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CASE REPORT

Acrania with placental adhesion: a case report

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ABSTRACT

Acrania is a rare developmental anomaly characterized by a partial or complete absence of the skull bones along with absence of skin and other soft tissues over the exposed brain tissue. Such cases may be detected during antenatal ultrasonography or in aborted fetuses. It is a lethal anomaly and post natal survival is very rare. Placental adhesions with the exposed neural tissue in anencephalic babies had been reported earlier. A rare case of acrania born with placental adhesion surviving for 35 days after normal vaginal delivery is reported here.

Keywords: *Acrania, anencephaly, placental adhesion*

INTRODUCTION

Acrania is a rare developmental anomaly characterized by a partial or complete absence of calvarium, which may be associated with complete, but abnormal development of brain tissue.[1] Most of the reported cases were detected during antenatal ultrasonography or in aborted fetuses.[2] As this is a lethal anomaly, very few cases surviving for a brief period after delivery had been reported. Unlike anencephaly, in acrania there is almost complete development of the brain tissue, which is directly exposed to the exterior. Placental adhesions with the exposed neural tissue in anencephalic babies had been reported.[3,4] A rare case of acrania born with placental adhesion surviving for 35 days after normal vaginal delivery is reported here.

CASE REPORT

A female baby with multiple congenital anomalies was born by normal vaginal delivery in hospital. The mother's age was 20 years, and she had no antenatal ultrasonography. The birth weight of the baby was 2.2 Kg. The baby's head was attached to the placenta, which was delivered along with the baby (Figure 1a). The baby also had cleft lip and palate, abnormal development of the right eye and syndactyly in the right hand. The spine was normal. The genitalia and anal openings were normal. There was no obvious cardiac anomaly on clinical examination; however echocardiography was not done as the facility for the same was not available.

Figure 1a Showing baby's head attached to the placenta

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The baby was immediately taken up for surgery as it was observed that because of establishment of vascular connections between the placental vessels and the meningeal vessels, there was continuous blood loss through the placenta. After the vascular connections were ligated and divided, it was observed that the brain was exposed and protruding through a large deficiency in the skull and was devoid of any covering (**Figure 1b**). Since it was not possible to raise a skin flap to cover the very large defect in the skull, amniotic membrane was taken for covering the exposed brain tissue as a biological dressing, which was then covered with wet dressing. The baby was kept on antibiotics and intravenous nutrition. However, from the second day onwards the baby accepted breast feeding. The dressing was opened on the fifth day and it was found that the amniotic membrane has provided a temporary, but good biological cover for the exposed brain (**Figure 1c**).

