Prenatal Ultrasonic Diagnosis of a Thoraco-Abdominal Ectopia Cordis

TOIUKO-ABDOMIN. , KORDİS'İNPRENA PAL UETRASONİK TANISI

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ÖZET

Skalp anomalisi de olan bir torako-abdominal tip eklopia kordis vakası rapor edilmiş ve bu nadir görülen vaka ışığında literatür taraması yapılarak özetlenmiştir.

Eklopia kordisin prenatal ultrasonografik tanısı ve bunun önemi ile tedavisi tartışılmıştır.

Analılar Kelimeler: Ektopia kordis, Fetal ultrasonografi

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SUMMARY

A case of thoraco-abdominal type ectopia cordis which had scalp abnormalities is reported, and the pertinent literature is briefly reviewed. We discuss the importance of prenatal ultrasonographic diagnosis and the management of this rare condition.

Key Words: Ectopia cordis. Fetal ultrasonography

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Prenatal ultrasonic diagnosis of congenital abnormalities can raise delicate decision problems. The finding of a defect in the sternum and the anterior abdominal wall is often associated with other anomalies. The most severe form is ectopia cordis including intra cardiac abnormalities. The present report describes the prenatal diagnosis and the clinical management of a fetus with ectopia cordis and scalp abnormalities. To our knowledge this is the fifth intrauterine diagnosis of this rare anomaly reported.

CASE REPORT

A 21-year-old woman attended the antenatal of the Ministry of Health Hospital, Ankara in the sixth week of her first pregnancy with emesis gravidarum. No abnormalities were reported at the routine physical examination and laboratory findings. This pregnancy had been uneventful until the seventeenth week. There was no history of infectious disease or use of drugs. In the seventeenth week, however, a routine ultrasound examination was done, and this revealed that there was a midline defect of the sternum and the upper abdominal wall (Figure 1 and 2). The pulsating heart was protruding out of the chest wall into the amniotic fluid. There were also some more strange findings in front of the midline defect in the thoraco-abdominal wall which later proved to be some part of the small bowell and the liver outside the abdomen (omphalocele). There were also associated anomalies found in the cranium. There was a large defect in the back region of the cranium, and a mass detected to be broin was present.

After consultation with pediatric cardiologists and a neurosurgeon, the intra uterine findings were concluded to carry a very poor prognosis for neona-

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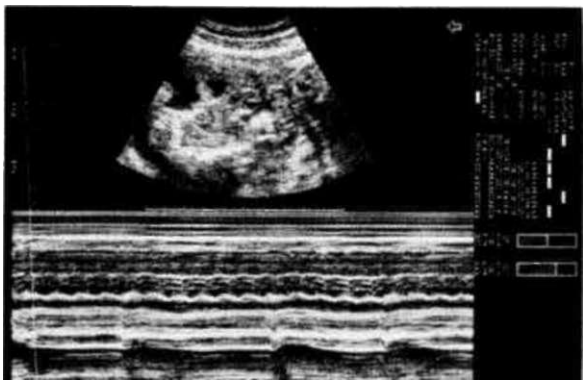

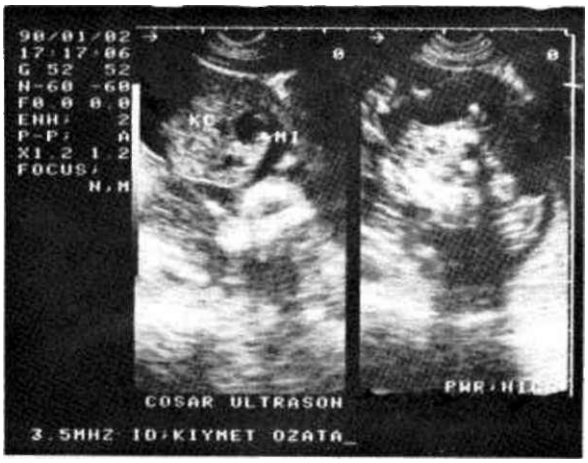

Figure 1. U'rcnalal ultrasonic examination ol Ihe iciu.s with thor.io hdnminul ectopia cordis, omphalocele, and cranial anomalies.

tal survival. It was the ultimate decision of the patient and her husband to terminate the pregnancy. In the eighteenth week of gestation labour was induced by intraeervieal prostaglandin E. The patient delivered a female fetus of 120 gr with suspected anomalies (Figure 3). The fetus died during the procedure. On the examination of the fetus: there was a large defect in the occipital region of the cranium. Heart dissection showed a double outlet right ventricle with perimembranous ventricular septum defect. The placenta showed no abnormalities. After two days the patient could be discharged in good physical health. Postpartum laboratory findings showed that Cytomegalovirus IgG (+), Cytomegalovirus IgM (-), Rubella IgG (+), Rubella IgM (-), Toxoplasmosis IgG (-), Toxoplasmosis IgM (-). Chromosomal analysis of the fetus revealed no abnormalities.

