

Fetal Acrania - Prenatal Sonographic Diagnosis and Imaging Features of Aborted Fetal Brain

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ABSTRACT

We report a 35 year old female patient referred to our ultrasound department to rule out congenital anomalies. The fetus was found to have a completely formed brain, base of the skull and facial structures but lacking a cranium. The fetus was therapeutically aborted. We correlated our antenatal sonographic findings with gross pathological features and CT Scan of the fetal head. Despite an extensive search, CT features of aborted fetal brain and base of skull were not found in the literature.

CASE REPORT

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A 35 year old female was referred to our ultrasound department to rule out congenital fetal anomalies. A single alive fetus of 30 weeks gestational age was seen in cephalic presentation with longitudinal lie. Ultrasound revealed well formed fetal brain without cranium (acrania) (Fig. 1, 2). Brain convolutions, interhemispheric fissure and sulci were clearly identified. A thin membranous structure was covering the brain. Fetal brain was easily compressed on probe pressure due to lack of cranium. The brain appeared to be floating in the amniotic fluid over the base of skull (Fig.3). Increased amount of amniotic fluid was seen (Fig.4). Fetal brain showed normal vascular pattern on color Doppler scan (Fig.5). No other fetal anomaly was seen. Facial structures were normal. The fetus showed normal cardiac activity (137 beats /min) with normal rhythm. The movements of the fetal body parts were normal and well coordinated. Placenta was anterior in position and

was of normal thickness. Both orbits were symmetrical in size and shape. Fetal nose and naso-labial folds were recognized and showed symmetry. Cervical, thoracic and lumbar spine appeared normal with normal morphology of the spinal canal. No mass was seen attached or arising from the spine. Fetal long bones were normal (Fig.4).

Fetal stomach, liver and gall bladder appeared normal. No abnormal intestinal loop was seen. Fetal kidneys were normal in size, shape and contour with normal calyceal system. Urinary bladder was recognized. Skin thickness of foetus was normal. Umbilical cord appeared normal with 1 vein and 2 arteries. The patient was counseled regarding termination of the pregnancy and a therapeutic abortion was carried out.

Aborted fetal examination revealed lack of fetal cranium (Fig. 6, 7). Fetal facial features were normal. Brain was covered by a thick membrane.

X-ray of the fetus revealed acrania (Fig.8). Rest of the fetal axial and appendicular skeleton was well formed with normal mineralization of visualized bones.

CT scan of the aborted fetus was performed. Basisphenoid, basitemporal and Basifrontal bones were formed (Figure 9-13). Metopic suture was identified. However fetal cranium was absent. Few structures of fetal brain were also identified.

DISCUSSION

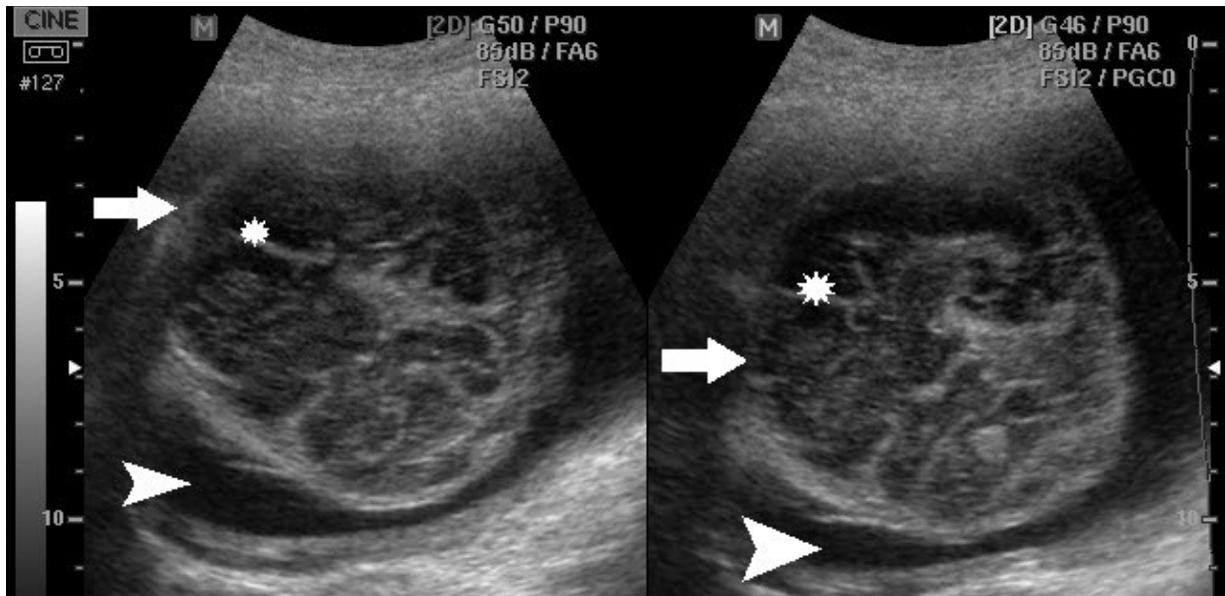
Fetal acrania (exencephaly) is a congenital abnormality characterized by the complete or partial absence of skull bones surrounding the fetal brain. Lack of mesenchymal migration in the fourth week of embryological age is a proposed mechanism (1). Acrania is a uniformly lethal entity that is characterized by a partial or complete absence of the calvarium with complete, but abnormal development of brain tissue. The fetal cranium is not fully calcified before 10-11 weeks; therefore, a first-trimester diagnosis must be made with caution (2).

