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Case Report

Mermaid Syndrome (Sirenomelia): With Use of Mifepristone and Misoprostol in Early First Trimester with Failed Medical Abortion and Continuing Pregnancy in A Multipara with Bad Obstetric History

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Abstract: Mermaid syndrome (Sirenomelia) is a congenital structural anomaly characterized by an abnormal development of the caudal region of the body with different degrees of fusion of the lower extremities giving them the appearance of a mermaid's tail; resembling the mermaid of Greek mythology. This deformity is also known as Symmelia, Symposia, Sympus, Uromelia and Monopodia. It is a rare congenital deformity with an incidence of approximately 1 case per one lakh births with male: female ratio of 3:1. The precise aetiology is not known but diabetes, genetic, irradiation and teratogenic factors are often incriminated. We are reporting a case of mermaid syndrome associated with unsuccessful medical abortion after Mifepristone and Misoprostol used at 6th wk of gestation in a multipara with bad obstetric history which is probably the first such case in our knowledge.
Keywords: Mermaid syndrome (Sirenomelia), congenital structural anomaly

INTRODUCTION

Mermaid syndrome or Sirenomelia named after the mythical Greek Siren was first described by Rocheas in 1542 and Palfya in 1553[1].This condition is seen in approximately one out of every 100,000 live births[2]. The complete or partial fusion of the lower limbs is the most striking feature of this malformative disorder.

It is a congenital anomaly including renal agenesis, ambiguous external genitalia, imperforate anus and blind intestinal loop[3]. Occasionally double inferior Vena cava and angio matous lumbosacral myelocystocele are reported as well[4].This condition is 100 times more likely to occur in identical twins than in single births or fraternal twins[5]. About 300 cases have been reported in the world literature so far of which eight have been reported in India.

We are reporting a case of mermaid syndrome associated with unsuccessful medical abortion after Mifepristone and Misoprostol used at 6thwk of gestation and with bad obstetric history and higher birth order which is probably the first such case in our knowledge.

CASE REPORT

A 28 year old 9th gravida female with history of 4 abortions and 4 live female children consulted the co-author and gynaecologist for termination of pregnancy at 6th week of amenorrhoea and was prescribed Mifepristone with Misoprostol for medical abortion. The mother had 2 female children (alive) followed by 2 spontaneous abortion both at 3 months of pregnancy followed by 3rd female child (alive) followed by 2 spontaneous abortion both at 2 months of pregnancy and a 4th female child (alive).She consulted after 4 days for no bleeding or abortion, but she refused the advice of surgical abortion and decided to continue pregnancy despite adequate counselling. She was tested negative for HIV&VDRL blood pressure and blood sugar levels were normal with haemoglobin level of 7.5 gm/dl. Other anti natal checkups including urine examination were also normal, follow up ultrasound done at 10th week suggested single live foetus with

suspected congenital malformation (Caudal regression) and 4 D serial scan was advised. Further imaging was refused by parents. The patient consulted at 32 weeks of gestation and the Ultrasound done at 32 weeks was suggestive of heart shifted to right side, non visualisation of kidney and underdeveloped genitourinary system with non visualisation of bladder, spine showed multiple deformities with fusion of cervical vertebrae and poor visualisation of lower spine, Sacrum was absent with poorly visible iliac bones with decreased distance between two femur ,two tibia and two fibula were present but foot bones were not visualised . The patient decided to continue pregnancy till term despite adequate counselling and presented with labour pains at term, the baby was in vertex presentation and normal vaginal delivery was conducted in hospital(fig 1).

Fig-1: Mermaid baby

The baby with weight 2.9 kg was shifted to NBSU for weak cry, dullness and central cyanosis and congenital malformations. The examination of umbilical cord revealed a single umbilical vein and single umbilical artery .Physical Examination of the infant revealed upward slant of eyes, low set ears, anteverted nostrils, high arched palate, widely spaced nipples, absent external genitalia, urogenital and anal orifice and completely fused non-rotated lower limbs without feet with five malformed digits. Spine had curvature with malformed caudal region but skull was normal. These findings were consistent with the diagnosis of sirenomelia. The baby was declared dead after 1hr of protocol management. The parents refused x ray and autopsy of the baby.

DISCUSSION

The precise aetiology of sirenomelia is not known. Many inconclusive theories have been proposed .Teratogenic agents have been implicated in some reports. The likely potential teratogenic effect of vitamin A is mentioned by Vonlennep et al and cocaine or irradiation exposure as well[6]. Hibelink *et al.;* demonstrated that the intravenous administration of cadmium and lead can produce sirenomelia in the golden hamster [7]. VonLennep *et al.;* described the possible teratogenic effects of Vitamin A[8].There has been reports of foetal abnormalities in pregnancy of women exposed to both Mifepristone and Misoprostol and Misoprostol when the pregnancy was continued unintentionally[14]. The anomalies noted Sirenomelia (Mermaid syndrome) with Mifepristone and Mobius syndrome with Misoprostol were unlikely to have been provoked by either drug; the need for continuous vigilance is evident[9].

Maternal diabetes mellitus, genetic predisposition and vascular hypo perfusion have been proposed as possible causative factors. However in our case mother was not diabetic. Lynch *et al.;* realized an autosomal type of caudal dysgenesis, but no chromosomal defects are discovered in mermaid syndrome[10]. Genetic counseling should be suggested because the mentioned risk of recurrence is 3–5%[11]. In our patient, twin gestation was aborted in gravida 10^{th} at 2 months of gestation after the above case, but none was suggestive of sirenomelia, she is now in follow up at 4^{th} month of pregnancy. Reports indicate a 100–150 times higher incidence in monozygotic twins relative to dizygotic twins or singletons. Duhemmel *et al.;* consider this anomaly as a manifestation of the caudal regression syndrome that is a consequence of abnormal development of structures derived from the caudal mesoderm of the embryo before the fourth week of gestation and extended to various cranio-caudal levels[12].

Potter facies found in our case may be secondary to renal agenesis and are associated with sirenomelia. Prognosis depends on visceral anomalies

and is generally poor because of complications associated with abnormal kidney and urinary bladder development and function. Death is usually due to renal agenesis or dysgenesis, which is also likely in our case[13].

CONCLUSION

Vigilance is advised on continuation of pregnancy after exposure to Mifepristone and Misoprostol and such continuation should be strongly discouraged. Prenatal diagnosis on USG is desired in this lethal entity so that termination of pregnancy can be offered at the earliest. Further studies are needed to establish association of Mifepristone, multiparity, or bad obstetric history with sirenomelia.

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