Research Paper Medical Science

A Rare Case Report -Acrania

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ABSTRACT

Acrania is a rare congenital anomaly and characterized by partial or complete absence of the calvarium with abnormal brain tissue development. Ultrasound allows early diagnosis of this anomaly. The fetus was found to have a completely formed brain, base of the skull and facial structures but lacking a cranium. Although acrania associated with anencephaly is a well recognized entity but isolated acrania is a rare anomaly.

KEYWORDS Acrania, brain, cranium

CASE REPORT

A 30 year old non-diabetic, non-alcoholic primigravida with normal obstetric examination came for fetal sonography to rule out congenital fetal anomalies.There is no family history of congenital anomalies .There was no history of intake of teratogenic drugs and other relevant past illness.During ultrasonographic scan a single live fetus of gestational age of about 34 week was seen with cephalic presentation. Femur length, abdominal circumference were matching with menstrually calculated gestational age of about 34 weeks. Liquor was excessive with closed internal os.The fetus showed normal cardiac activity (140 beats/min) with normal rhythm. The fetal movements were normally Coordinated. Ultrasound showed well formed cerebral hemispheres with lateral ventricles but without the echogenic rim of skull bone around it . Brain tissue cover with thin membrane and present outside of cranial fossa posteriorly with normal color doppler study.

Facial feature were normal with normal inter and intra orbital distance according to gestation ,nasal bone ,lips were seen normally formed . Cervical, thoracic, lumbar spine appeared normal with normal morphology of the spinal canal. No appreciated defect and no mass was seen attached or arising from the spine. Fetal limb bones were normal.Intra thoracic and intra-abdominal organs were normal. Placenta was anterior in position and was of normal thickness.

Live fetus born through normal vaginal delivery without any complication ,having well formed brain without skull bone covering [acrania (exencephaly)]. Death of neonate was occure after one hour of delivery in peadiatic nicu

DISCUSSION

Fetal acrania (exencephaly) is a congenital abnormality characterized by the complete or partial absence of skull bones surrounding the fetal brain with complete, but abnormal development of brain tissue (1).. Although acrania associated with anencephaly is a well recognized entity with an incidence of about 10:10,000 births, isolated acrania is a rare anomaly.

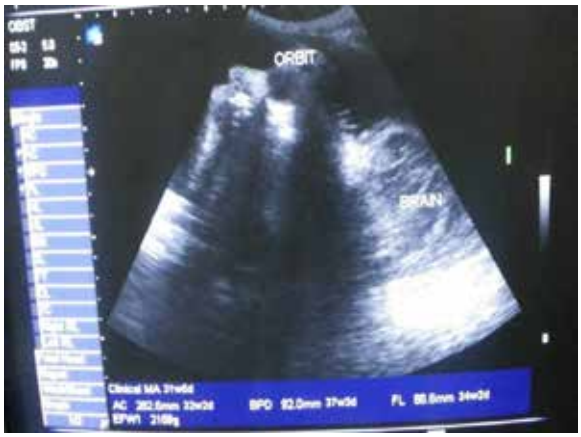
The fetal cranium is not fully calcified before 10–11 weeks; Fetal acrania can be diagnosed from 11 weeks onward (2). At 11–14 weeks gestation, the majority of cranial ossification is in the lateral aspects of the frontal bones and lower parietal bones, and no vault ossification is visible in the midline on a perfect midsagittal image. Hence, misdiagnosis may occur if only midsagittal views of the fetus are obtained. . It is important to look specifically for frontal bone ossification in the axial and coronal planes (3,4).

The differential diagnosis includes anencephaly and large cephaloceles. In anencephaly, cerebral tissue is completely absent; while in cephaloceles, the cranial vault can always be detected and a part of the brain is intracranial.

CONCLUSION

Fetal acrania is a rare and lethal congenital anomaly that warrants the identification of fetal skull and cranium around the brain that should be normally calcified. Antenatal identification allows the clinician to make appropriate and timely management decisions.

LEGENDS
Figure1



Fetal Acrania. Gray scale ultrasound of the fetal [Gestational age 34 weeks] brain (5.5 MHz probe) in sagittal plane showing no echogenic skull bone is seen around the brain .

http://vesalius.northwestern.edu/ixmlquery/hilight?parm1=chapters/FA.1.19&keyword=rib&scrollpoint=FA.1.19.07&-Figure 2:

Fetal Acrania.. live fetus [Gestational age 34 weeks] showing brain without cranium protruding posteriorly outside the base of the skull in a rounded configuration.

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