Fetal acrania can be diagnosed from 11 weeks onward. 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. The absence of cranial ossification may not be noted, and the head may appear relatively normal. It is important to look specifically for frontal bone ossification in the axial and coronal planes (3,4).

3-D US may contribute to early detection of fetal acrania and provide a novel visual depiction of this defect after reconstruction (5).

FIGURES

Figure 1: Fetal Acrania. Gray scale ultrasound of the fetal brain (3.5 MHz probe) in axial plane. Gestational age 30 weeks. No skull is seen around the brain (arrows). Arrowhead=amniotic fluid around the brain. Asterisk indicates inter-hemispheric fissure.



Severe Osteogenesis imperfecta and congenital hypophosphatasia result in poor mineralization of the calvarium. In these cases sonography may demonstrate poorly defined calvaria which can be conceivably difficult to differentiate from acrania. However although thin and possibly deformed fetal calvaria in Severe Osteogenesis imperfecta is usually seen on sonography. In addition, sonographic demonstration of fractures and knowledge of family history aid in differential diagnosis (6).

TEACHING POINT

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.

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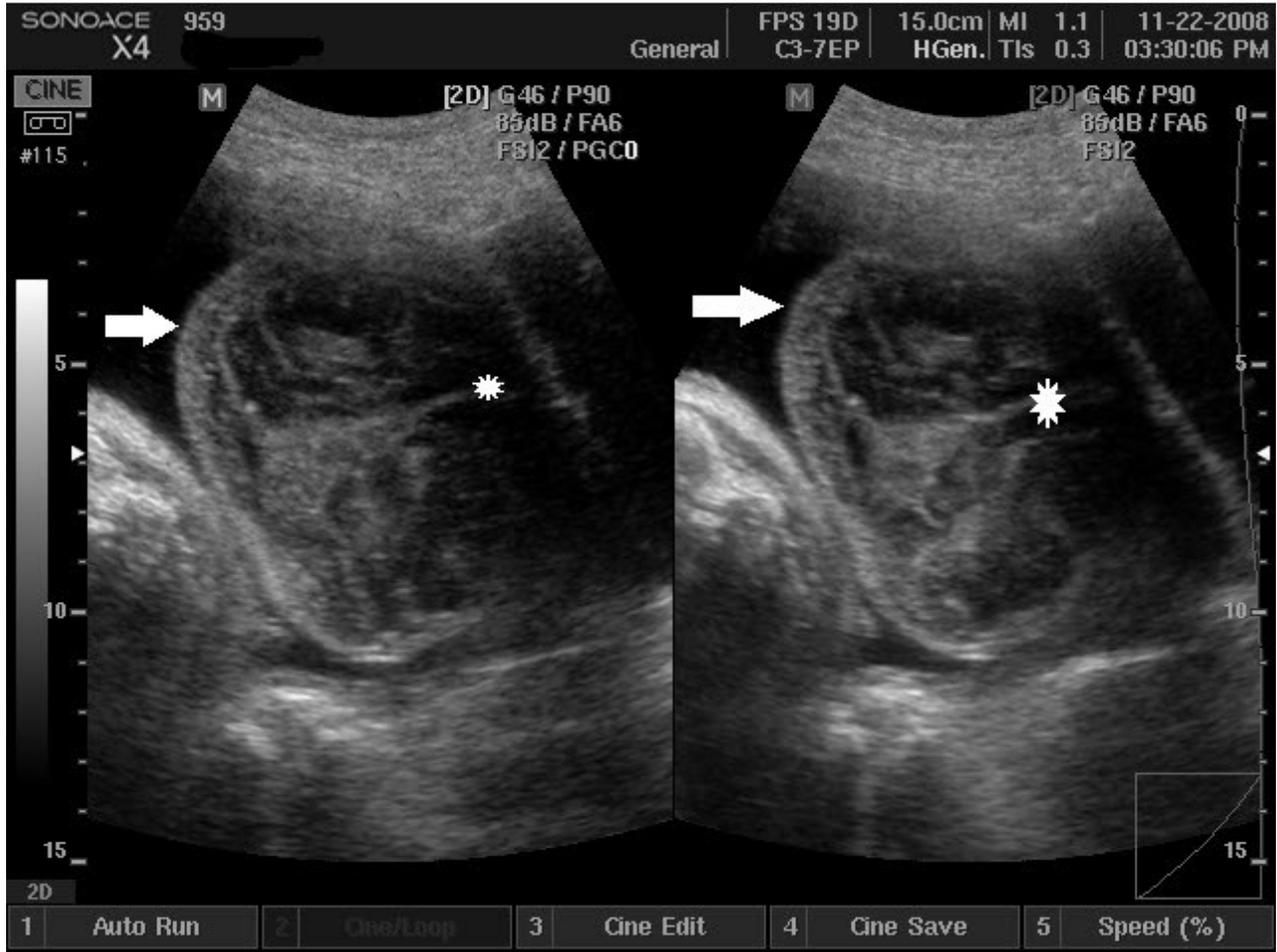


