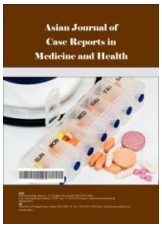

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Twin Babies, One Born with Gastroschisis & the Other with Sirenomelia in Fallujah Maternity & Children Hospital, Fallujah City, Iraq

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Author's contribution

The sole author designed, analysed, interpreted and prepared the manuscript.

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Case Report

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ABSTRACT

Back Ground: Both Sirenomelia & Gastroschisis are very rare congenital anomalies & it is extremely rare to find each of the two anomalies in both twins of one pregnancy, here we reported the two anomalies in one twin pregnancy.
Case Report: We reported twin pregnancy in G3p0A2 19 years old healthy woman who had irregular antenatal visits, ultrasound exam done 4 times during pregnancy, all shown monochorionic, diamniotic twins with polyhydromnia & Gastroschisis, abnormal spine and sacrococcygeal teratoma in the 1st twin and oligohydromnia in the 2nd twin in which the anomaly was not clear & later on the baby born with Sirenomelia.
In all the references we reviewed we cauldn't find the two anomalies in one twin pregnancy and this case was considered to be the 1st reported case globally.

Keywords: Twin; sirenomelia; gastroschisis; Fallujah.

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1. INTRODUCTION

Gastroschisis is abirth defect of the abdominal wall which can be small or large through which the intestine and some times other organs such us stomach & liver protrude outside the babie's body. It occurs early during pregnancy when the muscles that make up the babie's abdominal wall do not form correctly. Because the intestines are not covered by aprotective sac & are exposed to the amniotic fluid, the intestines can become irritated causing them to shorten, twist or swell [1]. The incidence is 0.4-3/10.000 live [1], It's etiology is unknown in most cases, some studies suggested some risk factors for it's occurrence like young age (teenage) mothers, Alcohol & tobacco smoking during pregnancy. It can be diagnosed early during pregnancy by ultrasonography [1].

Sirenomelia, also called Mermaid syndrome, is arare congenital anomaly of uncertain etiology, characterized by fusion of the lower limbs & commonly associated with severe urogenital & gastrointestinal malformation, it should be suspected in antenatal cases presenting with severe oligohydromnias & IUGR [2]. The incidence is about 1.5-4.2/100.000 births, it is 100-150 times more likely in identical (monozygotic twins) than in singletons or fraternal twins [3].

In our hospital we previously reported 3 cases of unusual anterior abdominal wall defect in an article published at 2012 [1] and 2 cases of Sirenomelia also published at 2013 [2], but it was the 1st time to document the 2 conditions in 2 twins babies in the same pregnancy and it is the 1st reported case globally.

2. CASE PRESENTATION

The mother was 19 years old, previously healthy, G3p0A2, of medium socio-economic status, non consanguinous marriage. No family history of any kind of congenital anomalies from both paternal sides. She gave history of 2 previous miscarriages (at 6 & 8 wks successively) due to unknown cause about 1y before the last pregnancy. She had negative antenatal serology, not diabetic nor hypertensive, and gave no history of having smoked or drank alcohol which is not allowed in our culture, there was no history of exposure to toxins or drugs or any significant febrile episode in the antenatal period. She had irregular antenatal visits, ultrasound exam done 4 times during pregnancy, all shown monochorionic, diamniotic twins with polyhydromnia & Gastroschisis, abnormal spine and sacrococcygeal teratoma in the 1st twin and oligohydromnia in the 2nd twin in which the anomaly was not clear and the mother was told that the condition of the 2nd twin was suspicious, that is why it was difficult to take the decision for the termination of pregnancy.

EDD was 10/7/2019. The last antenatal us done on 30/3/2019 in privet Obstetric Us clinic & it was decided that the 2nd twin looks normal (Figs. 1 and 2).

