TWIN PREGNANCY WITH OMPHALOCELE AND ACARDIAC MONSTER

(Case Report)

by

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The following case is reported for the several interesting features it presents. The first twin had a rupture of omphalocele with prolapse of the foetal intestines and liver. The second was an acardiac monster. Acardiac monsters have been reported and discussed because of their rarity and embryological mystery.

Case Report

Mrs. G. S., aged 28 years, was seen at the antenatal clinic of Dr. Balabhai Nanavati Hospital on the 26th June 1964. She had 3 full-term normal deliveries, the last delivery being 2 years ago. The birth weights of the children were between 7 and 8 pounds. There was no history of diabetes or multiple pregnancy in the family. Her last menstrual period was on 5th January 1964 and the expected date of delivery was calculated as 12th October 1964.

On general examination she was fairly well built and well nourished. The cardiovascular and respiratory systems were normal. Her blood pressure was 130/90 and her haemoglobin was 10 gms. Urine did not contain albumin or sugar.

On abdominal examination the uterus was about 26 weeks' size, which corresponded with the period of amenorrhoea. The foetal heart sounds were well heard. The patient was advised to take calcium, vitamins and iron throughout the pregnancy and she was asked to report after a fortnight for check up.

She presented herself on 16th July 1964, three weeks after the first visit. The uterus had definitely grown much more than it should have. The height of uterus corresponded to 32 weeks. The foetal heart sounds could be heard in the right lower quadrant and were 140 per min. Doubt was raised about multiple pregnancy or a large baby. The woman on further questioning replied that she was habituated to having large babies and hence x-ray at this trimester was postponed. She was advised to have a glucose tolerance test done, but she failed to go to the laboratory for the test.

She came to the hospital after a month, on the 18th August 1964, with labour pains. The uterus was about 36 weeks' size. The foetal head was felt in the right iliac fossa. It was definitely small for the size of uterus. The foetal heart sounds were heard in the right quadrant, and were regular in rhythm, the rate being 140 per minute. One foetus was lying transversely. On the top of this foetus a firm mass was felt which occupied the major portion of the uterine cavity. There was no evidence of distinct foetal parts. A diagnosis of twin pregnancy was made clinically. Second foetal heart sounds were not heard. An internal examination revealed the cervix to be fully dilated with bulging bag, which ruptured accidentally, with drainage of liquor. Cord-like structure prolapsed which the medical officer on duty thought to be the umbilical cord. The foetal heart stopped at this stage. The consultant was informed immediately. By careful examination a diagnosis of transverse presentation was made with added complication of prolapse of the foetal in-

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testines and liver. An intra-partum skiagram (Fig. 1 a.) confirmed twin pregnancy with the first foetus lying transversely. The second twin showed a large head rudimentary ribs, vertebral column and 2 long bones (Fig. 1. b.).

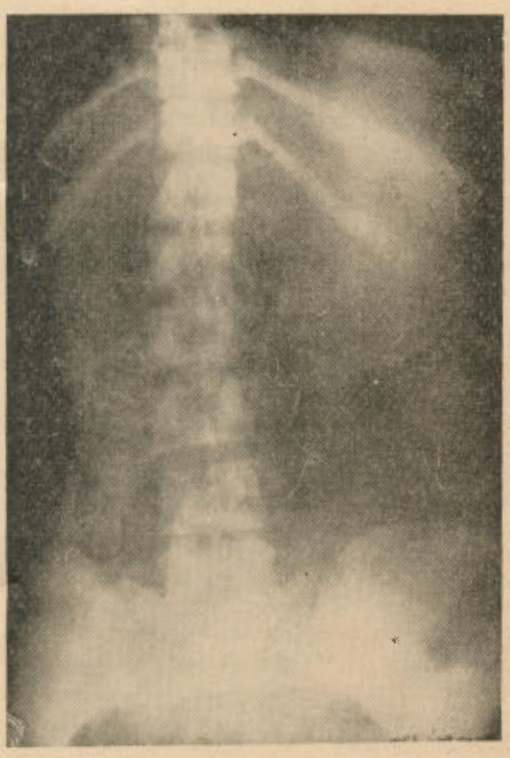

Fig. 1 (a)
X-ray.

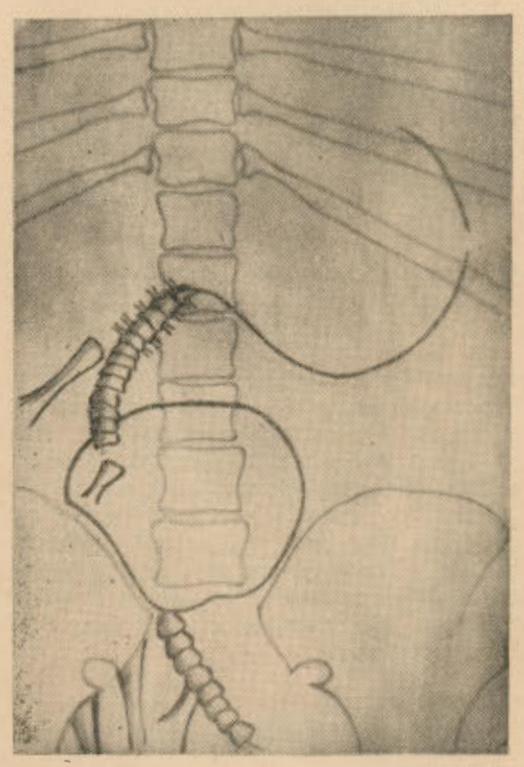

Fig. 1 (b)
Diagramatic representation.
The first child is lying transversely. The second twin shows a large head, rudimentary vertebral column, ribs and two long bones.