Figure 2: Fetal Acrania. Gray scale ultrasound of the fetal brain (3.5 MHz probe) in coronal plane. Gestational age 30 weeks. No skull is seen around the brain (arrows). Asterisk indicates inter-hemispheric fissure.

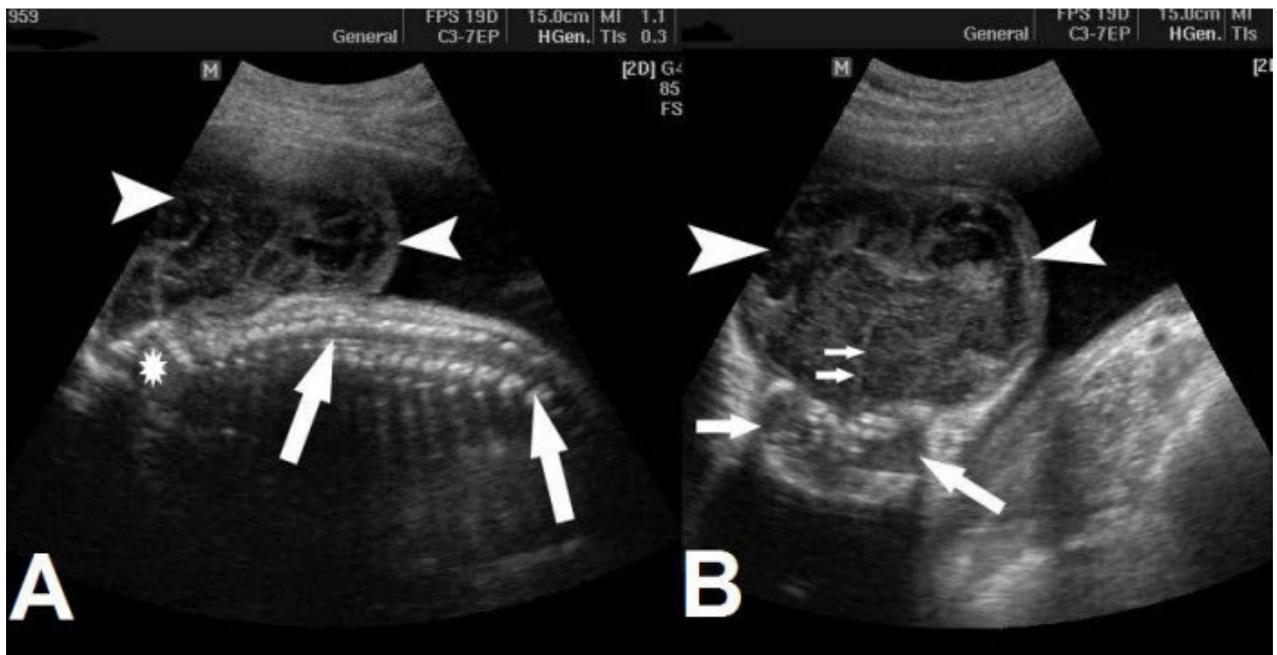


Figure 3: Fetal Acrania. (A) Gray scale ultrasound of the fetus (3.5 MHz probe) in sagittal plane. Gestational age 30 weeks. White arrows = Normal looking spine. Fetal brain is seen floating in the amniotic fluid now placed posterior to the cervical spine without covering skull. Asterisk denotes craniocervical junction. (B) Coronal gray scale ultrasound scan showing normal orbits. No skull is seen around the brain. Small Arrows = Inferior Interhemispheric Fissure. Arrows = Orbits. Arrowheads = Fetal brain lacking covering skull.

