Case Report

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Ectopia cordis with multiple ventricular septal defect and sternal cleft in a newborn: a case report

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Introduction and importance: Ectopia cordis is a rare type of malformation where the heart is not located normally. It may be partially or completely located outside the thoracic cavity and can be associated with other congenital abnormalities.
Case presentation: This case report is of a 34-week- and 6-day-old female fetus with a birth weight of 2040 g, a height of 41 cm, and a head circumference of 32 cm. The initial physical examination revealed a responsive newborn with an externalized heart outside the chest, with pericardium protection. In addition, a thoracic wall defect was detected suggesting incomplete formation of the septum bone. Moreover, in this case, the echocardiography report showed a multiple ventricular septal defect.
Conclusion: The management of ectopia cordis is a challenge for any obstetrician and pediatric surgeon due to its rarity. It causes mental agony and anxiety to the parents. With an early diagnosis, termination of pregnancy can be one of the options. Once it is diagnosed late, it needs a multidisciplinary approach, and the services of a very experienced pediatric surgeon to improve the prognosis.

Keywords: congenital cardiac anomaly, ectopia cordis, sternal cleft, ventricular septal defect

Introduction

A congenital heart defect (CHD), which also called congenital heart anomaly and congenital heart disease, is defined by a defect in the structure of the heart or great vessels that is present during birth[1]. The classification of these type of abnormalities is cardiovascular disease, and signs and symptoms depend on the kind of defect could vary from little to life-threatening[2]. Chest pain could not be presented in CHD, but the complications can lead to heart failure[1]. A rare example of CHD diseases is ectopic cordis, which is a congenital condition in which a part or the entire of the infant's heart does not have the typical coverage of the breastbone or the heart is abnormally located either partially or entirely outside the thorax[3]. The prevalence of ectopic cordis is 8 per million childbirths[4]. Furthermore, the causation of ectopic cordis is a failure of appropriate maturation of mid-line mesoderm and ventral body wall (chest) formation during the development of embryonic, but the precise reason for this abnormality is still unknown[5].

Another one of the congenital heart conditions is Ventricular Septal Defect (VSD). VSD is the most common CHD and constitute 20–30 present of children seen in large pediatric cardiology clinics[6]. The exact prevalence is unbeknown to researchers, but estimates ranging from 2 to 5 out of every 1000 newborns[7]. It is noteworthy that in rare cases there could be several VSD, which is defined as multiple VSD[6]. In this condition, the presence of more than one foramen in the muscular septum of ventricular is expected, and the extent of the openings may vary from pin size to substantial ones[7].

Another rare chest wall malformation is congenital absence of the sternum, or a complete sternal cleft, which resulting from failed mid-line fusion during the development of embryonic[8]. Significant morbidity can cause by this abnormality and, like other congenital anomalies, associated defects can be derived

HIGHLIGHTS

- A rare example of congenital heart defect diseases is ectopic cordis.
- Ventricular septal defect is the most common type of congenital heart defect.
- Management of ectopia cordis is a challenge for any pediatric surgeon.

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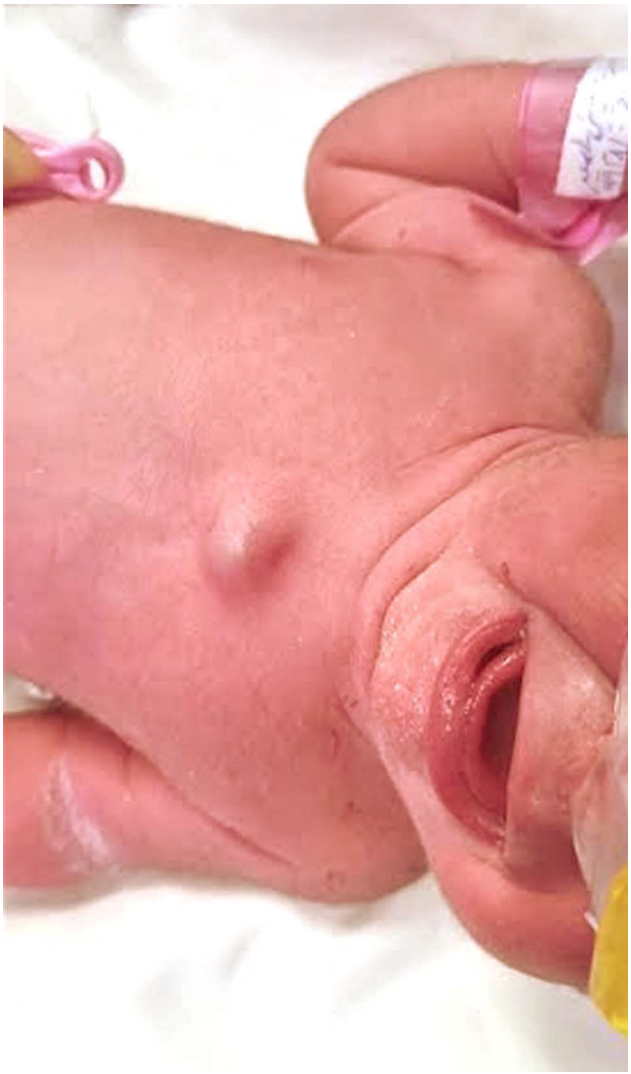

