CASE REPORT

Thoracic Ectopia Cordis in an Ethiopian Neonate

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ABSTRACT

BACKGROUND: Ectopia Cordis is defined as complete or partial displacement of the heart outside the thoracic cavity. It is a rare congenital defect with failure of fusion of the sternum with extra thoracic location of the heart. The estimated prevalence of this case is 5.5 to 7.9 per million live births.
CASE PRESENTATION: We had a case of a 16-hour-old male neonate weighing 2.9kg with externally visible, beating heart over the chest wall. Initial treatment included covering the heart with sterile-saline soaked dressing, starting systemic antibiotics and supportive care. A staged surgical approach to this defect with the initial aim of replacement of the heart to the thoracic cavity was opted. The neonate died twenty minutes after the surgical intervention due to cardiogenic shock despite adequate resuscitative measures.
CONCLUSION: This case report underscores the missed opportunity of antenatal ultra-sonographic diagnosis and the challenge of Ectopia Cordis treatment in Ethiopia.
KEYWORDS: Ectopia Cordis, case report, antenatal ultrasound, Ethiopia, treatment challenge

INTRODUCTION

Ectopia Cordis (EC) is one of the rare congenital anomalies characterized by complete or partial displacement of the heart outside the thoracic cavity. The estimated prevalence of EC is 5.5 to 7.9 per million live births (1). Prenatal diagnosis is established by ultrasonography by visualizing the heart outside the thoracic cavity (2-5).

EC is classified into cervical, thoracic, thoracoabdominal and abdominal. Pentalogy of Cantrell is considered when a combination of thoracoabdominal EC, anterior diaphragmatic hernia, lower sternal defect and midline supraumblical defect occurs. Although surgical techniques have evolved, the prognosis and survival are limited; thoracic type has the worst prognosis while the thoracoabdominal EC has a better prognosis (6-8).

An EC case neonate was seen and intervened at Hawassa University Referral Hospital, Hawassa, Ethiopia. It necessitated literature review and case report. Ectopia Cordis (EC) is reported rarely from African settings.

CASE PRESENTATION

A 16-hour-old, vaginally delivered full-term male neonate, born to a 29 years old Para IV mother was referred from a primary hospital after exposed beating heart was found over the chest wall with the patient experiencing difficulty of breathing. The delivery was attended at home by a traditional birth attendant. All siblings were alive and healthy. There was no history of consanguinity, infection, radiation, drug or any known herbal exposure. There was no family history of any such or related congenital heart diseases. Previous pregnancy courses were uneventful. Health center antenatal care was uneventful although no ultrasonography study was done.

Physical examination showed a full-term neonate of 40 weeks with heart rate- 124/ min, respiratory rate- 60/min and SpO_2 - 77%. The upper sternum was deficient with the heart lying outside the thoracic cavity and without pericardial protection with cephalic orientation of its apex (Figure 1, Figure 2). The abdomen was intact. Laboratory test revealed: hemoglobin- 19g/dl, total leukocyte count- 20.5x 10^3/ml, differential count: L-25.5 %, P-64.3 %, Platelets count- 173x10^3/ml, blood group of O +ve and imaging studies were not done.

The patient was kept in a temperature regulated room. Initial prompt treatment included covering of the heart with sterile-saline soaked dressing, systematic treatment and supportive care. Then, a staged surgical approach to this defect with the initial aim of replacement of the heart to the thoracic cavity was chosen. The neonate died twenty minutes after the surgical intervention due to cardiogenic shock despite adequate resuscitative measures. Postmortem examination could not be processed due to familial reasons.

Figure 1: *Lateral view of the Ectopia cordis neonate*

Figure 2*: Frontal view of the Ectopia cordis neonate*

DISCUSSION

EC is a rare congenital abnormality with reported point prevalence of 5.5 to 7.9 per million live births (1)**.** The burden of the disease is inadequately known in Africa due to limited reports (2-5). It is classified into Cervical, thoracic, thoracoabdominal and abdominal (6). Only few patients with thoracic type have survived and the thoracoabdominal EC has a

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better prognosis. Our case was compatible with the thoracic type (6-8).

Thoracic EC was explained embryonically by the rupture of the chorion at 3 weeks of gestation with resultant compression of the thoracic cavity and failure of descent of the heart at this stage. The possibility of amniotic bands is also ascribed. EC may occur in isolation or in association with other ventral body wall defects (7).

Antenatal ultrasound during the first trimester helps in the diagnosis of EC. In our case, ultrasonography study was not performed during pregnancy. EC was diagnosed postnatally after a home delivered neonate was brought to our hospital (2,3,9).

There was no history of consanguinity, similar history or any congenital heart disease in our case. Although the genetic causes for EC are not exactly known, certain associations with chromosomal abnormalities have been reported (7). Testing facilities for such cases are nonexistent in Ethiopian Settings.

Survival of thoracic EC case is limited despite advances in care, and its management is challenging. Aggressive surgical procedures are recommended to increase the survival (7,10). In our case report, death occurred within twenty minutes of the initial surgical intervention. Earlier studies showed a lethal course of the thoracic EC.

In conclusion, this report underscores the missed opportunity of antenatal ultrasound diagnosis and the challenges in the management of EC in Ethiopia.

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