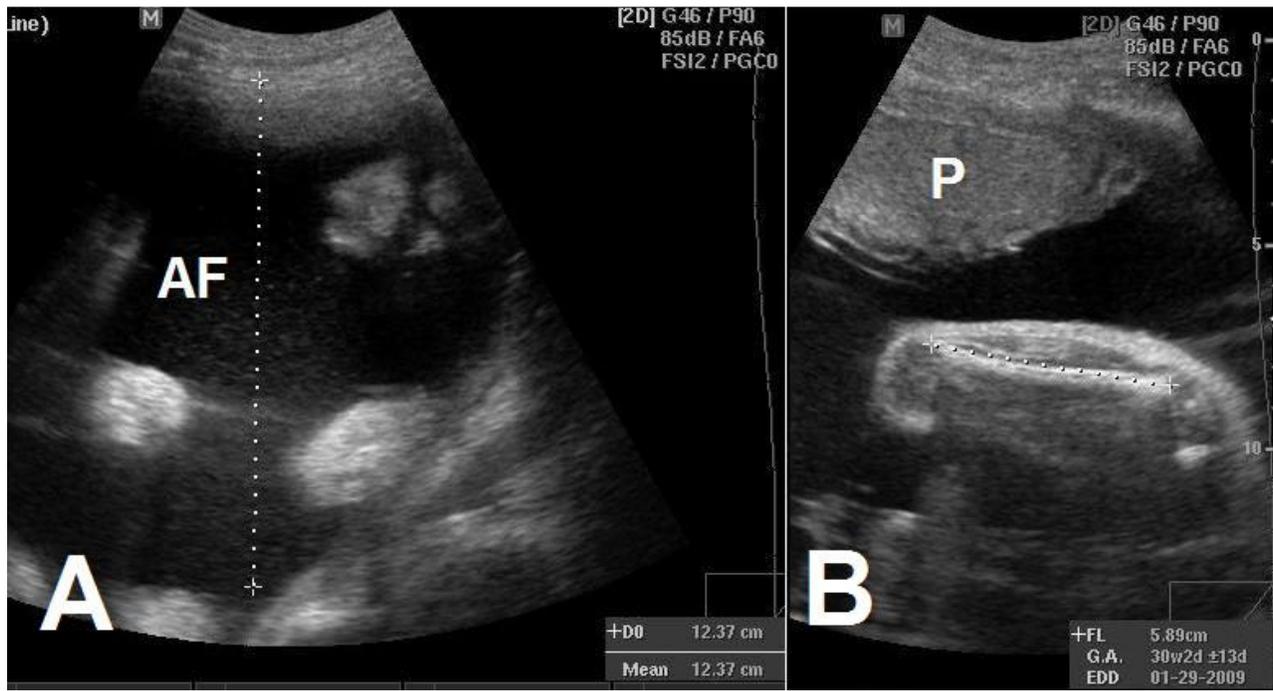


Figure 4: Fetal Acrania. Gray scale ultrasound (3.5 MHz probe). Gestational age 30 weeks. (A) Increased amount of amniotic fluid was seen .AF=Amniotic fluid. (B) Normal femur equal to gestational age of 30 weeks. P= Placenta

