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Antenatal Diagnosis of Acrania

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Sonography with its capability to diagnoses lethal fetal anomalies at an early antenatal age, has radically changed the management policies in pregnancy with fetal abnormality. A case of acrania diagnosed by ultrasound at thirteenth week of gestation, confirmed subsequently by follow up scan and the delivered fetus is reported.

Case Report

A 23-year-old seventh gravida (consanguineous marriage) was referred for sonographic examination at thirteenth week of gestation. She gave a history of six consecutive abortions with no detectable cause; all occurring in the late third trimester except for one which aborted in the fifth month and was a case of anencephaly.

Sonogram revealed a near normal sac volume and the fetus was active. The crown-rump length was less (48 mm) due to the small cephalic region representing the brain. Neither the midline structures nor the ventricles were visualized at this time, probably because of the distortion produced by the absence of the calvaria. The usually echoic skull contour was absent (*Fig. 1*).

Though the diagnosis of acrania was made, she was reassessed at 16 weeks of gestation for confirmation. At this time (*Fig. 2*), the brain volume was corresponding to the gestational age but without skull bones. There was a thin membrane over it. The bones forming the base of skull and the face were visualized and the other definable bones were also within the normal limits. No associated significant gross abnormality was detected on repeated attempts.

Therapeutic abortion was performed which confirmed the diagnosis of acrania

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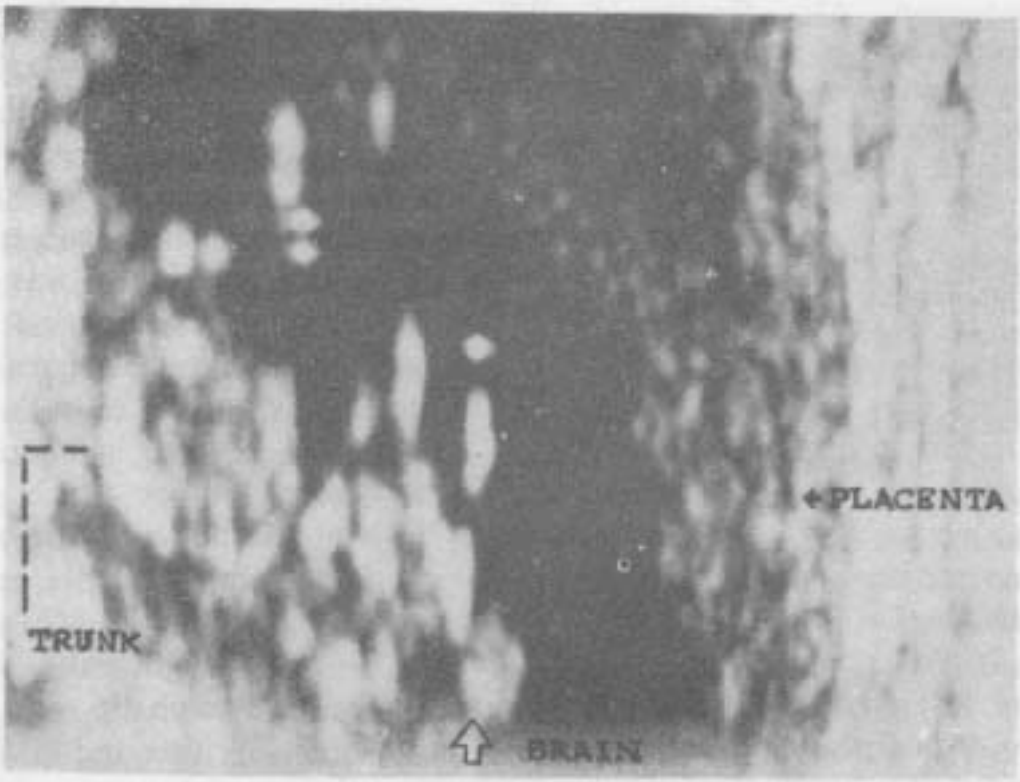

Fig. 1. Uitrasonogram of the fetus at 13 weeks of gestation showing the absence of calvaria.

