SIRENOMELUS (MERMAID)

by

P. C. BANSAL

C. H. SATHE*

and

A. M. VARE

Sirenomelus, Symmelus, Mermaid or Sympodia is by no means a common monstrosity reported in world literature. Of all the monstrosities it will probably be 4th or 5th in order of occurrence. It is interesting, both because of the malformation of the lower limbs and because of the commonly coexisting anomalies of the gastro-intestinal and urogenital tracts. It can, however, be said to be a rule to find, in such cases, a totally normal foetus above the level of the diaphragm except for some negligible malformations.

The following case, which has certain points of interest, is presented.

Case Report

K. K., aged 25 years a resident of — Aurangabad, was admitted to the local medical college hospital on 20-11-63 with complaints of labour contractions for approximately 4 hours. She reported to have passed a large quantity of fluid at home. She gave a history of amenorrhoea of 9 months' duration with regular periods previously. She was a 4th gravida, and had had two normal deliveries with an abortion in between. The youngest child was 3 years old and healthy. She emphatically denied having taken any drugs during her pregnancy, even after careful subsequent questioning.

On Examination. She was pale with a pulse of 100/min. and blood pressure of 110/75 mms. of mercury. The uterus was full-term, with a floating breech presenting. Foetal heart sounds could not be heard.

About 2 hours later she delivered a macerated foetus. Her puerperium was entirely uneventful except for a feeling of slight fever and cough on 2nd and 3rd day which was successfully treated. The usual post-natal treatment was given and she was discharged on the 4th day.

Investigations. Haemoglobin 8.2 gms. per 100 c.c. Total leucocytic count 10,000/cmm. urine — normal. Fasting blood sugar 84 mgm./100 c.c.

Foetus. 35 cms. in length weighing 1400 grammes, had only one leg with imperforate anus, and questionable genitalia.

External Appearances. While the external appearances of the face and the trunk were perfectly normal, the abdomen suddenly tapered with a rightward tilt into a single lower limb. The thigh was stout and well formed but the leg and foot were rudimentary and vestigial. There was no anus but it was replaced by a nearly imperceptible dimple in the region. The external genitalia were represented by shallow, empty pouches of overgrown skin and a rounded knob which could have been a rudimentary penis. There were no testes in the pouches and on cutting open the knob a very narrow urethra was seen in the midst of some cavernous tissue and was observed, on probing, to be blind distally. There was

*Asstt. Professor, of Obstetrics & Gynaecology, Medical College Aurangabad (Maharashtra State).

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a small pilonidal sinus with an overhanging flap of skin, concealing it. The neck was moderately extended and was, indeed, very small. There was a sessile body in front of the helix of the left auricle.

Internally. Above the diaphragm: The right lung showed absence of horizontal fissure (hence bilobed). The left lung showed interrupted oblique fissure. The heart was found to be completely normal even after a scrupulous search. The diaphragm was also complete and well-formed. Below the diaphragm: gastrointestinal tract: The stomach and small intestines were normal, but the distal part of the ileum could be traced to the subhepatic region. The caecum and appendix were situated just beneath the liver. The transverse colon traversed the abdomen in an irregular fashion. The descending colon was situated normally but ended blindly just below the place where there should have been a pelvic brim. This blind end was just dangling in the midst of surrounding fascia and peritoneum. Urogenital tract: It was seen that there was complete absence of anything that could have been called kidney, testis or ureter, nor was there anything to represent these structures. The urinary bladder was a cord-like structure from whose apex a fibrous band was traced towards the umbilicus (? urachus). This bladder was fusiform, more or less a solid cord. On opening it, only a capillary tubular lumen was found to exist in its interior.

The suprarenal glands were both present and were of usual size, shape and in position.