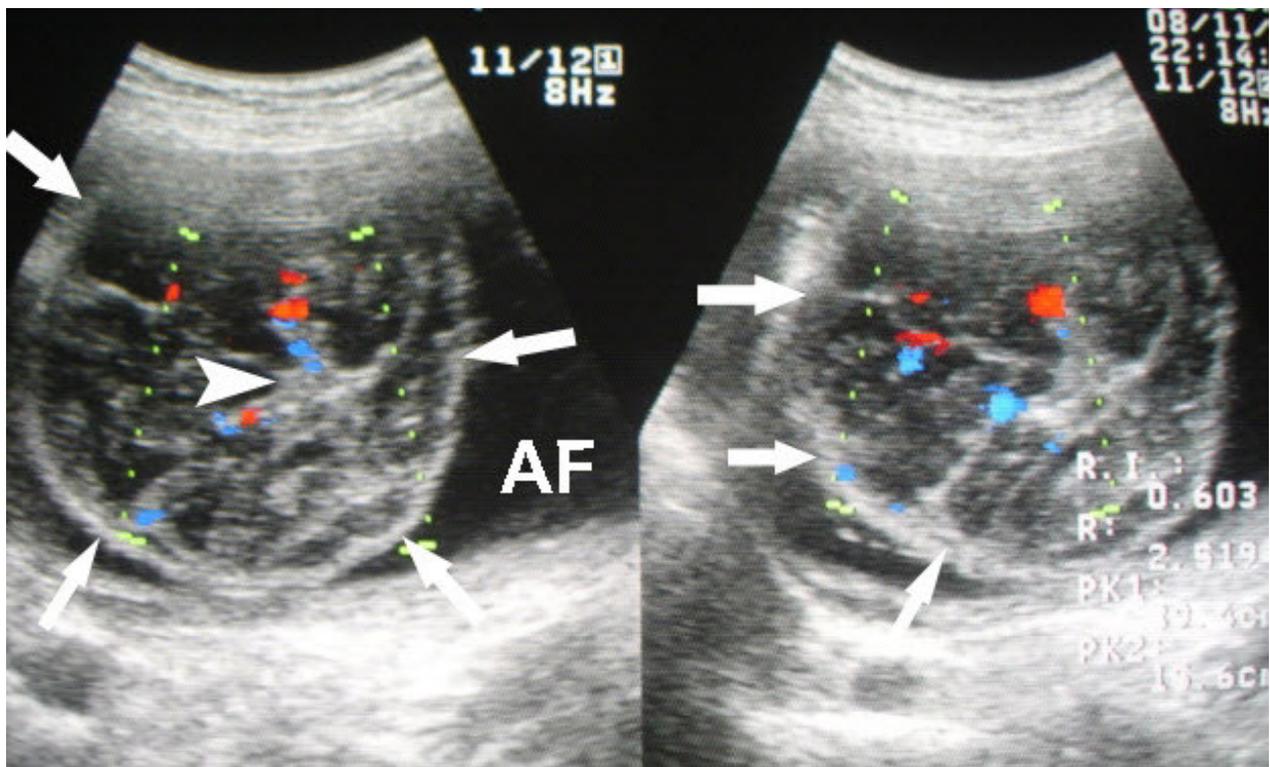


Figure 5: Fetal Acrania. Color Doppler ultrasound scan of the fetal brain in axial plane performed with 3.5 MHz probe. Gestational age 30 weeks. Intracerebral vessels are seen. Arrowhead = Star shaped suprasellar cystern, Arrows = Margins of brain without covering skull. AF= Amniotic Fluid.

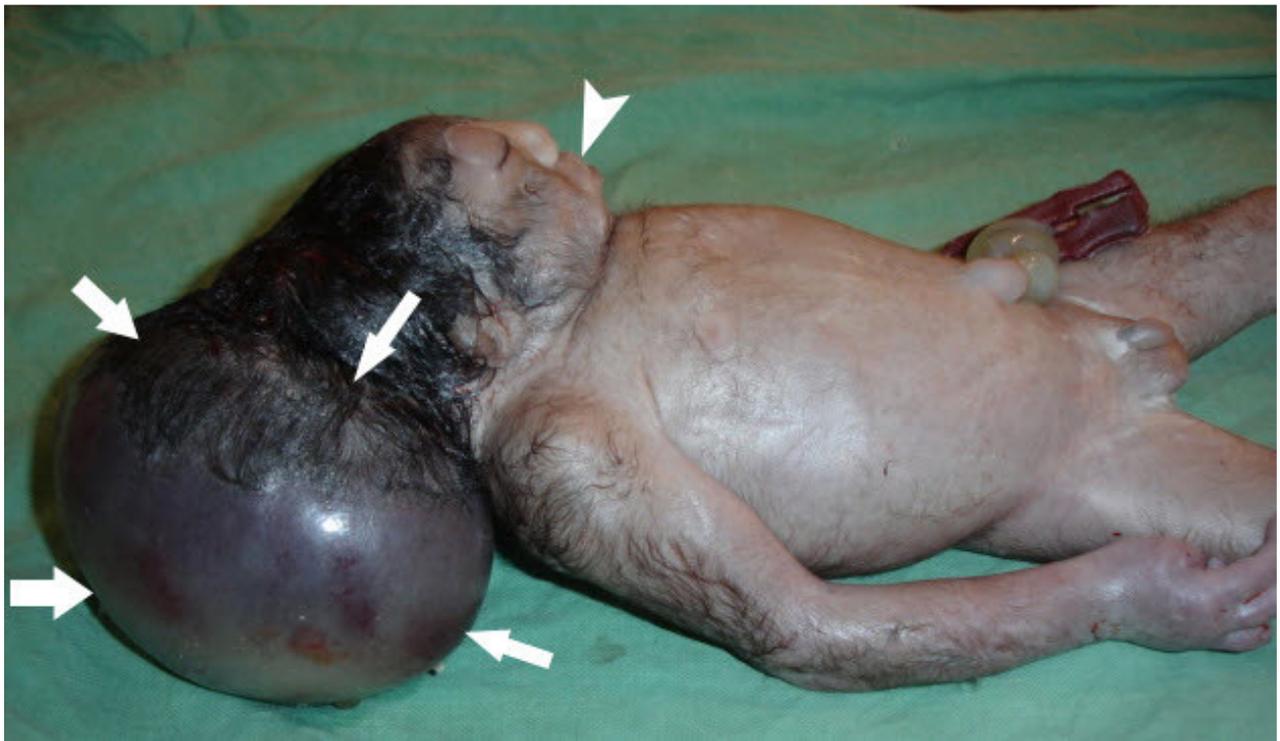


Figure 6: Fetal Acrania. Gestational age 30 weeks. Aborted fetus showing brain without cranium protruding outside the base of the skull in a rounded configuration. Arrows = fetal brain, Arrowhead = fetal face



Figure 7: Fetal Acrania. Gestational age 30 weeks. (A) Lateral view .Close up picture of aborted fetal face and brain. (B) Posterior view of the fetal brain without covering skull.

