femur, single tibia

VII: Single femur, absent tibia [9].

This case of sirenomelia was normal in upper extremities, head and neck, spinal cord and spine, visceral abdominal organs. Due to the presence of kidneys, the amniotic fluid was in normal range. Considering that fused femur and fused tibia were seen, our case belonged to the first group of this classification.

Genetic factors and sporadic cases are responsible for sirenomelia. Two hypotheses are recommended for mermaid pathophysiology [2]. The first is due to hypoperfusion of the caudal part of the embryo and the second is based on mesoderm development deficiency [10].

Color Doppler is helpful for vascularization assessment. In such cases, abnormal vascularization in lower parts is seen in aorta and iliac arteries [9].

Considering that 50% of sirenomelia cases are not diagnosed by ultrasound due to oligohydramnios in the third trimester, MRI can be helpful [8].

Conclusion

2D-US, 3D-US in the first stage of pregnancy can detect this kind of fatal anomaly for better decision and sooner termination of pregnancy. MRI and 3D-US can help with earlier exact prenatal diagnoses of this type of abnormality. Special consideration should be given to ART (Assisted Reproductive Technology) patients for early detection of anomalies.

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