Figure 1. Ectopia cordis in the newborn.

from[9]. The work has been reported in line with the Surgical CAse REport (SCARE) 2020 Criteria[10], Supplemental Digital Content 1, http://links.lww.com/MS9/A154.

Case presentation

The present case report is of a 34-week- and 6-day-old female fetus born to a 40 year-old Kurdish mother through normal vaginal delivery at Kawsar Obstetrics and Gynecology Hospital, Urmia, Iran. She (his mother) had a history of cesarean delivery at the age of 34, where the newborn was completely healthy. She was from a family with low socioeconomic status and lives in a village far from Urmia city. She had seen a gynecologist only twice during this pregnancy. She also mentioned a history of taking levothyroxine for her hypothyroidism. She had no history of smoking and denied a history of alcohol and drug abuse. Moreover, she did not state any history of exposure to teratogens during pregnancy, especially in the first trimester. During history-

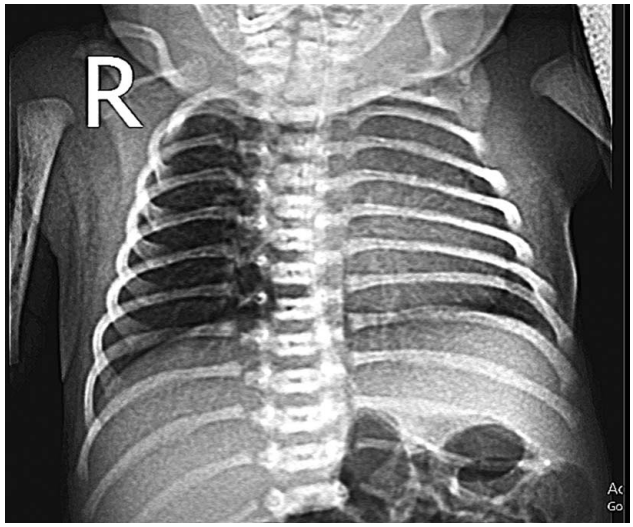

Figure 2. The chest radiograph of the newborn.

taking at admission, the parents did not mention the history of congenital anomalies or close family marriage in their relatives.

After the onset of labor signs, had an emergency lower segment caesarian section with an outcome of alive male baby born with an Apgar score of 9 at 1 min and 10 at 5 min. The mother gave birth to a 34-week- and 6-day-old female fetus with a birth weight

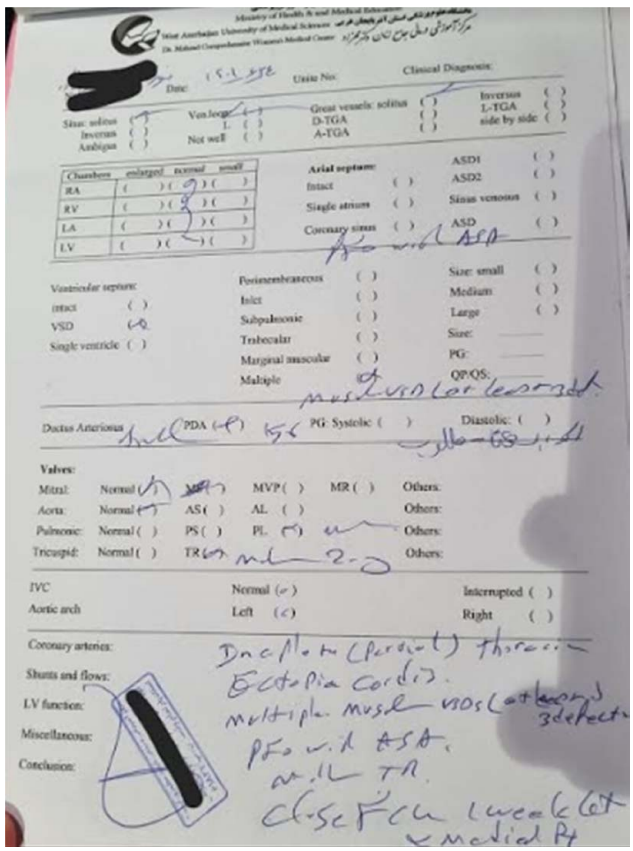

Figure 3. The echocardiography report of the newborn.