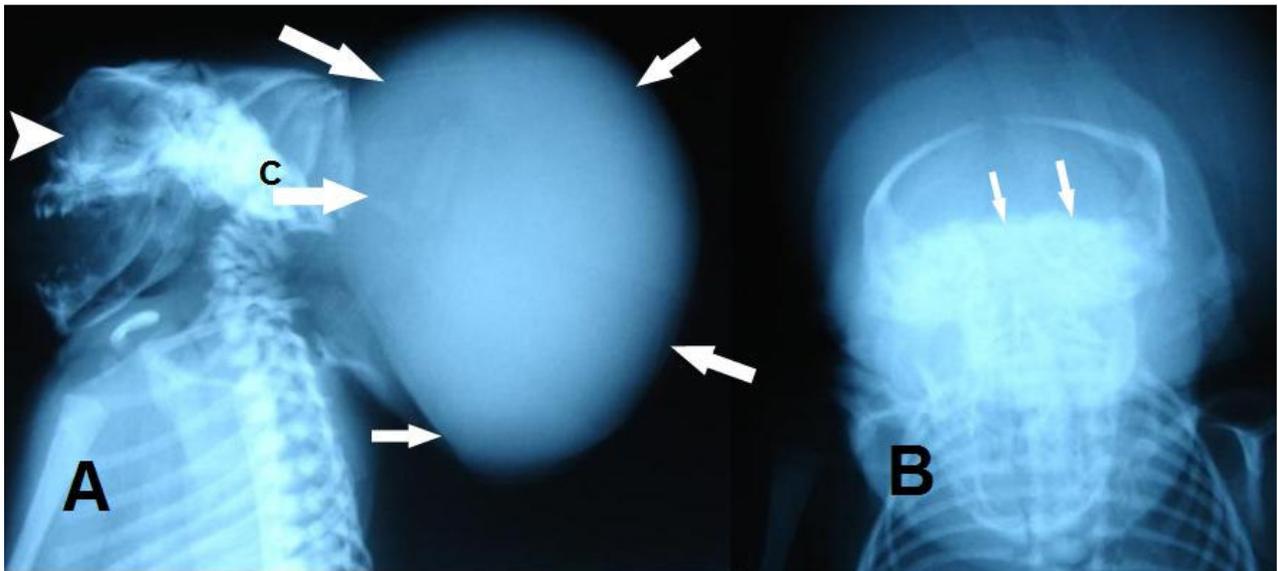


Figure 8: Fetal acrania. Gestational age 30 weeks. (A) Plain x-ray of the aborted fetus showing face (arrowhead) and brain (arrows) without any evidence of skull around the fetal brain. C=craniocervical junction. (B) Frontal projection showing only base of the skull (arrows).

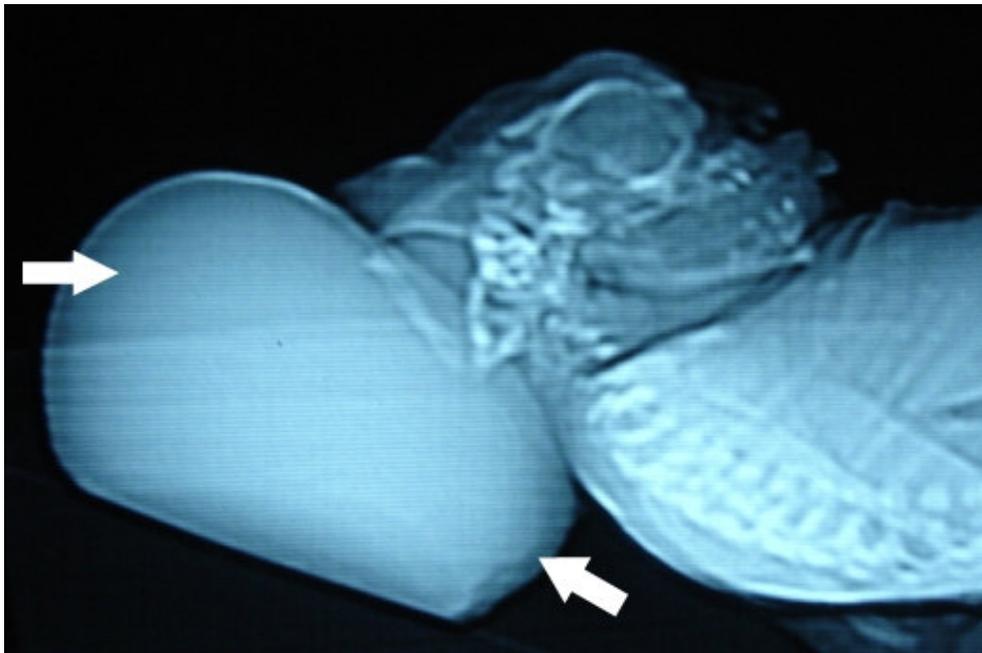


Figure 9: Scout view of the CT Scan showing acrania (white arrows).

