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Complete Thoracic Ectopic Cordis-A Rare Case Report

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Abstract

Ectopic cordis is a condition where heart is located in an extra thoracic position. The incidence is 5-8 per million. Hence, we are reporting this case.

Keywords: Ectopia; Cordis; Thoracic; Complete

Introduction

Ectopic cordis is a rare congenital malformation in which the heart is displaced completely are partially outside the thoracic cavity [1,2]. It may occur as an isolated malformation or associated with other anomalies such as omphalocoele, congenital heart disease or Cantrell syndrome. It is classified into 4 types- cervical, thoracic, thoracoabdominal and abdominal. Here we are reporting a case of complete thoracic ectopia cordis (Figure 1) [3].

Case Report

A 25 year, 2nd gravid mother presented to our hospital with labour pains, with a 24 week antenatal USG suggestive of cardiac anomaly. There was no known consanguity or family history of any congenital abnormalities. The antenatal course was uneventful and no medication were taken during her pregnancy. a live, 2.1 kg, male child was delievered at 37 weeks of gestation through normal vaginal delievery. The baby was received limp and cyanosed with a defect in the anterior chest wall and heart protruding out through it. Baby was immediately intubated and IPPV started. Spontaneous respiratory efforts appeared at 1 minute 30 seconds of life with an Apgar score of 3/10 and 6/10. But the neonate expired within 3 hours of birth. The examination revealed a defect of about 4-5 cm in the thoracic wall near 3rd-4th intercostal space on left side with evisceration of the heart through it. USG, X ray or CT scan could not be done due to poor general condition of the baby and autopsy could not be done as the attendants refused for it.

Discussion

When the heart is located outside the chest wall, it is known as ectopia cordis. It is derived from Greek word 'ectopic' which means out of place and Latin word 'cordis' which means heart. The heart is not covered by any pericardium, bony cover and skin. Failure of proper maturation of midline mesoderm and ventral body wall formation during embryonic development may be the probable etiology. The exact etiology is not known but abnormalities in the lateral body wall fold are believed to be involved. Normal lateral folds are responsible for fusion in midline to form ventral wall [4]. It is classified as thoracic (60%), abdominal (30%), thoraco abdominal (7%), cervical (3%), cervicothoracic (<1%). The thoraco abdominal type is regarded as a distinct syndrome known as Cantrell's pentology which includes distal sternum defect, midline supraumbilical abdominal wall defect, ventral diaphragmatic hernia, defect of the apical pericardium and congenital intracardiac defects [5]. Most common intracardiac defects include VSD and Fallot's tetralogy. The prognosis is grave and depends on location, extent and presence of congenital defects. The main causes of death includes infection, cardiac failure and hypoxia.

Treatment options are limitated. Numerous corrective and palliative operations have been proposed but overall prognosis is poor.

Figure 1: Showing complete thoracic ectopia cordis.

Conclusion

Ectopia cordis is a rare congenital malformation which can be diagnosed with an antenatal USG. Due to its poor prognosis, termination of pregnancy prior to viability should be considered.

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