On 30/4/2019, the mother was at her 32nd wk of pregnancy presented to our hospital with labor pain and was admitted to the labor room, the twins were delivered by spontaneous vaginal delivery, the 1st born alive, weighing 2200 gm, with fused lower extremities, absent genitalia

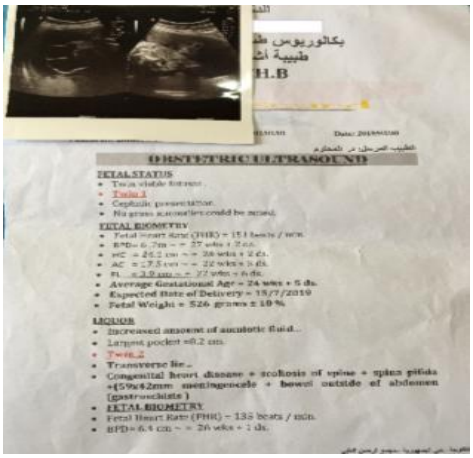

Fig. 1. 2nd twin looks normal

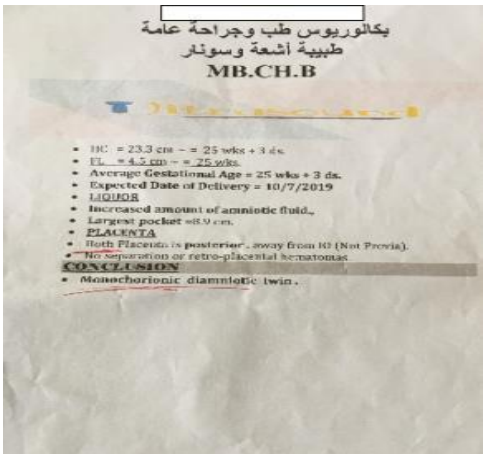

Fig. 2. 2nd twin looks normal

Fig. 3. 1st twin born with Sirenomelia, the 2nd with Gastroschisis

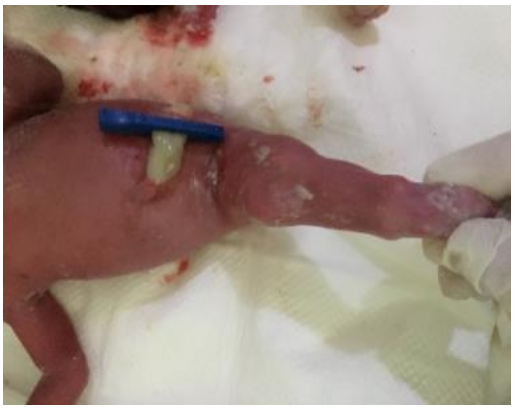

Fig. 4. 1st twin with fused lower limbs and absent genitalia

Fig. 5. The 1st twin, head, face and trunk look normal

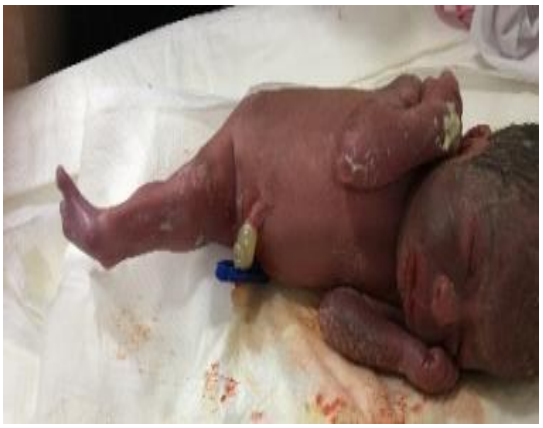

Fig. 6. Appearance of single foot with 3 abnormal separated toes

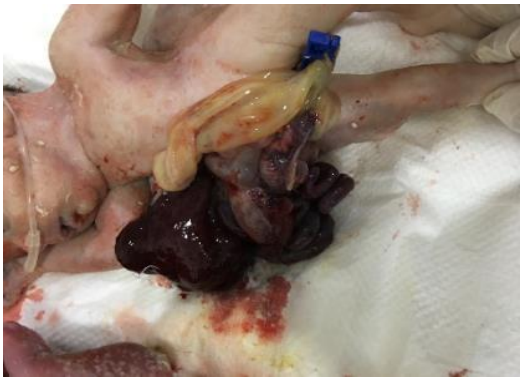

Fig. 7. The 2nd twin, liver, stomach and bowel herniated through abdominal wall defect