Great vessels. The abdominal aorta was not applied to the vertebral column but was rather on a more ventral level. In the upper abdominal region it suddenly drifted away from the lumbar column and travelled downward and ventrally to enter the umbilicus. Its lumen suddenly diminished after the origin of the mesenteric arteries. Beyond this there was nothing on the vertebral column which could even remotely resemble the aorta. This new vessel, which had all the appearances of an umbilical artery of a high origin, was found to give origin to all the arteries that are normally given by the aorta. Thus the left gastric, splenic, hepatic, superior and in-

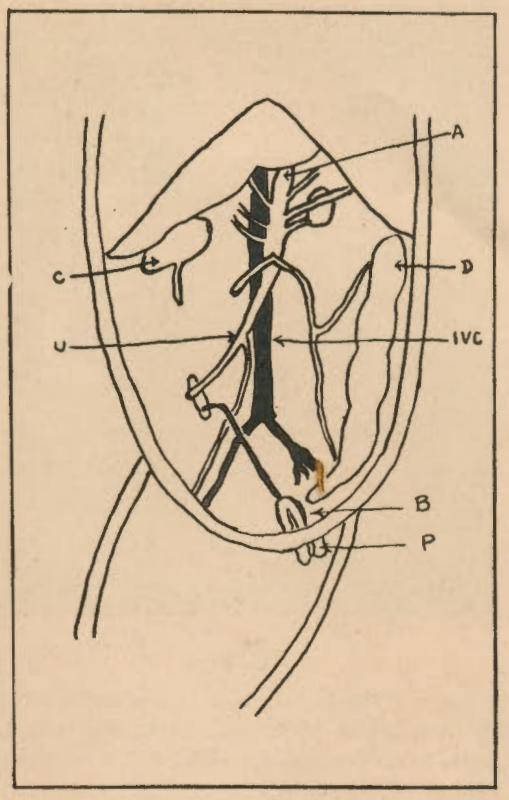

Fig. 1

Sketch showing the anomalies associated with the case.

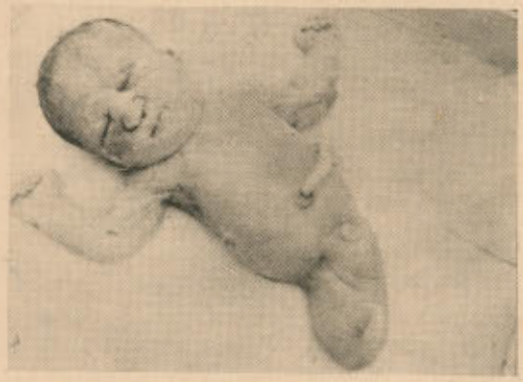

Fig. 2

Photograph showing the Sirenomelus. The limb is deviated to the right and rudimentary external genitalia seen.

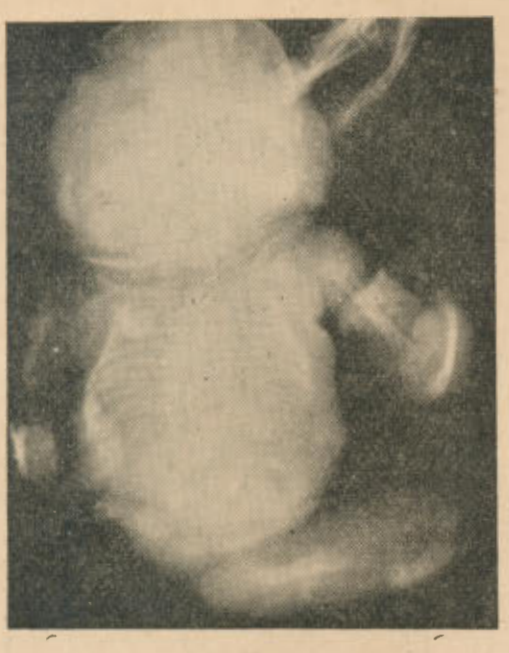

Fig. 3

Skiagram of the foetus. 2 centres of ossification. The tibia is also visualised. Right hip bone is seen as a hazy shadow. Left one is absent.

ferior mesenteric and the suprarenals were all given out by this aberrent trunk. A few arteries which were directed towards the diaphragm were also seen but could not be traced towards their destination because of their extreme fragility. About 5 cms. before entering the umbilicus it gave a branch which went to the only limb present. This may be called the external iliac artery as there was no internal branch.

The inferior vena cava was formed by union of two veins at the usual level. The right common iliac could be seen and the left was issued from an plexus of veins near the lower left part of the trunk.

Limbs. There was no pelvis, nor any public symphysis. A very small tubular interval, just wide enough to admit the tip of the little finger was all that could be called the pelvic cavity.

