Prenatal diagnosis of thoracic ectopia cordis: a case report

Maithem I. Sabeh[1], Ahmed AR. Ammar[2*], Ilham Al-Rubaei[3]

Abstract

Twenty years old primigravida lady has visited sonar clinic during the second trimester as a part of routine prenatal care after Shirodkar procedure for cervical weakness. The sonographic study revealed a single active viable fetus, of 15 weeks' gestational age, in slight hyperflex position. The placenta was normal and situated posteriorly, with normal amount of amniotic fluid. The fetus had anencephaly associated with heart extrathoracically. Family and maternal history revealed exogamous marriage with uneventful obstetric history. A second sonographic follow-up examination was done at 25 weeks' gestational age.

Keywords: Shirodkar procedure; Sonographic; Thoracic ectopia cordis

*Corrospondance author: ahmed.ar.ammar@gmail.com
[1]Ibn Al-Bitar center for cardiac surgery. Baghdad.
[2]College of Medicine, Muthanna University.
[3]Gynecological and Obstetrics department. Al-Hakim teaching hospital. Najaf, Iraq.
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Introduction

Thoracic ectopia cordis is a rare congenital anomaly in which the heart located outside the thoracic cavity through a sternal defect. It may be partial or complete depending on degree of displaced heart [1, 2]. In this review we reported a case with prenatal ultrasound diagnosis of "thoracic ectopia cordis."

Case Report

Twenty years old primigravida lady has visited sonar clinic during the second trimester as a part of routine prenatal care after Shirodkar procedure for cervical weakness. The sonographic study revealed a single active viable fetus, of 15 weeks' gestational age, in slight hyperflex position. The placenta was normal and situated posteriorly, with normal amount of amniotic fluid. The fetus had anencephaly associated with heart extrathoracically. Figure 1. Family and maternal history revealed exogamous marriage with uneventful obstetric history. A second sonographic follow-up examination was done at 25 weeks' gestational age.

Discussion

Ectopia cordis is an extremely rare and fatal congenital anomaly being described 5000 years ago. It occurs in range between 5.5 - 7.9 per million live-births [3, 4].

The hypothetical etiologic concept is thought at the establishment of demarcation between intra and extra-embryonic coelom about third week of development [5].

The improper progress of cephalic and lateral folding in addition to incomplete development of the body wall structures i.e. muscles, bone and skin is the basic embryologic defect. This will lead to lack of complete fusion of the lateral folds in the formation of the thoracic wall during fourth week. The sternum may be either absent, in most variant, or present with large defect. Sometimes be bifid and heart protrudes through the anterior wall. The heart is covered by skin or pericardial sac or coated with visceral pericardium only. Published cases suggest family inheritance and show evidence of a mutation in a gene mapped in Xq25 – q26 6.

Ectopia cordis associated with a number of anomalies which may be either intra or extra-cardiac. The intracardiac anomalies form 80.2% of the cases. These include: ventricular septal defect, aterial septal defect, tetralogy of Fallot and diverticulum of ventricle. The non-cardiac anomalies include: abdominal wall defects, cranial and facial malformations, cleft lip and palate, anencephaly, hydrocephaly, neural tube defects, pulmonary hypoplasia, genitourinary malformation, gastrointestinal defect, and chromosomal abnormalities [7-9]. The mortality rate remains high for neonate with untreated thoracic ectopia cordis especially in the presence of cardiac anomalies or other malformations, with only a few reported survivors [10].

Prenatal sonographic imaging remains the simplest available diagnostic tool.

Many reports have been diagnosed ectopia cordis during the first trimester even as early as 8 – 9 weeks of the pregnancy; however, the modest diagnostic time is at 10 weeks [11-14]. Repondek-Liberska et al [4], suggested that 26th week was the average gestational age for diagnosis. We think that this limit may be late because of earlier delineation of anatomical defect. The diagnostic modality used was Doppler sonography. However, three-dimensional ultrasound and its combination with Doppler allows for a more accurate early prenatal diagnosis [15]. In accordance to our society and cultural background, there is a controversy about the termination of the affected pregnancy owing to religious and customary reasons. Ectopia cordis carries high mortality with overall dismal prognosis especially in patients having complete forms or associated with extra and intracardiac anomalies [15]. We place an emphasis on earlier diagnosis of ectopia cordis in first trimester or first weeks of second trimester in order to assist the concern family to take a decision for terminate of pregnancy. The diagnosis is settled by outside abnormal location of the heart in association with other intra and extracardiac anomalies, which are common. In case of non-terminated pregnancy despite early or delayed diagnosis to third trimester early surgical palliation should be performed, still it carries high mortality.

Successful interference depends on absence of significant intra and extracardiac anomalies and availability of adequate skin to cover the midline defect. In this review, the mother opted to complete her pregnancy, however, it ended with spontaneous abortion in 26th weeks of gestation.

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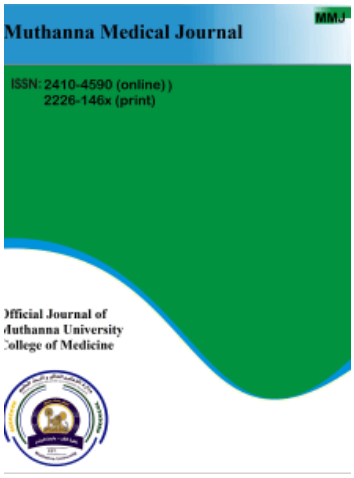

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Contact us
Postal Mail
Muthanna Medical School
Samawah
Tel: +964 (782) 542-5669
Office's business hours: Sunday-Thursady 9.00 am – 1.00 pm
Email: yousif_ghaly@yahoo.com
Email for technical assistance: shatha.farhan@gmail.com