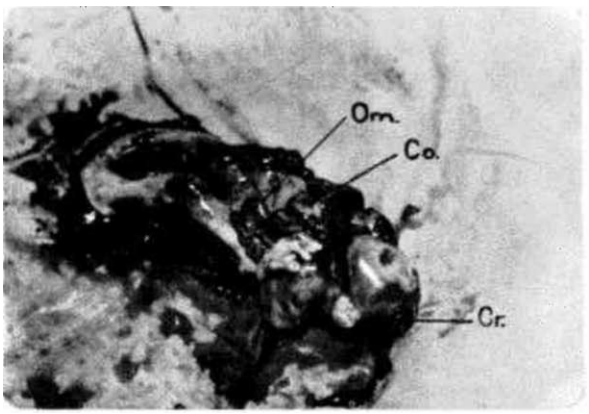

Figure .1. Postnatal phologruph demonstrating the Ictus vuth thoraco abdominal type ectopia cordis and omphalocele in combination **with** cranial anomalies

DISCUSSION

Ectopia cordis was defined by Abbott as a congenital malformation in which the heart is located out of the thorax (1). The first observation of it is generally attributed to Haller in 1706, but such malformations were recorded in the writing of the ancient Babylonians (2-3). Four types of ectopia cordis are described: cervical, thoracic, abdominal, and thoracoabdominal (1,4-5). Cantrell established the last group as a syndrome: a midline supraumblical abdominal wall defect, a defect of the lower sternum, a deficiency of the anterior diaphragm, a defect in the diaphragmatic pericardium, and congenital intracardiac defect (6). To our knowladge thoraco-abdominal ectoia cordis has been described in 78 cases (3,5,7-10). Omphalocele is the most common associated abdominal wall defect. Other congenital abnormalities were present in about 70% of the cases reported. They are mostly cranial (as in the case presented) and facial anomalies (3).

The life expectancy varies considerably with the presence of associated abnormalities. Although life with an cxtralhoracic heart is usually short (in the scries repotcd-the average duration of life was 36 hours) there arc cases reported in which the patients lived some months and even many years. The latter patients all had an abdominal or thoracoabdominal hearth with life (3). The presence of omphalocele and/or intracardiac abnormalities increases mortality and morbidity dramatically as seen in the case presented in this report (7). The unique aspect of the present case seemed to be the presence of a cranium defect. Even during

recent years treatment of thoraco-abdominal ectopia cordis has often been unsuccessful. The greatest progress in the treatment seems to be a correct fetal diagnosis of the abnormalities. Improvement of equipment along with introduction of equipment along with introduction of systematic scans of the fetal heart have permitted reliable diagnosis of congenital heart diseases. Extrathoracic heart is readily diagnosed in this fashion. The heart is easily visualized swinging in the amniotic fluid or within an omphalocele in cases of thoraco-abdominal type, a through examination of the fetal heart by echocardiograpy and a collaboration between obstetrician and pediatric cardiologist are the prerequisites for a careful prognosis, in selected cases sometimes leading to termination of pregnancy (3,11). The treatment of omphalocele and abdominal wall defect varies according to their extent and content (12). Immediately after birth one can consider coverage of the defect by a silastic prothesis in order to perform diagnostic workup of the intracardiac defects during the postpartum days (3,8).

The etiology of the mentioned malformation complex is not known. There is no proven causal relationship between ectopia cordis and chromosomal abnormalities, nor an increased risk for recurrence in future pregnancy (13).

In conclusion, prenatal ultrasonography is of great value for the assessment of the midline defects of the thoraco-abdominal wall. When the diagnosis has been made by fetal echocardiography, two possibilities should be considered with the parents participating actively in the decision: interruption of pregnancy or attempted rurgical repair. Partial protrusion of the heart outside the thorax without an omphalocele or intracardiac abnormalities can be considered for successful postnatal repair (14). However if an intracardiac abnormality is present particularly in combination with an omphalocele, the prognosis must be considered very poor.

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