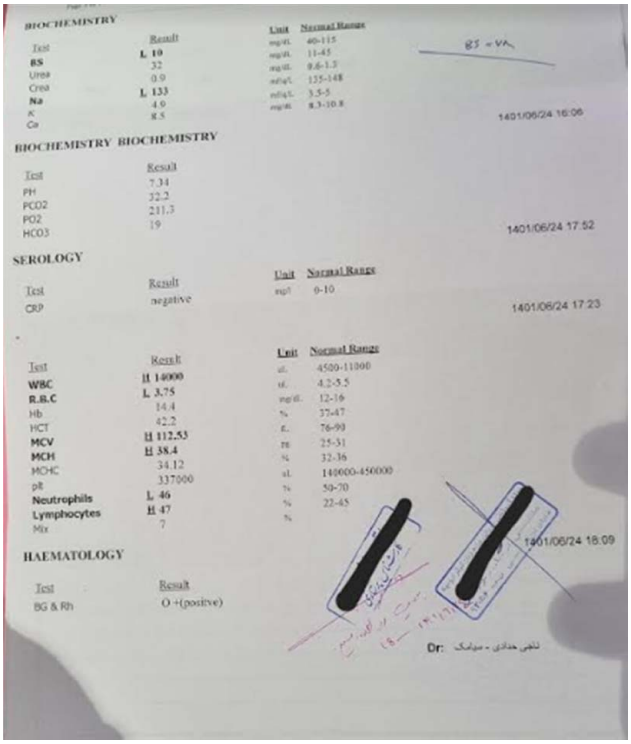

Figure 4. The laboratory results of the newborn.

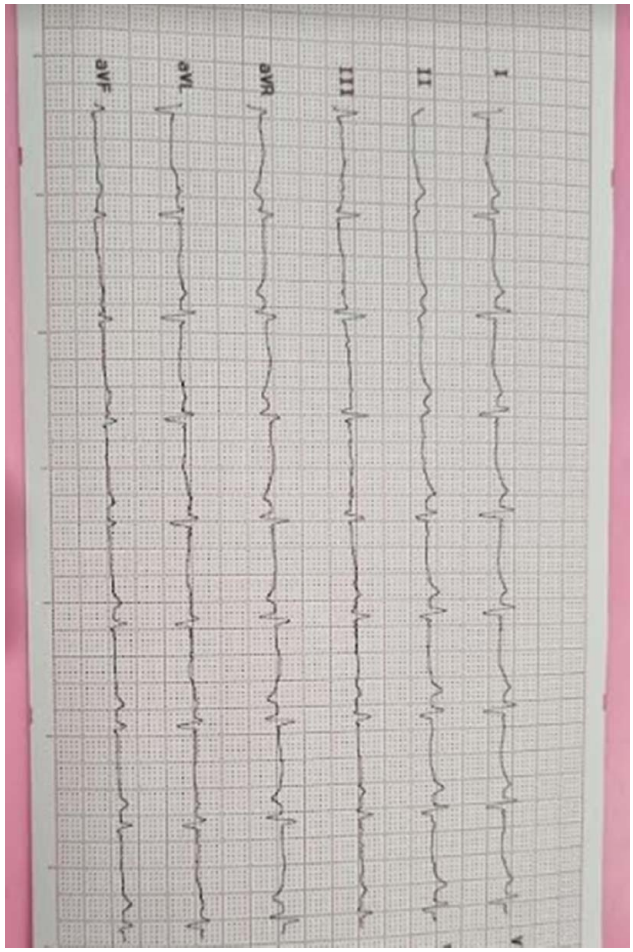

Figure 5. The ECG of the newborn.