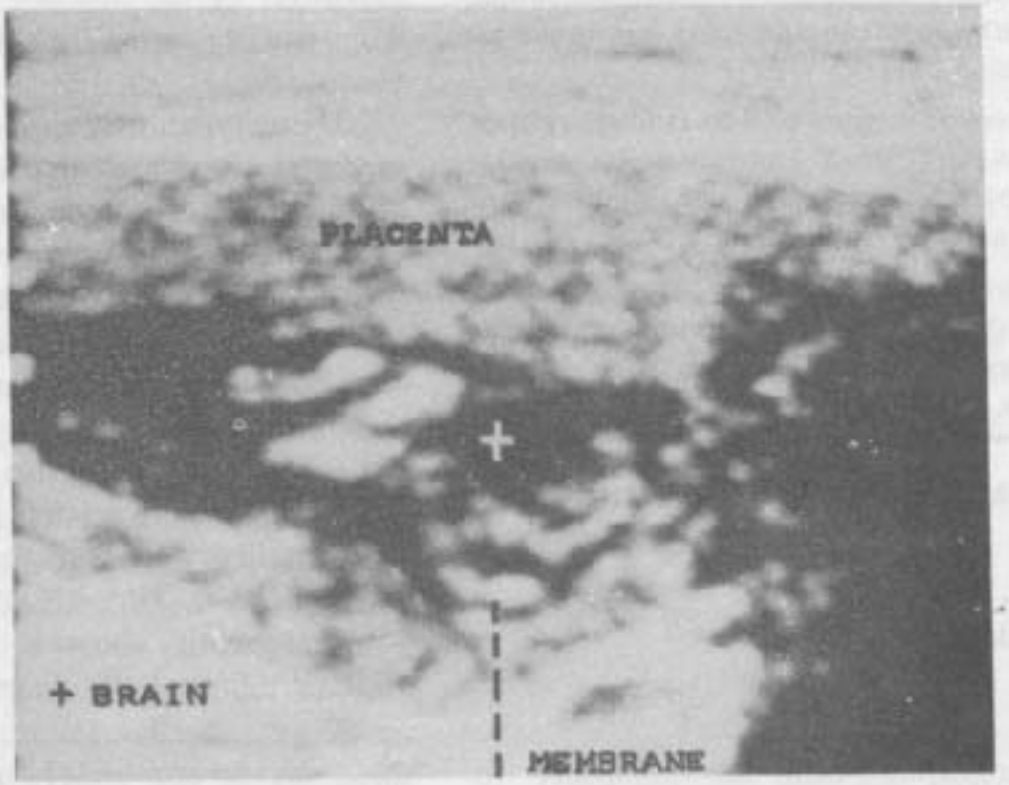

Fig. 2. The appearance of fetal head at 16 weeks when the brain was seen covered by a thin membrane instead of the skull bones.

(*Fig. 3*). The *X*-ray of the fetal skull showed absent calvaria and the presence of base of the skull (cartilagenous neurocranium) and the facial bones (viscerocranium).

Fig. 3. Post abortal appearance of the fetus confirming the diagnosis of acrania.

Discussion

Regarding the antenatal diagnosis of acrania, only few cases are reported in the literature(1). The first sonographic demonstration was at 18 weeks of gestation by Mannes *et al.* in 1982(2).

Embryologically, this condition results from the failure of mesenchymal migration under the ectoderm which initiates the ossification of the membranous neurocranium(3) in the fifth week of intrauterine life. As a result of this the future vault of skull is not formed and the scalp muscles and the duramater are represented by a thin membrane. The sonographic appearance was consistent with these features.

In the differential diagnoses, those conditions in which the skull bones are poorly defined are to be considered. Most important among them is anencephaly which can be made out by the absence of normal brain tissue. In the later weeks of pregnancy, the presence of rhomboencephalic components helps to confirm the diagnosis of anencephaly(4). Cephalocele can be identified by demonstrating a bony defect in the skull outline. Rarely, a large encephalocele may mimick acrania. But in the former case the usual supportive evidences are the presence of hydrocephaly(5), spina bifida(6) and microcephaly(7). Along with these features the maternal as well as the fetal alpha feto protein levels are usually elevated like in any neural tube defect. Conditions such as osteogenesis imperfecta and hypophosphatasia were excluded since these conditions are associated with normal intracranial anatomy and generalized bone involvement. Exencephaly is difficult to be distinguished by only echo features(2,6).

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Thymic Hyperplasia Masquerading as Cardiomegaly: MRI Validation

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On the posteroanterior chest roentgenogram the thymus appears as a bilateral smoothly outlined superior mediastinal mass merging almost imperceptibly with the cardiac silhouette(1). However, the presence of a notch on posteroanterior(PA) projection and a sharp inferior border on lateral projection helps delineate the thymus. Thymic hyperplasia, a benign condition in which there is massive enlargement of the normal thymus may simulate cardiac disease(2). Here, we report a case of thymic hyperplasia presenting with marked cardiomegaly on plain *X*-ray chest, where a suspicion of thymic enlargement was raised on two dimensional echocardiography and the diagnosis was confirmed on Magnetic Resonance Imaging (MRI). Awareness of this cause of pseudocardiomegaly will help in preventing misdiagnosis of heart disease.

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Case Report

A two-month-old child was referred to us with a diagnosis of congenital heart disease. He had a history of repeated respiratory tract infections. On clinical examination there was cardiomegaly with grade 2/6 vibratory murmur in the second left intercostal space. Plain *X*-ray chest (PA view) showed a cardiothoracic ratio of 0.7 (*Fig. 1*). Pulmonary vasculature and lung fields were normal. Two dimensional echocardiography revealed normal sized cardiac chambers. A moderate sized paracardiac mass was visualized in the subcostal four chamber view (*Fig. 2*) around the left ventricle and left atrium. This was thought to be either a pericardial lipoma or granuloma or an enlarged thymus. The possibility of a congential pericardial cyst was also entertained but considered unlikely in view of the mass appearing to be of soft tissue echogenecity and not fluid echogenecity. To characterize this mass more definitively, MRI was performed. This showed a medium signal intensity of T2 weighted image of this mass which had pointed ends and was draping itself over the heart, confirming it to be thymic hyperplasia (*Fig. 3*).