Under general anesthesia, an internal podalic version was done and a dead foetus was easily extracted. From the amniotic sac covering the foetal liver there were vessels about 8 cm. long simulating the cord and these ended in the edge of the placenta which was felt dipping into the lower uterine segment.

The hand was introduced into the uterus to deliver the second twin. The second bag of waters was absent. It was found that the foetus appeared to be a square mass with two small projections, occupying the upper uterine segment which was stretched. On pulling at the projections, the foetus could not be brought into the pelvic brim. Embryotomy was thought to be dangerous, since only a solid mass with no polarity was felt and it was held high up at the fundus. The true nature and the extent of the monstrosity being uncertain, the abdominal route of delivery was chosen. A caesarean section was done and the monster with exomphalos was extracted. It had a short umbilical cord measuring 4 x 0.5 cm. arising from the amnion of the exomphalos. There was a large placenta. The patient was sterilised as she did not desire more children. She made an uneventful recovery.

Description of the First Foetus: (Fig. 2). A male weighing 1250 gms. presented the following features:—The skin was normal and had no sign of maceration. The spine

Fig. 2
Intestines and liver outside the abdominal cavity with hyperextension of the spine. This is as a result of abnormal formation of the body stalk.

was scoliotic and completely extended, with hyperextension at the hips, flexion of the knees and hyperextension of the feet. The head was normal but extended. Greater portion of the anterior abdominal wall was absent. The amnion was torn leaving exposed the liver, the small and large intestines. The cord was represented by the vessels leading from the amnion covering the liver. No other abnormality was found in this foetus.

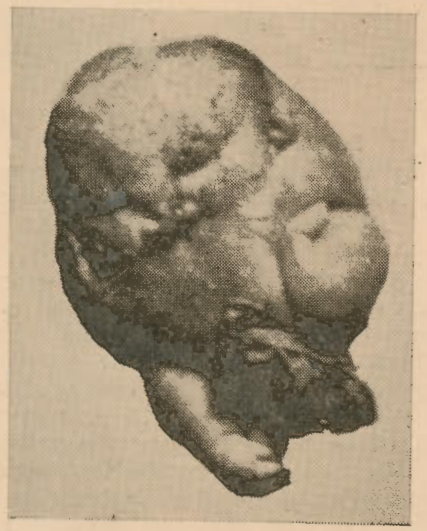

Fig. 3
Monster Acardius Parasiticus.

Monster: (Fig. 3.). There was a large firm pink mass which weighed 2270 gms. and appeared like a foetus that was arrested in its development. There was no evidence of maceration. Head to rump measurement was 21 cm. and the largest transverse diameter was 20 cm. The skin and the soft tissues were saturated with serous fluid having a spongy texture. The head was large, circumference being 40 cm. globular in shape and flattened anteroposteriorely. There was no hair except for a few at the back of the head. The facial features were poorly developed. The eyes were represented by ridges, the nostrils were represented by two bulges with a cleft below them. Two projections represented the ears. The skull bones were not united and at places only the thickened dura was present. The anterior portion of the trunk showed an irregular nodular surface. There were three projections representing the extremities. The right bud which was 6 cm. long resembled the upper limb and it had a long bone. The left bud was very small. The lower bud was 3 cm. in length and had a long bone. The spine and the ribs were present but poorly developed (Fig. 1 b.). Just above the rudimentary leg there was an omphalocele. It showed remnants of the gastro-intestinal tract in the form of tubular organs about 20 cm. in length beginning in a blind end and terminating in an imperforate anus. This appeared to be small intestine. On tracing the umbilical cord stump, there was a single artery and a single vein. It was traced posteriorly near the vertebral column and ended into the general mass of tissue. Apart from the cavity which contained the intestines, no other body opening was found.

Placenta: The specimen consisted of a large placenta which weighed 1135 gms. It measured 2.5 cm. thick and 22 cm. in diameter. There was only one amniotic cavity. The umbilical vein and arteries of the first foetus were attached paramarginally 2 cm. from the edge of the placenta. The second umbilical cord was 4 x 0.5 cm. in length. It consisted of a single artery and a single vein and was very close to the attachment of the vessels of the first foetus. There was a large blood vessel connecting the site of

the insertion of both umbilical cords. The maternal side of the placenta was complete and normal.

