FETAL AKRANİA VE EŞLİK EDEN ANOMALİLERİN ULTRASONOGRAFİK TANISI*

ULTRASOUND DIAGNOSIS OF FETAL ACRANIA AND ASSOCIATED ABNORMALITIES

Selami SUMA, Pınar PO LAT, Hanefi YILDIRIM, Ayhan AKÇALI, Suat EREN
Fevzi HARORLU, Adnan OKUR
Atatürk University, Department of Radiology (SS, PP, HY, AA, SE, AO) and Pathology (FH) Faculty of Medicine, Erzurum / TURKEY.

* 17-20 Aralık 1995 tarihinde Uludağ'da yapılan 5. Ulusal ultrasonografi kongresinde poster olarak sunulmuştur.

Özet
1991-1996 yılları arasında akrani ve birçok diğer anomalisi olduğu düşünülen yedi fetus real time ultrasound ile incelendi. Yedi olguda da akrani tespit edildi. Üç fetusta sadece akrani mevcuttu, başka bir anomali yoktu. İki olguda anensefali ve polihidroamnios tespit edildi. Üç olguda polihidroamnios mevcuttu. En erken tespit edilen olgu femur uzunluğu ve son menstrüel periyoda göre 13 haftalıktı. Beş olgu 2. trimester, 2 olgu ise 3. trimesterde tespit edildi. Kesin tanılar patologlar tarafından doğumu takiben koyuldu.

Anahtar kelimeler : *Akrania, Ultrasonografi, Konjenital anomali*

Summary
Seven fetuses who had been suspected acrania and multiple other associated anomalies were examined by real time ultrasound in the period of 1991-1996. Acrania was demonstrated in all cases. Three fetuses had only acrania and no other associated anomally. In two cases anencephaly and polyhydroamnios were associated anomalies. There was polyhydroamnios in three cases. The earliest demonstrated fetal age was 13 week according to femur length and last menstrual period. Five cases were diagnosed at second, two were at third trimester. The last diagnose was made by pathologists after delivery.

Key words : *Acrania, Ultrasonography, Congenital anomaly*

AÜTD 1997, 29:470-472 **MJAU 1997, 29:470-472**

Acrania is a rare developmental anomaly in which flat bones of the cranial vault are absent with abnormal development of brain tissue. The presence of substantial amount of brain tissue differentiates it from anencephaly. These two conditions may be seen together and with other associated anomalies. We try to present the ultrasonographic findings of 7 fetal acrania cases that were confirmed by pathological examination after delivery during 5 years period. To our knowledge there are about 8 cases that were reported in the literature (1-7) This condition is lethal. The early diagnosis of this condition will provide termination of pregnancy as soon as possible.

Subjects and Methods
Our study group included 7 fetal acrania cases that attended ultrasound (US) unit for routine obstetric examination in the period of April 1991-January 1996 and they had pathologically confirmed acrania. US examinations were done by using Toshiba SSA-270A US unit with 3.75 mHz convex probe.

Table 1. *The Results of Ultrasound İmaging In Seven Acrania Cases*

Case number	Maternal age (Year)	Number of gravida	Number of para	Fetal age (Week)	Assosiated fetal anomalies
1	28	8	7	32	Polyhydroamnios Anencephaly
2	38	3	3	15	Polyhydroamnios
3	18	1	0	13	-
4	23	1	0	14	-
5	30	2	1	17	Spina Bifida
6	27	4	2	14	-
7	18	1	0	26	Spina Bifida Anencephaly Polyhydroamnios

Figure 1. *The Fetus Is 32 Menstrual Weeks. There Is No Brain Tissue and Bone Tissue Above the Level of Orbits. Anencephally and Polyhydroamnios Are Associated Anomalies In this Case .*

