

SIRENOMELIA: A CASE REPORTS. K. Wankhede¹, S. Shrivastava²¹Professor, Department of Anatomy, Amaltas Institute of Medical Sciences, Dewas.²Professor, Department of Anatomy, Amaltas Institute of Medical Sciences, Dewas.**ABSTRACT**

The incidence of Sirenomelia is thought to be about 1 in 65,000 live births (Oxford Medical database, Dysmorphology). In Sirenomelia, lower limbs are fused together. Common associated malformations include absent genitalia, imperforate anus, lumbosacral vertebral pelvic abnormalities and renal agenesis. Sirenomelia with craniorachischisis totalis (CT) is a rare fetal malformation, only six cases have been reported in English literature (1-5) and none yet from Asia. Our case is of a 26 weeks fetus born to a 20-year-old primi mother and 24-year-old father. It was a lower category Hindu couple. Mother was anemic and was undergoing treatment for anemia. On USG examination of mother showed a single line fetus of 26 weeks placental calcification, marked oligohydramnios. The fetus weighs 250 grams of unidentifiable sex on external examination. There is absence of pubis, definite buttocks and single fused lower limb ending in a bifid foot with 5 toes. X-rays show no skeletal abnormality or absence of bones. Study of internal structures will be discussed later on.

KEYWORDS

Sirenomelia, Anomalies, Agenesis, Primi, Fused, Bifid, Imperforate, Gestation, Amalia.

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INTRODUCTION

The incidence of Sirenomelia is thought to occur 1 in 65,000 live births (Oxford Medical Database, Dysmorphology). In Sirenomelia, lower limbs are fused together. Common associated malformations include absence of external genitalia, imperforate anus, lumbosacral vertebral pelvic abnormalities and renal agenesis.^{1,2,3,4} Sirenomelia with Craniorachischisis Totalis (CT) is a rare fetal malformation. Only six cases have been reported in English literature and about three from Asia.^{5,6,1,2}

CASE REPORT

This case is of a 26 weeks fetus delivered in the Christian Hospital of Indore. The fetus was born to a 20-year-old primi mother and 24-year-old father, belonging to a lower category Hindu couple.

Mother was anemic and was undergoing treatment for it. Her haemoglobin was 7.0. Gram, V.D.R.L. non-reactive, her blood group was AB +ve. At 26 weeks of gestation an Ultra sound examination was carried out which showed a single live foetus of 26 weeks with placental calcification, marked oligohydramnios.

Patient came in II stage of labor and was delivered by breech presentation, gave birth to a fresh still birth.

EXTERNAL EXAMINATION

The weight of foetus is 250 grams and is of unidentifiable sex. Absence of pubes (Fig. 5 and 6), has a definite buttock. There is a single fused lower limb ending in a bifid foot with 5 toes in each foot (Fig. 1-4). The upper limb, head, face and chest shows no abnormality. (Fig. 5-6).⁴

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Fig. 1: External genitalia not formed.³



Fig. 2: Shows both Lower Limbs Fused Together, but feet are free.³



Fig. 3: Shows both Feet Apart

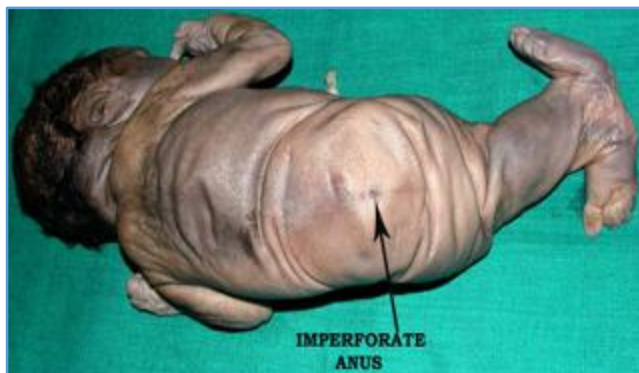


Fig. 4: Shows Imperforate Anus

THE INTERNAL EXAMINATION

It reveals total absence of urogenital system. Liver and spleen appears to be normal. Genitalia not formed. Lungs consolidated. Umbilical cord shows signs of calcification.^{1,3}

RADIOLOGICAL EXAMINATION

The radiological examination was done by taking two radiographs in different positions, which reveals no skeletal abnormality, except the absence of pubic bone.



Fig. 5: Shows Absence of Pubic Bone



Fig. 6: Limb Bones Present

DISCUSSION

This case is somewhat similar to one reported by Ashutosh Halder of S.G.P.G.I.M.S., Lucknow. The only complication during pregnancy was anemia and oligohydramnios. Sirenomelia and craniospinal defect originate much earlier in pregnancy.

Sirenomelia is characterized by single fused lower limb includes multiple defects of contiguous organs such as Urogenital System, rectum and lower vertebrae which constitute a single developmental field.⁴

This sequence is known to be associated with non-contiguous anomalies, such as CNS defect and radial dysplasia, which do not lie in the same field.^{7,8,9,10}

On seeing this case, it can be attributed that during pregnancy every woman should take full precaution during pregnancy regarding drugs, diet, regular exercise, etc. so that the complications or anomalies can be avoided.

CAUSES OF LIMB DEFECTS

1. Genetic: Genetic syndromes, Autosomal disorders
2. Teratogenic: Drugs like thalidomide, dimethadione, cadmium, chemicals, viruses, radiation, hypothermia and hyperthermia.
3. Mechanical: Amniotic bands, Oligohydramnios, Uterine Defects, etc.

CONCLUSION

After the study of this case, we can reach to a conclusion that taking full precautions during pregnancy a woman can avoid

the anomalies which are caused by various teratogenic drugs, unnecessary radiations, oligohydramnios, uterine defects, etc. Every pregnant woman should have a good nutritional diet, regular exercises and regular sleep, should be tension free. By following this she can avoid the complications.

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