Discussion

Since the report of the first case by Benedetti (1944) there have been 152 cases (153 with this one) of acardius reported in the world literature according to Napolitani and Schreiber (1960). Kappleman (1944) reviewed 63 cases of this condition up to 1944. Guttmacher (1944) records one case at the Johns Hopkins Hospital in 606 twin pregnancies. The incidence cited by Gilliam and Hendricks (1953) (using the statistics of Guttmacher) is about 1 in 34,000 deliveries. The present case is the first recorded in 9,748 deliveries in the obstetric department of Dr. Balabhai Nanavati Hospital.

The acardius is a variety of monster intimately related to monovular twins. In order that the acardius may live it must receive blood supply from the twin with a functioning heart. The acardii amorphi have been described as being of various shapes, as egg shaped, reniform, globose, triangular, oval etc., their length varying from 4.5 cm. to 40 cm. and the weight ranging from 120 gms. to 2020 gms.

The descriptive classification of acardii is based mainly on gross anatomical appearance. It is difficult to classify this monster because all the classifications exclude the presence of a skull. A monster similar to this has been described by Boronow and West (1964). Their case revealed "a skull, definite axial skeletal development and some effort at limb formation blighted though it appears". This case is also similar to the one described by Kappleman (1944).

According to Schwalbe (1906) acardius is a free parasitic double monstrosity, the chorio-angiopagus parasiticus. The definition is well chosen because it emphasizes the complete parasitic dependence of the acardius on the circulatory system of the normal twin. As suggested by others (1964) it is reasonable to accept the term "Monster acardius parasiticus" for this class of monsters.

Prenatal diagnosis is by no means easy. In all cases of twin pregnancy where atypical findings and abnormal skeletal shadows are present, the condition should be suspected. If an acardiac monster has in it any tissue which is radio-opaque it should be possible with the aid of good radiological technique to diagnose the malformation prenatally. The abdominal delivery may be justified because of the abnormal size of the foetus and embryotomy under the circumstances would have been dangerous. Rupture of the uterus (1957) and abdominal delivery (1960) have been reported.

Aetiology of Acardius and Omphalocele

In the aetiology of foetus acardius the fault does not lie in the germplasm, since a normal co-twin develops from the same ovum. In an exhaustive study of the vascular anastomoses of monochorial twin placentae Schatz (1898), concluding that the development of acardiac foetuses occurred only in the presence of a large artery to artery and vein to vein

anastomoses, hypothesized that a restriction to the venous return of the acardiac is the primary event for the reversal of circulation. This obstruction was held to be the one frequently encountered in omphalocele.

Hempel (1850) was the first to recognise the reversal of circulation. Consequently any accidental or more or less constant impediment in the venous return to the only heart of so intimately interlocked circulatory system of both twins results in critical passive hyperaemia in the acardiac twin. The passive congestion explains the marked oedema and pseudohypertrophy of the tissues of the acardiac monster. The increase in volume resulting from the 'peculiarities' of reversed blood stream of the acardiac foetus may lead to critical difficulties in the delivery.

In this case the first twin had an extensive abdominal hernia which was fatal. According to Potter (1961) it is due to profound alteration in the development, and dates from an early stage of the embryonic life. It is related to the formation of the body stalk by separation of the embryo from the wall of the blastocyst, which normally takes place during the third week. Normally a cleft appears in the mesoderm between the amniotic sac and the wall of the blastocyst and enlarges until the embryo is attached to the blastocyst only by a small mass of mesoderm that gradually elongates into the umbilical cord. In this variety of eventration the area of cleavage is incomplete, the umbilical cord is rudimentary or absent and a portion of the embryo remains in contact with the wall of the blastocyst. This is the area of the yolk sac, and as a result the region of the anterior abdominal wall is not covered by skin but remains open and adjacent to the placenta, the latter derived from the portion of the blastocyst wall to which the body stalk is attached. The abdominal viscera lie outside the abdominal cavity in a space made up on one side of placenta and on the other of amnion. The umbilical vein and arteries usually course through the amniotic portion of the sac and are usually grouped together in a mass simulating a cord.

The abdominal portion of the foetus is attached directly to the placenta, and consequently to the uterine wall, and the spine is severely kyphotic and scoliotic owing to forward protrusion and fixation of the abdomen. The limbs are abnormal as a result of their cramped position.

The systematic and prospective study such as that by Walker (1957) clarifies the point that the defects are not due to genetic factors. The doubled incidence of malformations in twins, the variety of malformations in these twins, as well as the occurrence of various unilateral anomalies in identical twins suggest that the malformations are related to the twinning process itself rather than to the genetic factor.

Summary

1. A case of acardius monster is presented with intranatal x-ray photographs and description.

2. The occurrence of the monster is rare. A simple, single name of monster acardius parasiticus should be given to this entire group.

3. If the monster is large, dystocia

is an important clinical consideration.

4. Aetiological theories of the monster and omphalocele are reviewed. The double incidence of malformations in twins, the variety of malformations in these twins, as well as the occurrence of various unilateral anomalies in identical twins suggest that the malformations are related to the twinning process itself rather than the genetic factor.

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