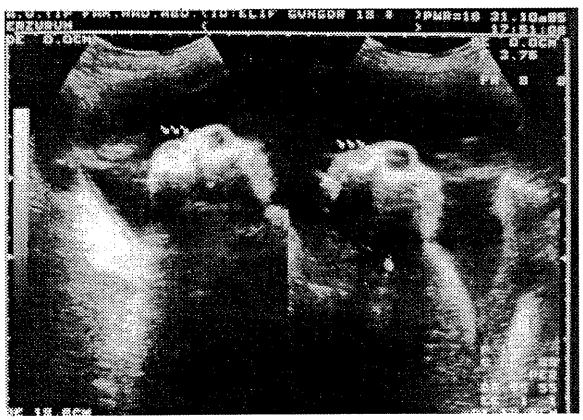
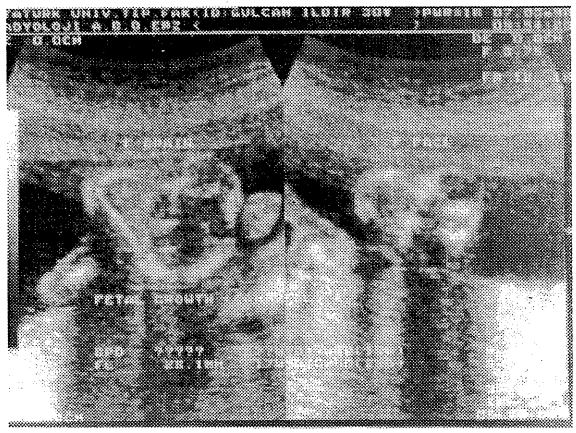


Figure 2. *Frontal Bone and Ossified Calvarium Are Not Depicted. The Fetus Is 17 Menstrual Weeks. In This Case Acrania Is Only Finding. There Is No Associated Anomally.*



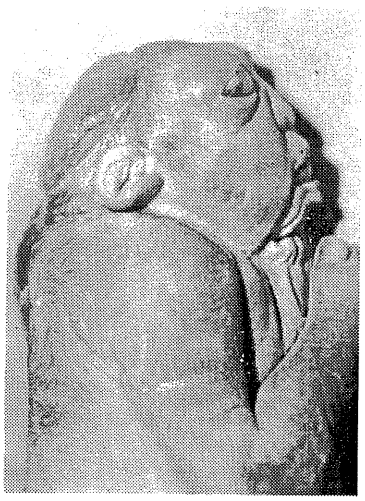
Results

Maternal age, number of gravida and number of para were listed in Table 1. The youngest maternal age was 18 year and the oldest was 38 year. Four patients were multigravidas, three were primigravidas. There was no abnormality in historical evaluation except case 1 whose two infants died immediately after birth. In detailed evaluation it was suggested that these two infants died because of anencephaly or acrania (fig 1,2). But reliable information could not be taken from this patient. Other three patients had their first gravida and in their historical evaluation there was no positive family history, other medical disease, drug uptake and radiation exposure during gravida. The youngest fetal age was 13 week according to femur length and maternal last menstrual period. In this case US evaluation was repeated two weeks later and same diagnosis was confirmed. Five cases were diagnosed at second trimester, two were at third trimester. In all cases delivery was induced immediately after diagnosis was done and all died after 5-10 minutes of delivery (fig3). In US evaluation we demonstrated soft tissue mass similar to echogenicity of brain tissue in 5 cases and no soft tissue mass similar to brain tissue in 2 cases. In all cases the bony structure above the level of the orbits were absent. We observed normal meninges in one case and normal falx cerebri in two cases. There was no other anomaly in four cases. We demonstrated polyhydroamnios in 3 cases, anencephaly in two cases and spina bifida in two cases at the level of cervical vertebrae and cervicothoracal vertebrae. Placental maturation was in accordance with gestational age. There was no other visceral anomaly in our cases.

