ACEPHALUS ACARDIUS CERVICO-THORACHOPHAGUS CONJOINED TWIN — A CASE REPORT

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SUMMARY

A rare case of acephalus acardius cervico-thoracophagus conjoined twin associated with polyhydramnios and delivered by Caesarean section is reported. The diagnostic and management problems and a brief review of the pathophysiology of conjoined twins is outlined.

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INTRODUCTION

The pathophysiology of the twinning process not only interests the obstetrician and paediatrician but also the radiologist and geneticist. Conjoined twins are very uncommon, and more so is one with an acephalus[1], still rarer is the acardiac twin. Radiology and ultrasonography play a vital role in the evaluation of conjoined twins with regard to the route of delivery[2] and feasibility of corrective surgery.

CASE REPORT

A 26 year old Indian Tamil was referred from a peripheral hospital for evaluation of her third pregnancy because of excessive uterine enlargement. The period of grstation was 36 weeks.

Examination of the abdomen revealed moderate polybydramnios wit hevidence of multiple fetal parts. Further examination to exclude multiple pregnancy was not easy because of the tense uterus due to polyhydramnios.

Ultrasound examination showed a singel fetal head with four upper and lower limbs and a single heart. No further details could be evaluated. Examination of the abdomen revealed one head at the pelvic inlet, two almost parallel spinal columns and multiple limbs with evidence of polyhydramnios. Fusion of the spines was noticed in the ventral aspect of the upper thoracic and cervical region.

In view of the ultrasound and radiological findings, an elective Caesarean section was performed at 38 weeks gestation. The fetus at birth had a single head with fusion of the spines in the cervical and upper thoracic region. The fetal head appeared large but was normal in other respects. One pair of the upper limbs was shorter compared to the other. Both pairs of lower limbs were of equal length and size. One placenta with a single umbilical cord and only one artery noted. The baby died soon after delivery inspite of resuscitative measures. Radiography of the fetus (Fig.1) confirmed the above features.

Post mortem exanination revealed a single heart, two aortic arches, one being rudimentary. There were two oesophagae and two stomachs. The small bowel was unusually long with a communication at the mid gut level. The large bowels were separate, both ending blindly.

Two pairs of kidneys were noticed, one being under developed. The external genitalia appeared female even though internal genital organ were not clearly identifiable.

DISCUSSION

Conjoined twins are rare and to date about 300 cases have been reported. More than 75 per cent of them are of the thoracophagus type in which the twins are joined at or near the sternum[1]. This case is intersting in that it was an acephalus acardius cervico-thoracophagus conjoined twin. The cause of conjoined twinning in human is not known. Failure of splitting of the yolk sac has been incriminated[1].

Modern ultrasoninc and radiographic[2] facilities have made diagnosis of conjoined twins and other associated abnormalities easier. However with the presence of polyhydramnious , radiographic clarity may be poor and diagnosis difficult. The roentgenographic criteria of heads at the same level, unusual extension of the spines, proximity of the fetal spines and no change in the relative position of the featuses

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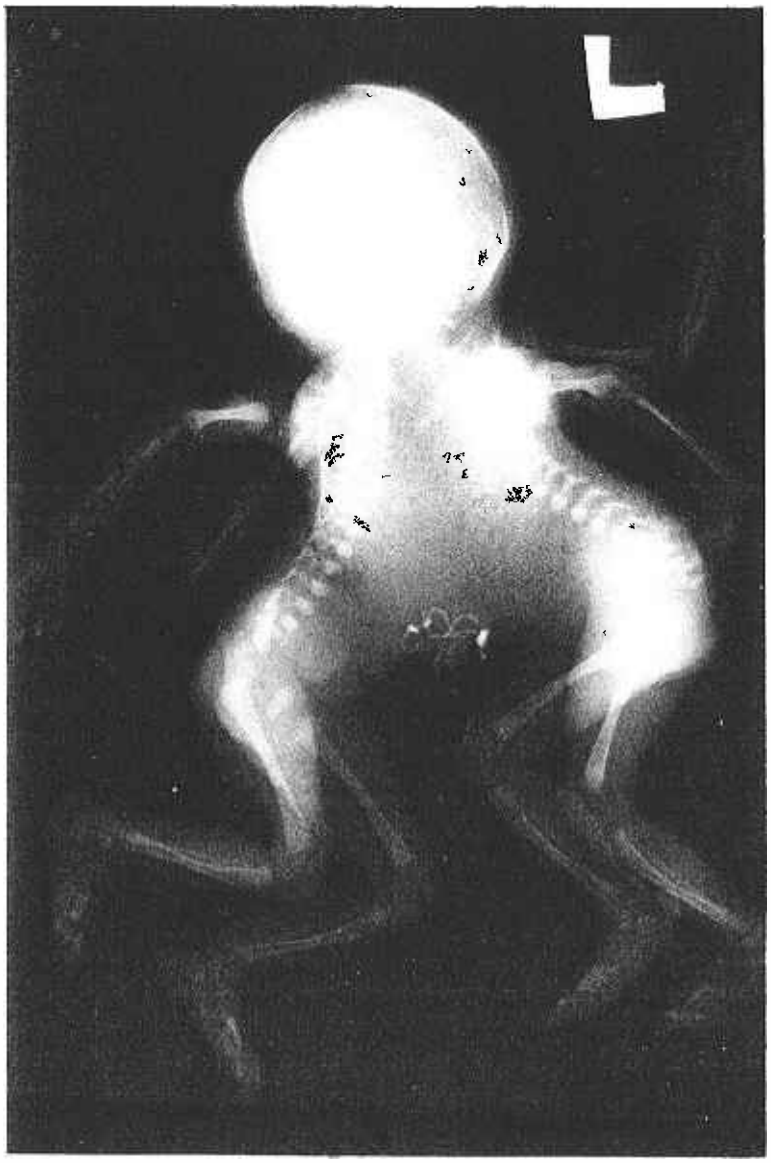
may not be seen in all cases. In our case, there was a single skull and fusion at the cervico-thoracic level. Amniography is useful not only as a confirmatory procedure, but also in demonstrating the extent of soft tissue fusion anomalies of the gastro-intestinal system and the location of the placenta. Pre-operative evaluration of conjoined twins with regards to the organ systems of the body and other anomalies is essential in order to ascertain operative viability.

Conjoined twins are invariably monozygotic and this may be associated with interplacental anastomosis from the circulation of its normal monochorionic twin. This placental circulatory aberration may influence fetal cardiac development and produce extracardiac defects[1]. In the normal fetus, the head requires a larger blood flow than the rest of the body. As a result of interplacental anastomosis, this may be reversed and the head may fail to develop. This possibly explains the mal-development of the fetal head and the single heart in our case. All the acardiacs so far studied have had a single umbilical artery which supports the haemodynamic factor[3] also evident in this case.

Fig 1: Post Mortem X-ray showing the acephalus cervico-thoracophagus conjoined twin.



REFERENCES

1. Bergsma D. Conjoined twins. In: Bergsma (ed): Birth defects original article series. New York; The National Foundation 1976; 3:1.

2. Borden S, Rider RF, Pollard JJ, Harden WH. Radiology of conjoined twins. Am J Roentgen 1974;120:424-30.

3. Noonan JA. Twins conjoined twins and cardiac defects. Am J Dis Child 1978:132:17-24.