The left half showed no bony point and there appeared to be a complete absence of hip bone on this side. The right side showed obscure bony landmarks on the upper part but the femur could be well palpated below.

X-Ray of the malformed foetus. The femur was clearly seen in the x-rays, with two centres of ossification at the distal end. The leg consisted of one single tibia; the fibula was not seen at all. The centre of ossification for the upper end was also seen. The hipbone could not be seen clearly on either side, nor were the tarsals or metatarsals visualized. No radiological deformity worth mention was found in the upper part of the skeleton.

Discussion

The description of the case fairly agrees with the classical description of sireniform monsters. Previously it was thought that such an anomaly occurred in female infants, but a review of a large number has established that it is commoner in males. It is difficult to decide on the sex in this case, as though the gonads were missing, there was a tiny knob of skin suggesting a penis with a urethra, though blind, and a little cavernous tissue, which however could not be confirmed by microscopy. The external orifices i.e. genital, urinary or anal were all absent suggesting a complete agenesis of cloaca.

Urogenital Tract

Anomalies of this system are more or less essential accompaniments in sirenomelia. The only difference is that these may manifest themselves in a variety of ways. There may be horseshoeing of the kidney, complete agenesis, non-functioning kidneys, or double ureters etc. In this case there was a total bilateral agenesis of the whole urinary tract except for a small urinary bladder. Moreover, there was a complete absence of gonads.

Gastrointestinal Tract

As is the case with urogenital tract so also is it with the gastrointestinal tract. Usually there is an imperforate anus of various degrees. In this case it was noted that in addition to blind ending of colon there was a subhepatic position of the caecum and appendix, showing that the final stages in gut fixation have failed to take place.

Amniotic Fluid

A point of some interest in this case was the history of passing a large quantity of fluid. Usually such cases are attended with scanty fluid-oligo—hydramnios which is regarded, more or less, as a rule, in cases accompanied with renal aplasia. It has been suggested by various authors that the lack of amniotic fluid contributes the mechanical factor in causation of minor malformations of the limbs e.g. talipes. This lack is also blamed for causation of sirenomelia. The passage of large quantity of fluid in this case is contrary to this popular belief and, as many workers have put, it is hard to believe that this factor plays or could play any role in causation of sirenomelia.

Non-rotation of the leg is another point which is said to be always present. It cannot be confirmed here because of the absence of the fibula.

Umbilical Artery

Kampmeier (1927) stresses that a single umbilical artery was an essential feature of sirenomelia and might be an aetiological factor. In general it is regarded to be a persistent vitelline or omphalomesenteric artery connected with the aorta.

The interesting feature in our case was that there was complete agenesis of the abdominal aorta below the superior mesenteric artery. It seemed to have been replaced by this so-called umbilical artery. All the branches of abdominal aorta were forthcoming from this vessel, which was placed at a more ventral aspect. There was, similarly, complete absence of internal iliac on both sides.

Maternal age and parity. It is supposed that most of the cases are primiparous and the maternal age ranges from 19 to 41. The ratio of primiparous and multiparous incidence is 4:1 approximately, as stated by Kampmeier (27). Our case was a 4th gravida and 25 years of age.

Parental Subfertility

This has been blamed as a cause of malformations. It could not be ascertained, however, whether the father was oligospermic or otherwise. Moreover, there is no definite evidence that paternal subfertility is one of the causes.

Drugs

Usually thalidomide group of drugs is blamed for production of limb anomalies. In our case there was no history of such drug administration. There was, however, a history, of fever in early period of gestation but we wonder whether it can be regarded as a cause.

In view of complete absence of one of the limbs and the distal stunting of the available limb our case could be termed as *'ECTROMELIA'* combined with *'HEMIMELIA'*.

Summary

(1) A case of sirenomelus is described where there was complete absence of left lower limb and distal stunting of the right.

(2) The mesenteric arteries arose high up in the abdomen from a vessel which was more ventrally placed and continued towards the umbilicus. All the subsequent branches of the abdominal aorta arose from this vessel.

(3 There was complete absence of kidneys, ureters and gonads. The bladder was nearly solid with only a capillary lumen.

(4) There was a urethra traversing a rudimentary penis which did not communicate with the exterior.

(5) The caecum and appendix were subhepatic in position and the rectum and anal canal were completely absent with an imperforate anus.

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