JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH

How to cite this article:
NIRUPAMA M, ARAVIND R, RUCHI S, SANDHYA N. BIRTH OF A MERMAID IN THE COASTAL REGION!-SIRENOMELIA. Journal of Clinical and Diagnostic Research [serial online] 2010 December [cited: 2010 December 10]; 4:3680-3682.

Available from
http://www.jcdr.in/article_fulltext.asp?issn=0973-709x&year=2010&volume=4&issue=6&page=3680-3682&issn=0973-709x&id=822

LETTER TO EDITOR

Birth Of A Mermaid In The Coastal Region!-Sirenomelia

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The word 'Sirenomelia' is defined as any condition where the two posterior appendages are fused with each other to form a single conical mass which is suggestive of a body which is fish like in the lower half. It is known as 'mermaid' or 'caudal regression syndrome'. Sirenomelia is a rare congenital anomaly which is characterized by fused limbs or a single lower limb and several visceral anomalies, with an incidence of 1.5-4.2 per 100,000 births. [1]

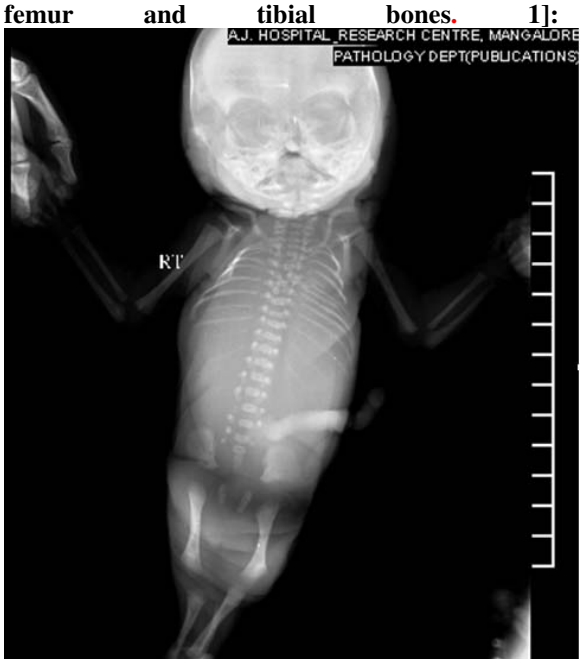
We report here, a similar case in a 26 week old foetus. A 30 year old female gravida two, Para one came with 26 weeks of gestation and was admitted to the hospital for pain in the abdomen. Her past history revealed gestational diabetes in her first pregnancy. After her ultrasound examination, the scan showed oligohydramnios with a foetus having severe intrauterine growth retardation and limb abnormalities and there was bilateral absence of the foetal kidneys.

The pregnancy was terminated and the foetus was submitted for autopsy along with the placenta.

The still born foetus weighed 800 gms. The foetus measured 23 cms. There were facial anomalies in the form of cleft lip and low set ears. There was absence of vertebral anomalies with atrophic muscles at the tibia and both the feet were fused. The external genitalia were absent [Table/Fig-2]. There was no anal opening. The autopsy of the intestine, brain, thymus, heart, lungs, liver and spleen were unremarkable. The vertebral column was unremarkable. The umbilical cord was unremarkable. There was a defect in the anterior abdominal wall, but it was not complete. The placenta was unremarkable. The foetus appeared like a mermaid, with fused lower extremity.

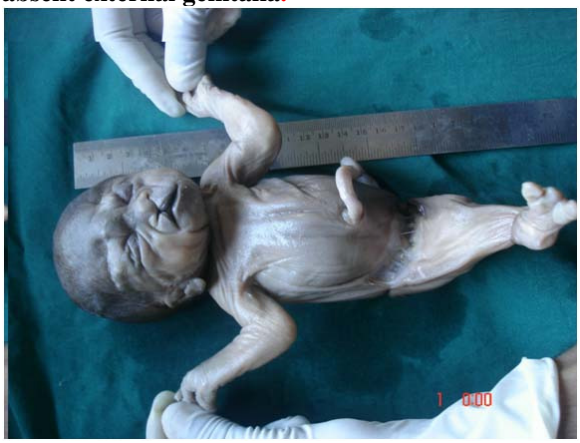
There were fusion of both the lower limbs and the two feet were seen, but were indistinct due to the fusion. Three formed toe buds were seen.

[Table/Fig1]-X-ray of fetus showing separate femur and tibial bones. 1]:



The foetus was subjected to radiological examination, which revealed the fusion of both the lower limbs and the partially fused feet [Table/Fig 2,3].

[Table/Fig 2]: Gross photograph of fetus with absent external genitalia.



The common feature in sirenomelia during development is the presence of a single large artery arising high up in the abdominal cavity, which assumes the function of the umbilical arteries and diverts the nutrients from the caudal end of the embryo, distal to the level of its origin. The steal vessel derives from the vitelline artery complex, an early embryonic vascular network that supplies the yolk sac. The arteries below the level of the steal vessels are underdeveloped and the tissues which are dependent upon them for nutrient supply, fail to develop, are malformed or are arrested in some incomplete stage[2,5].

This sporadic defect occurs more often in males and is more common in monozygotic twins, with the incidence being 150 times greater than in that in singletons. The incidence is very less and accounts to .01-0.16/10,000 live births.[7] It occurs due to a developmental field defect of the posterior axis caudal blastema, resulting in the apparent fusion of the lower limb buds. It occurs in the primitive streak stage, during the third week of gestation, before the development of the allantois and the allantoic vessels are usually absent [3] In many cases, a single umbilical artery that arises from the aorta is seen. In the present case, the umbilical cord was unremarkable. Other defects of the caudal axis include imperforate anus, lower vertebral defects and genitourinary and tracheoesophageal fistula in some cases, which suggest VATER association anomalies.

Cardiovascular respiratory and upper gastrointestinal tract malformation occurs in 20-35% of the cases. [4] The radial agenesis and oesophageal atresia suggest a lesser degree of caudal regression sequence. This sequence is a defect which is most commonly seen in the infants of a diabetic mother [4].

Present case did not show any features of oesophageal atresia or tracheoesophageal fistula, although the mother had a history of gestational diabetes. Renal agenesis or cystic renal dysplasia is known to occur in almost all cases, leading to the potter sequence[3].With experimental confirmation, 'polytopic' field defects were discovered, such as acrorenal field defect, Digeorge anomaly with associated third and fourth branchial arch, craniofacial and cardiac defects, all due to the defective cranial neural crest action and also the Wyer's type of acrofacial dysostosis, which explains the association of the facial anomalies in the present case too. [Table/Fig 2]

Banerjee et al reported two cases of Sirenomelia with skeletal abnormalities and blind loop with imperforate anus[4].

[Table/Fig 3]: Gross picture showing cleft palate and fused lower extremities.

Present case also showed that there was the complete absence of the anal opening [5].

Sirenomelia is fatal and early parental diagnosis is very important. Late diagnosis in the third trimester is difficult as the amniotic fluid levels are decreased. It is very much essential to screen all known diabetics or those with gestational diabetes by sonographical

examination. Early detection and termination helps in a smooth and uncomplicated procedure. Only three known cases of Sirenomelia have been reported, where the patients survived after surgery[7].

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