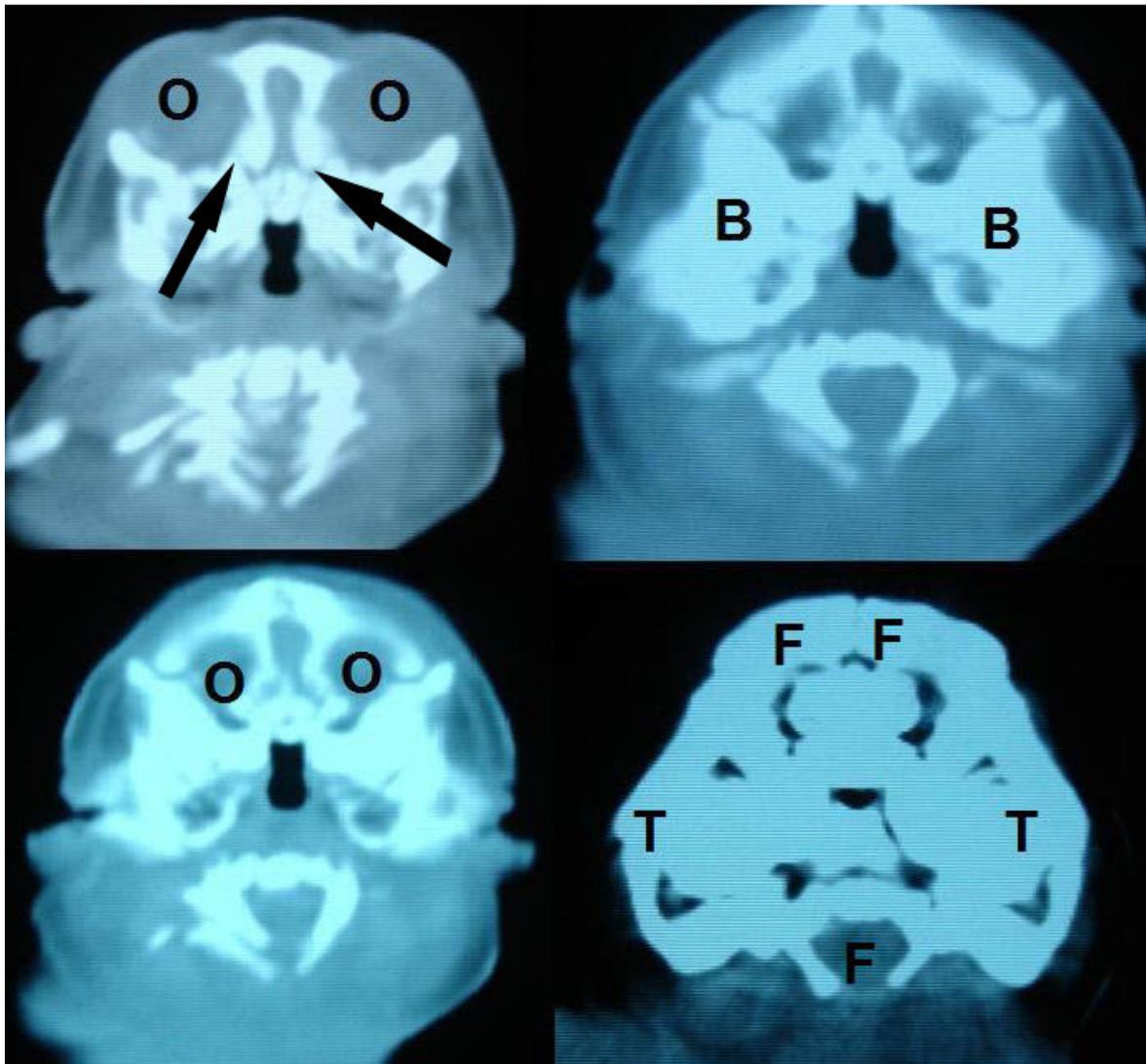


Figure 10: Fetal Acrania. Gestational age 30 weeks. CT scan of the aborted fetal face and base of the skull .All images are selected from bone window settings. O = orbits, F=frontal bones, B= basisphenoid, T= temporal bones, F= Foraman Magnum. Black arrows = Frontoethmoidal sutures

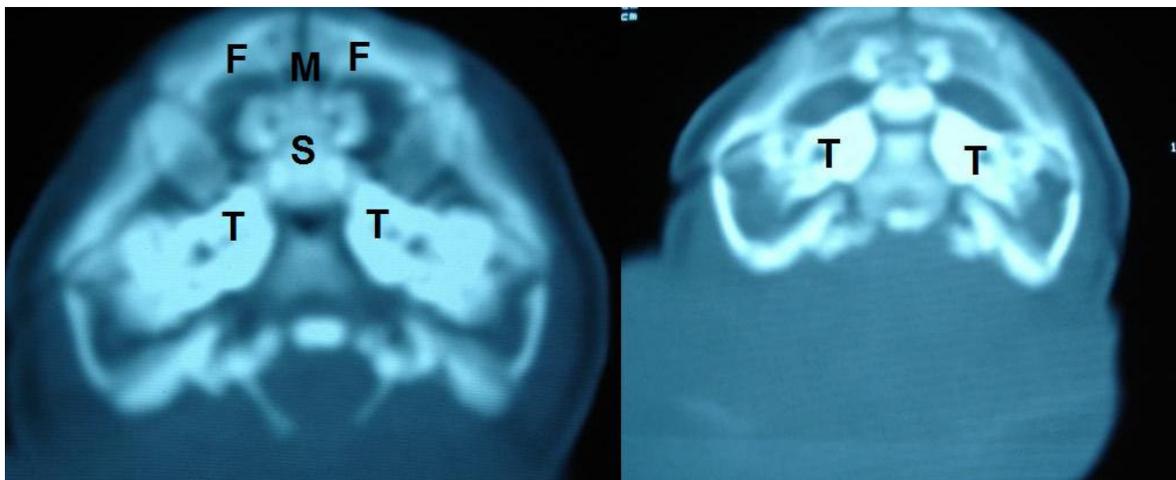


Figure 11: Fetal acrania. CT SCAN of the aborted fetal face and base of the skull .All images are selected from bone window settings. F=frontal bones, B= basisphenoid, T= temporal bones, M=Metopic suture. S= Sphenoid bone.