Fig. 8. Gastroschisis with deformed lower limbs

and absent anal and urethral orifices with appearance of single foot containing 3 abnormal separated toes, the babie's face, trunk and upper limbs were apparently looking normal, not dyspneic or cyanosed, investigation including skeletal x.ray exam and abdominal and pelvic sonography were requested but the father refused any interference and discharged the baby on his own (Figs. 3-6).

Fig. 9. The 2nd twin, spina bifida and sacrococcygeal swelling

The 2nd twin was male, weighing 2000 gm, born dead (stillbirth) with abig defect (about 3x4 cm) on the right para-umbilical area with the liver, stomach, small & large bowel herniated outside his body, distorted spine with big sacrococcygeal swelling shown by the prenatal ultrasound as spina bifida & meningocele and abnormal right knee with the leg & foot rotated upward in addition to congenital heart disease (Figs. 7-9)

3. DISCUSSION AND CONCLUSION

Sirenomelia is a rare congenital anomaly characterized by partial or complete fusion of lower limbs and usually associated with other severe anomalies. It is considered, by many authors, as severe form of caudal regression syndrome. The associated anomalies may include bilateral renal agenesis, complete or partial agenesis of genitourinary system, imperforate anus, absence or ambiguous external genitalia, single umbilical artery, lung hypoplasia and vertebral and cardiac anomalies [4]. Etiology of sirenomelia is uncertain and various theories have been proposed to explain its origin. An embryonic insult to caudal mesoderm between 28-32 days of gestation and vascular hypo-perfusion has been proposed as possible factors. Others associated the condition with maternal diabetes mellitus, exposure to teratogens and genetic predisposition Sirenomelia carries a grim prognosis, with survival dependent on the nature of the visceral anomalies [5].

In our patient (the 1st twin), the associated anomalies were complete agenesis of urogenital system and imperforated anus, Ultrasonography and skeletal radiology to assess for the association of other abdominal or pelvic anomalies were not done because the babie's father refused any interference and discharged the baby from the hospital on his own responsibility.

Abdominal wall defects can be identified prenatally via ultrasound or measurement of alpha-fetoprotein levels (AFP) within the amniotic fluid, Alpha-fetoprotein is the fetal analog of albumin, and maternal serum AFP (MSAFP) reflects the level of AFPin the amniotic fluid [6].

In gastroschisis, MSAFP is markedly abnormal; up to 9 times the median value [2&6]. Gastroschisis is aright paraumbilical defect involving all layers of the abdominal Wall6. The small bowel always eviscerates through the defect and is, by definition, non-rotated and lacking secondary fixation to the posterior abdominal wall [7]. The loops of bowel are never covered by a membrane; hence, they are directly exposed to the amniotic fluid [2,8, 9,10].

In our patient (the 2nd twin) in addition to the presence of big abdominal defect with herniation of most of the visera there was also spina bifida and meningocele in addition to the abnormal right knee with the leg & foot rotated upward.

Having these 2 major anomalies in the same pregnancy in addition for the history of two previous abortions, in the absence of any family history of congenital abnormalities, may suggest the presence of serious genetic damage which can not be confirmed with our very limited facilities.

From this and all our other previous reports and studies during the last 10 years [1,2,11,12,13,14] we conclude that the high prevalence of birth defects in Fallujah is impairing the population's health and it's capacity to care for the surviving children especially in the absence of sufficient facilities for the prenatal and postnatal management of cases of birth defects and also the absence of clear serious future plans for improvement of the health system or taking serious measures to clean the post wars contaminated environment.

CONSENT

Consent for publication was obtained by the patient's family.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Author has declared that no competing interests exist.

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