of 2040 g, height of 41 cm, and a head circumference of 32. The initial physical examination revealed a responsive newborn with an externalized heart outside the chest, with pericardium protection. The dynamic movement of heartbeats outside the chest cavity is shown in the supplementary file (SDC. 1), Supplemental Digital Content 2, http://links.lww.com/MS9/A155, and a protruding heart is obvious on the skin of the chest (Fig. 1). In the chest radiograph conducted at birth, it was shown to be an incomplete formation of the septum bone (Fig. 2). The remainder of the physical examination was normal. Her vital signs at birth were as follows: Temperature: 36.4, Heart Rate: 173 bpm, Respiration Rate: 65 bpm, Blood Pressure: 57/25 mmHg, Oxygen Saturation: 96%. The newborn was transferred to the Neonatal Intensive Care Unit and placed in an incubator. Moreover, in this case, the echocardiography report showed a multiple VSD (Fig. 3). Other organs were shown to be normal, and no other congenital malformations were found. In addition, some of the patient's laboratory information during admission is provided in Figure 4. The initial electrocardiogram on day 1 showed in Figure 5.

Discussion

One of the rare heart congenital conditions is ectopic cardis[2]. As a portion or all of a baby's heart is located outside the cavity of the chest, ectopia cordis occurs[3]. This condition can occur as early as the first trimesters and is often diagnosed by ultrasound device[4]. It becomes obvious as soon as the baby is born, if not discovered during pregnancy[1]. There is some type of this abnormality, which include, cervical (3% of cases), thoracic (64% of cases), thoracoabdominal ectopia (18% of cases), and abdominal ectopia (15%) [5]. It is quite important to note that the type of ectopia cordis can be depended on whether the affected heart is totally uncovered, covered by a serous membrane or covered by skin, which this categorization and as well as the associated heart defects are determining the treatment options[4]. In cope with ectopic cardis, in utero options and postbirth surgeries are considered. When a baby is diagnosed with this severe birth defect in utero, the parents can opt to terminate the pregnancy, as the ectopia cordis survival rate is around 10%[11]. Newborns with lethal congenital malformations, even if they survive longer than expected, become severely impaired and palliative care should be considered[12].

On the other hand, infants with ectopia cordis who are born with their hearts partially or completely outside of their bodies, usually have other organ structures that also develop abnormally[5]. Ectopia cordis often coexists with additional heart defects, such as abdominal wall defects[4]. VSD is a septal defect of the heart that is defined by the existence of a foramen between the two ventricular which by the oxygenated blood will mix with the nonoxygenated stream and the consequences could be mild to severe cyanosis[6]. Multiple VSD called the situation in which there are several holes in the septal of the heart, and the prognosis in such cases is poor[7]. One else abnormality which is seen in these newborns with ectopic cordis is the sternal cleft. Sternal cleft

is a chest wall malformation that can expose mediastinal viscera and other chest components to injuries. It can be classified into two complete and partial forms[8]. In the year of 2020 in Indonesia, Limanto, and Soebroto did a palliative surgery on a 5-day-old full-term newborn for a complete thoracic ectopia cordis with the aim of covering the exposed heart with the use of bovine pericardium material that is commonly used in reconstruction in cardiac surgery. Unfortunately, the patient expired on the third postoperative day due to heart failure[13]. In the year 2021, Yıldız *et al.* in Turkey reported four cases of ectopia cordis with abdominal, thoracoabdominal, partial thoracoabdominal, and middle sternum defects that were treated surgically. They concluded that the degree and level of exposure of the heart preclude initial survival, and complex repairs of intracardiac defects may be difficult in cases requiring shunts for long-term palliation[14]. In the same year in Iraq, Aboud *et al.*[15] reported a case of complete thoracic ectopia cordis, but due to poor general condition, the newborn expired on 36 h of life before any intervention. Kebalo *et al.*[16] in the Togolese Republic reported a case of midsternal defect of ectopia cordis and the newborn expired 22 h after birth, which also made them unable to perform any procedure. The most recent case reported in the year 2022 from Saudi Arabia by Ishamiri *et al.*[17] was a case of incomplete ectopia cordis in a 23-year-old male, who expired due to coronavirus disease 2019 infection.

Conclusion

The management of ectopia cordis is a challenge for any obstetrician and pediatric surgeon due to its rarity. It causes mental agony and anxiety to the parents. With an early diagnosis, termination of pregnancy can be one of the options. Once it is diagnosed late, it needs a multidisciplinary approach, and the services of a very experienced pediatric surgeon to improve the prognosis.

Ethical approval

None.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

R.G., N.F., M.F., M.A.: study concept, data collection, writing the paper, and making the revision of the manuscript following the reviewer's instructions; L.H., S.M.B., P.N.: study concept, reviewing, and validating the manuscript's credibility; M.H.B. and M.A.: reviewing and validating the manuscript's credibility.

Conflicts of interest disclosure

None.

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