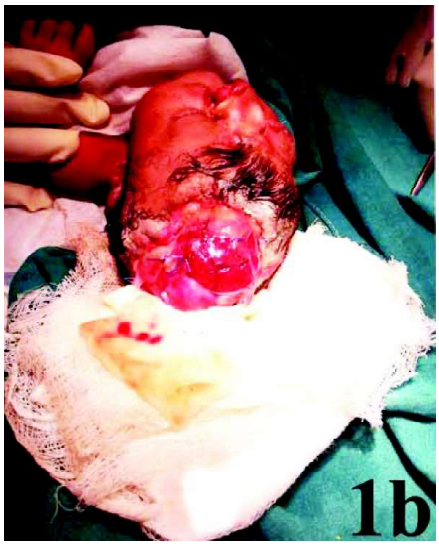

Figure 1b Exposed brain tissue after separation of the placenta

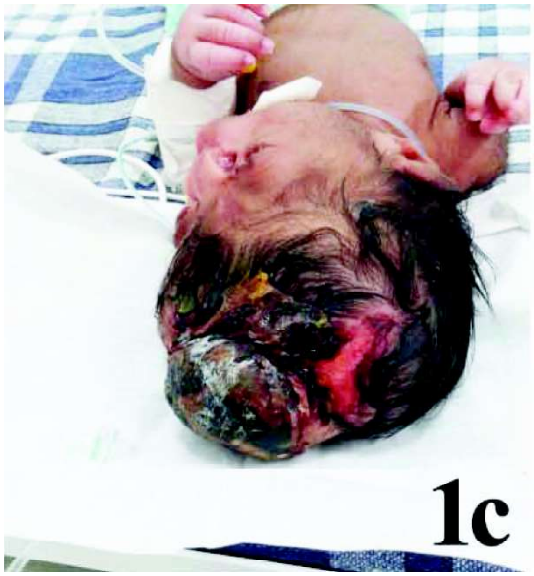

Figure 1c Amniotic membrane providing a temporary cover for the exposed brain

CT scan with 3D imaging of the head done in the post operative period revealed acrania (**Figure 2a**). It also showed significant development of the brain with gross hydrocephalus with most of the brain tissue not covered by skull or soft tissue (**Figure 2b**), and multiple facial and orbital deformities (**Figure 2c**). The baby survived for 35 days and then expired due to sepsis.