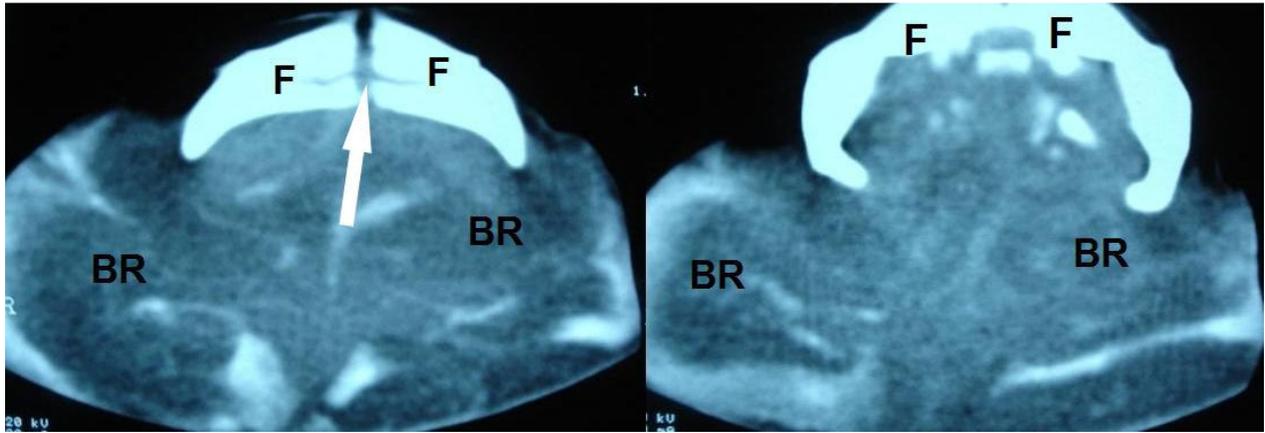


Figure 12: Fetal Acrania. CT Scan of the fetal brain. Gestational age 30 weeks. Arrow = Metopic suture, F=frontal bones, BR=Brain

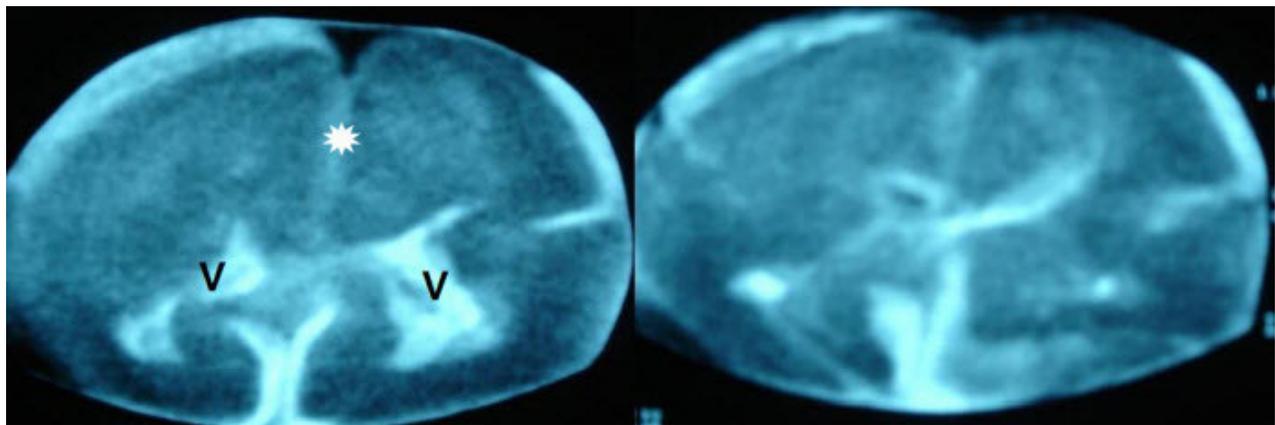


Figure 13: Fetal acrania. CT Scan of the aborted fetal brain. Gestational age 30 weeks. Fetal cranium is absent. V= ventricles filled with blood. Asterisk indicates inter-hemispheric fissure.

ABBREVIATIONS

CT SCAN = Computed Tomography Scan
MHz = Megahertz

KEYWORDS

Acrania, abortion, brain, cranium

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