Discussion

Fetal acrania is a rare congenital anomaly that results from embryologic maldevelopment at the fourth week of pregnancy when the cranial end of neural tube closes. It is characterized by the absence of cranial vault and presence of substantial amount of brain tissue usually surrounded by a thin membrane. Anencephaly is believed to result from primary nonclosure of the anterior neuropore at the fourth week of the pregnancy as acrania and is characterized by the absence of the brain tissue. These two entities may be together and seperately as well. Acrania is also considered as a varient of anencephaly and is found in all anencephaly cases in various degrees (4,5). Because of their lethal outcome, early diagnose must be done for timely termination of pregnancy. In our study, we demonstrated anencephaly and acrania together in two cases at second and third trimester. In these cases the brain tissue and cranium bones above the level of orbita were absent. In both cases polyhydroamnios was associated pathological finding. The earliest diagnosis of anencephaly is possible whenever nonclosure neural tube is demonstrated with transvaginal ultrasonography during 9 to 10 weeks, menstrual age (MA) (5). But acrania must not be diagnosed prior to 11 weeks or 12 weeks, MA at the time when normal ossification of the cranial vault occurs. In the report of Ekici and Gülmezoğlu (6) it is said that sonographic diagnosis of fetal acrania can be made during the second trimester. Goldstein et al (3) and Bronshtein and

Figure 3. *Pathologic Specimen of the Fetus. Substantial Amount of Brain Tissue Is Depicted Nut No Calvarium Above the Level of Orbits. Cervical Spina Bifida Is Also Present .*



Orsoy (5) found it difficult to diagnose anencephally prior to 14 weeks; MA with standard transabdominal US techniques. In our study we demonstrated acrania at 13 weeks, MA in one case and we repeated sonographic evaluation two weeks later that revealed the same findings. Cranial vault ossification occurs nearly in 12-13 weeks, MA but it is advanced at 14 weeks, MA. So we concluded that for certain diagnose by transabdominal US 14 weeks, MA must be waited. The incidence of acrania and the incidence of acrania among the cases of anencephaly is unknown. But anencephaly incidence appears to be about 1 per 700 liveborn infants in the Jewish, 1 per 500 among Arabs (5). Among our own 7 acrania cases we demonstrated two anencephaly. In the other reports the incidence of acrania among anencephalics seems to be higher (5). But this necessitates further evaluation. Associated abnormalities may be found together with acrania (metatarsus varus, bilateral cleft lip, bilateral absence of orbital floors and spina bifida (6). In our cases spina bifida were present in 2 of 7 (28.5%) cases, polyhydroamnios in 3 of 7 (42.8%) cases. Severe osteogenesis imperfecta, congenital hypophosphatasia, exencephaly and cranioschisis are included in the differential diagnosis (4,6). There is poor mineralisation of calvarium in severe osteogenezis imperfecta and congenital hypophosphatasia so in ultrasonographic examination, it may be difficult to differentiate low level echos of calvarium bones. Exencephaly is a neural tube defect in which brain lie outside an imperfect cranium. Postnatal examination is required for definitive diagnosis.

References

1. Sanders RC. Prenatal ultrasonic detection of anomalies with a lethal or disastrous outcome. Radiol Clin North Am 1990; 25 : 163-190
2. Vergagni P, Ghidni A, Sirtori M. Antenatal diagnosis of fetal acrania. J Ultrasound Med 1987; 6: 715 -717
3. Goldstein RB, Filly RA, Callen PW. Sonography of anencephaly pitfolls in early diagnosis.J Clin Ultrasound 1989; 17: 397-402
4. Ekici E, Gülmezoğlu AM. Sonographic diagnosis of fetal acrania. J Clin. Ultrasound 1991; 19: 363-366
5. Bronshtein M, Ornoy A. Acrania : anencephaly resulting from secondary degeneration of a closed neural tube: two cases in the same family. J Clin Ultrasound 1991; 19: 230-234
6. Eric JM, Edmund SC, John SH. Sonographic demonstration of fetal acrania. AJR 1982; 139: 181-182
7. Mannes EJ, Crelin ES, Hobbins JS, et al: Sonographic demonstration of fetal acrania. AJR 1982; 139: 181

Correspondence :
Selami SUMA MD.
Department of Radiology
Faculty of Medicine, Atatürk University
Erzurum / TURKEY