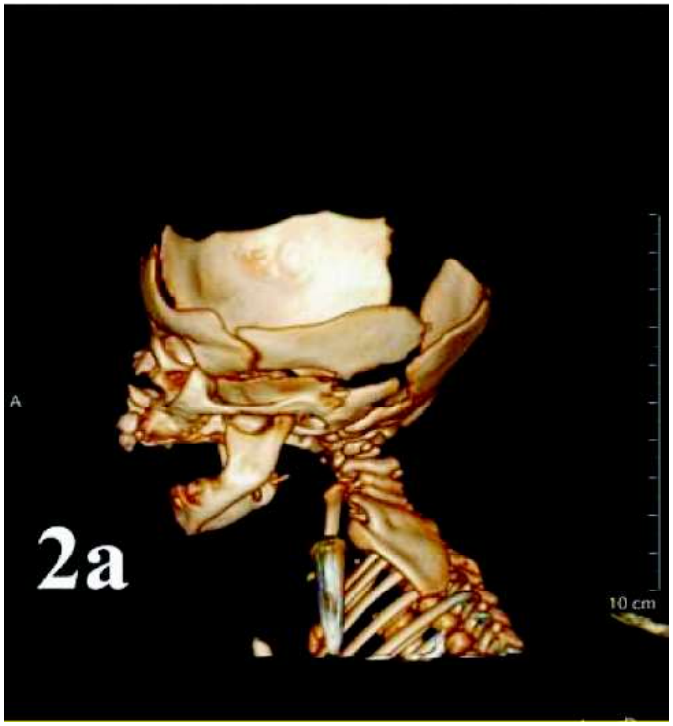

Figure 2a CT scan with 3D imaging showing acrania

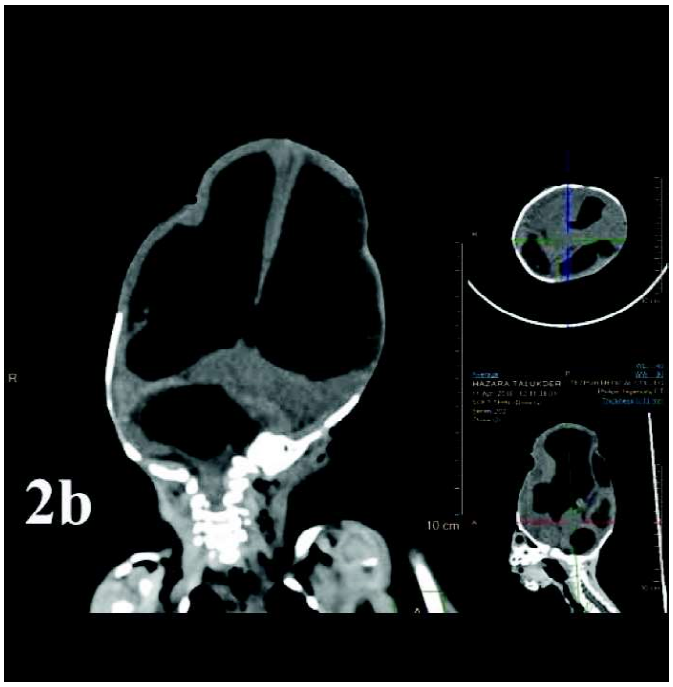

Figure 2b CT scan showing development of the brain with gross hydrocephalus

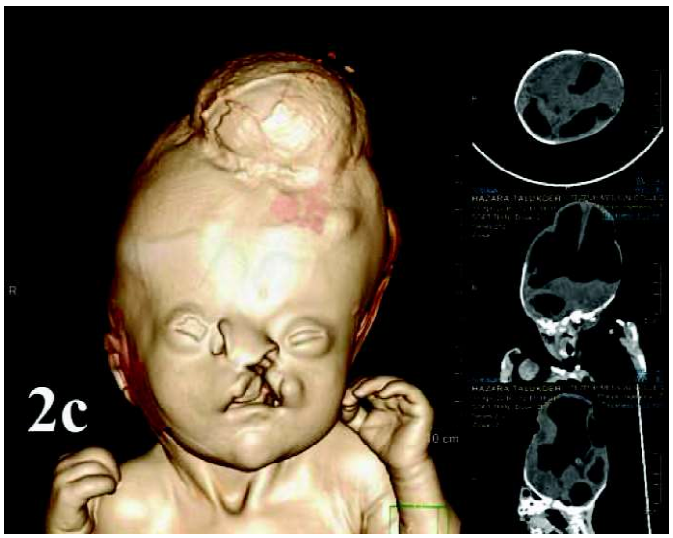

Figure 2c CT scan with 3D imaging showing cranial, facial and orbital deformities

DISCUSSION

Acrania can be diagnosed early in pregnancy during antenatal ultrasonography. Absence of a large part of the skull is diagnostic. Unlike anencephaly there is almost complete development of the brain tissue, which is directly exposed to the exterior. The exposed neural tissue during the early fetal period leads to adhesion with the placental tissue, the later being very vascular. As a result of this event taking place early in the fetal period, there may be establishment of vascular connections between the placental vessels and the meningeal vessels, leading to loss of blood after delivery through the placental vessels. This was observed in our case, which prompted us to take up the newborn baby for emergency surgery.

Anencephaly is a term which is defined as congenital absence of a major portion of the brain, skull, and scalp. The Centers for Disease Control estimates the incidence of anencephaly to be 0.3 per 1000 births.[3] Anencephaly with placental attachment had been reported earlier but with survival for a very brief period.[4,5] The exact incidence of acrania with placental attachment is not known, but must be extremely rare. The difference between anencephaly and acrania is merely the degree to which the brain tissue has developed. This case had almost fully developed brain tissue protruding through the skull defect, and hence may not qualify for the diagnosis of anencephaly. But the author feels that such differentiation of terminology may be only of academic interest.

Our case had multiple other defects like facial cleft and limb deformity. The cases reported earlier by Bisht et al[4] and Sasidharan et al[5] were similar to our case, although reported as anencephaly. Our case had no skin cover over the brain, which explains the intra-uterine placental adhesion with the exposed brain. Moreover the brain in our case was developed, although it had abnormal structure as was seen in the CT image. CT scan with 3D imaging in the postnatal period is very helpful in assessing the deficiency of skull bone, and this establishes the diagnosis of acrania. Embryogenesis of this anomaly is not well understood. Amniotic band encircling the developing brain during the early fetal period had been cited as a probable cause of this anomaly.[6]

Prolonged survival of such cases is not reported in literature. Most cases are aborted during early pregnancy or are stillborn. Separation of the placenta from the cerebral surface after careful ligation of the connecting vessels followed by amniotic membrane coverage of the exposed brain tissue in this case led to survival of the baby for 35 days.

CONCLUSION

Acrania with placental adhesion is a rare anomaly. Survival of such a case with multiple anomalies of face and limb for 35 days is reported. This is probably the longest reported survival of a case with such anomalies.

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Conflicts of interest: There are no conflicts of interest.

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Consent: Taken from parent for the publication of photos and other informations of the baby for academic use.

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