Asian Journal of Case Reports in Surgery

10(3): 1-5, 2021; Article no.AJCRS.70347

Thoracic Ectopia Cordis: Cardiothoracic Surgeon's Perspective

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Authors' contributions

This work was carried out in collaboration between both authors. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Yasushi Shibata, University of Tsukuba, Japan.
(2) Dr. Ashish Anand, G. V. Montgomery Veteran Affairs Medical Center, USA.
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Reviewers:
(1) Tommaso Hinna Danesi, University of Cincinnati, United States.
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(3) Antonio da Silva Menezes Junior Federal University, Brazil.
Complete Peer review History: http://www.sdiarticle4.com/review-history/70347

Case Study

Received 01 May 2021
Accepted 06 July 2021
Published 09 July 2021

ABSTRACT

Thoracic ectopia cordis is a rare congenital abnormality with a complete partial displacement of the heart outside the thoracic cavity. Prognosis is poor and depends on the severity of intracardiac defects. With its high mortality and <5% survival rate, management is challenging. Here we report a case of ectopic cordis in a pre-term newborn presented with a heart totally outside the thoracic cavity without pericardium protection. Due to the lack of antenatal diagnosis, cardiopulmonary complications and poor prognosis, we accepted masterly inactivity. This approach was best for the family, society and other available resources. Unfortunately, Neonate succumbed to death within 24 hours of life.

Keywords: Ectopia cordis; thoracic; prenatal; surgery; palliative; cardiothoracic surgeon.

1. INTRODUCTION

Ectopia cordis [EC] is a rare congenital malformation defined as a complete or partial displacement of the heart outside the thoracic cavity through a parietal pericardium, diaphragm, or sternal defect [1]. The prevalence is estimated to be between 5.5 and 7.9 per 1 million live births

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[2]. This sporadic malformation is believed to be a disorder of morphogenesis early in embryonic life, occurs due to failure of maturation of the midline mesodermal components of the chest and abdomen. Based on the degree of soft tissue coverage, it can be either partial EC or complete EC, where the heart lies outside the thoracic cavity and has neither skin nor pericardial covering. It can also be classified as cervical, cervicothoracic, thoracoabdominal, or abdominal [3,4]. Despite advances in neonatal cardiac surgery, complete thoracic EC has a poor prognosis, thus remains a surgical challenge with only a few long-term survivors [5]. Here, we present a case of complete thoracic EC in a newborn baby and a literature review from a cardiothoracic surgeon's perspective.

2. CASE REPORT

A preterm male neonate of 34 weeks gestation with complete thoracic EC is referred to the Department of Cardiovascular and Thoracic surgery from a remote hospital for further management. The neonate was born through normal vaginal delivery, appropriate for gestational age. The young mother was nulliparous, with no associated maternal history of intake of medications, drugs, cigarette smoking, or alcohol abuse. There was no family history of consanguinity or congenital malformation. However, her antenatal checkups were irregular.

On examination, the anterior chest wall defect was noted, measuring 8cm x 7cm in craniocaudal and horizontal diameters with naked heart outside the thoracic cavity. Anterior fontanelle was patent with no other midline defects and primitive reflexes showed a delayed response. Abdominal examination was unremarkable with well-formed external genitalia. Echocardiography showed ventricular septal defect and patent foramen ovale. As the neonate was dyspnoeic, tachypnoeic, and had a delayed cry, cardiopulmonary resuscitation was considered, and the neonate was put on mechanical ventilation and inotropic support. In view of severe cardiopulmonary insufficiency, surgical repair of chest wall closure was not attempted. However, we covered the exposed heart with saline-soaked gauze pad wrappings to prevent desiccation and heat loss. We accepted masterily inactivity because of the poor prognosis, socioeconomic status of the family and resources available. Unfortunately, cardiogenic shock worsened, and the neonate succumbed at 24 hours of life.

Fig. 1. Complete thoracic ectopia cordis

3. DISCUSSION

Ectopia cordis is a rare entity representing only 0.1% of all congenital heart anomalies [6]. The first case of ectopia cordis was reported by Stensen in 1671 and was described by Haller et al. in 1706 [7]. It is caused by failure of midline fusion of the paired cartilages of the embryonic sternum and cardiac descent arrest. The pathogenesis still remains an enigma; it is hypothesized that the rupture of the chorion and yolk sac at 3th-4th embryonic weeks with resultant compression of the thoracic cavity fails descent of the heart. The possibility of amniotic band syndrome is also ascribed [8,9]. It may also occur secondary to exposure to intrauterine drugs or in association with other congenital anomalies [2,10]. The mother, in our case, denied intake of any unprescribed medication, with no history of consanguinity.

EC can be partial with heart pulsating through the skin or complete, as in our case, with the displacement of the naked heart entirely outside the thoracic cavity with or without pericardium. Depending upon the location of the heart and underlying defect, Weese (1818) and Todd (1836) classified it into five types: cervical (3%), cervicothoracic (<1%), thoracic (60%), thoraco-abdominal (7%) and abdominal (30%) [5,7]. Around 80.2% of cases present with intracardiac defects such as ventricular septal defect (100%), atrial septal defect (53%), tetralogy of Fallot (20%), left ventricular diverticulum (20%), and pulmonary hypoplasia. It can also be associated with other congenital and chromosomal abnormalities [3,5]. Our case was a complete thoracic variant with no abdominal changes.

Although with ultrasonography, EC can be diagnosed as early as between 9-12 weeks of pregnancy, three-dimensional ultrasonography combined with doppler gives accurate findings. Alternatively, Magnetic resonance imaging (MRI) and fetal echocardiography can be used to monitor the development of the condition throughout pregnancy and postnatal analysis of associated cardiac anomalies [11]. Unfortunately, our patient belonged to a rural background with infrequent prenatal checkups and tests; hence they were unaware of the condition till birth. Prenatal diagnosis is important for parental counselling, planning delivery and postnatal management. It also provide parents with option for termination of pregnancy in view of its extreme poor prognosis.

Differential diagnoses include cyllosomus and amniotic band syndrome. The distinguishing feature is the position of abdominal wall defect with the umbilical cord insertion, organ evisceration, the presence or absence of membranes or bands, and associated anomalies. Cyllosomus have two of the three anomalies as the diagnostic criteria- thoracic or thoracoabdominal defects, limb defects, and craniofacial defects with associated cardiac anomalies [12]. At the same time, amniotic band syndrome presents as abdominal or thoracoabdominal defects with omphalocele and craniofacial defects with the characteristic finding of ring deformities of extremities affecting finger and toes, pseudosyndactyly appearance, unlike in that of cyllosomus [13].

Thoracic type is invariably associated with a dismal prognosis, with only <5% of neonates surviving beyond the first month of life [7]. Most infants are stillborn or die within the first few hours or days of life from infection, cardiac failure, or hypoxemia. To date, very limited long-term survivors were recorded, those who underwent early palliation and surgery [3,10]. Successful corrective or palliative surgery can be performed despite high mortality rates. Cutler and Wilens (1925) attempted the first EC repair surgery, followed by many attempts by surgeons after that [2,14]. In contrast, the partial thoracic EC can be repaired electively by primary closure of sternal cleft after freshening the edges and making oblique relaxing incisions in the costal cartilages bilaterally. However, complete EC requires a multistage approach involving a primary soft tissue coverage of the heart, replacement of the heart into the thoracic cavity, repair of the intracardiac defects, and reconstruction of the chest wall [5,6].

Neonates with this life-threatening anomaly require intensive care from birth. Covering the exposed heart and viscera with saline-soaked gauze pad wrappings to prevent desiccation and heat loss is the primary goal in the management strategy [15]. A primary approximation can be achieved by using split-thickness skin graft, cadaveric skin graft, or prosthetic material, following which a slow reduction of the defect can be achieved. During the neonatal period, due to limited mediastinal space, reduction of the heart into a small thoracic cavity often produces compression and kinking of the great vessels leading to low cardiac output and hemodynamic instability [2,5]. In such cases, a Blalock-Taussig shunt or pulmonary artery band can be placed to

allow expansion of the thoracic cavity. During this time, prostaglandins can be used for decreased pulmonary flow. Also, placing a traction suture in the apex of the heart helps to bring the heart through the chest wall into the thoracic cavity. With improved clinical development, the heart is returned to the left or right pleural spaces in the second stage of the corrective heart procedure performed between 6 months and two years of age. Finally, following the correction of intracardiac defects, final chest wall reconstruction is carried out at a later date in collaboration with a plastic surgeon. In our case, since the patient was referred after birth and in a critical condition, we could not carry out surgery [3,5,15]. Hence, we considered managing palliatively. Management of EC requires multidisciplinary approach by a team of gynaecologist, neonatologist, cardiac surgeon, thoracic surgeon, plastic surgeon and a pediatric surgeon with advanced neonatal care unit.

4. CONCLUSION

Despite the advances in neonatal surgeries, surgery of complete EC remains a challenge due to the small thoracic cavity and position of great vessels. Prenatal diagnosis is of utmost importance, which invariably guides the surgical team to estimate the prognosis and deduce a postnatal management strategy.

CONSENT

Inform consent obtained for the purpose of publication

ETHICAL APPROVAL

It is not applicable.

ACKNOWLEDGEMENT

I would like to thank Melisha R Pinto (melisha.pinto@gmail.com)for proof reading and editing the manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/70347