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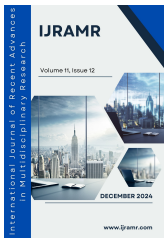

CASE REPORT

ECTOPIA CORDIS WITH FETAL HYDROPS AND SEVERE OLIGOHYDRAMNIOS: A CASE REPORT

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ABSTRACT

Ectopia cordis is a rare congenital anomaly where the heart is partially or completely extruded from the thorax. It can occur with other associated defects, especially as part of the Pentalogy of Cantrell (a collection of 5 congenital midline birth anomalies; involving the sternum, heart, pericardium, diaphragm, and abdominal wall). Mostly, the prognosis for infants with ectopia cordis is poordue to the complexity of management, inadequate expertise, and delayed diagnoses.We examine a rare presentation of ectopia cordis in a 34-year-old woman at 36 weeks gestation, complicated by fetal hydrops and severe oligohydramnios. Taking into account the poor prognosis, conservative management was chosen. The patient went into labor at 38 weeks and 4 days, and the outcome was a live male infant who passed away 5 minutes after delivery. We explore the clinical intricacy and significance of timely antenatal screening and multidisciplinary management in high-risk pregnancies.

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INTRODUCTION

The term "ectopia" is derived from the Greek word "ektopos," meaning "out of place" (1). Ectopia cordis (EC) is a rare congenital anomaly where the heart is either partially or completely extruded outside the thoracic cavity (1-4). Its prevalence is approximately 5.5-7.9% per million live births (2). EC is often associated with abnormalities such as omphalocele, congenital heart defects, and skeletal deformities (5). The prognosis is usually unfavorable, with most cases culminating in stillbirth or early neonatal death (6). This case calls attention to a rare presentation of ectopia cordis with fetal hydrops and severe oligohydramnios, prompting an exploration into antenatal diagnosis, management strategies, and outcomes.

CASE PRESENTATION

A 34-year-old woman, gravida 4, para 2+1 at 36 weeks and 2 days gestation, was referred from a peripheral facility due to concerning ultrasound findings. At 20 weeks gestation, she attended that peripheral facility for a routine anomaly scan; however, she received the sonographic findings report later at 36 weeks during one of her routine ANC visits. She had 3 ANC visits, the first at 16 weeks gestation, the second at 20 weeks, and the last at 36 weeks. This delay was occasioned by poor ANC attendance and failure for an active follow-up or referral to a maternofetal specialist. The ultrasound report revealed a single viable fetus in cephalic presentation with ectopia cordis, fetal hydrops, and severe oligohydramnios. An antenatal ultrasound scan of the anomaly is shown in Figure 1. Her antenatal history was unremarkable, with no history of illicit drug use, smoking, use of unprescribed medications, or chronic health conditions. However, the patient reported a history of miscarriage at 8 weeks of gestation in her preceding pregnancy.

She denied any family history of congenital or genetic abnormalities, and there was no consanguinity in the family. Given the poor prognosis and limited intervention options in advanced gestation, conservative management was planned. At 38 weeks and 4 days gestation, the patient was admitted to our facility in labor and later delivered a live male infant with an Apgar score of 2 at 1 minute and 0 at 5 minutes. The infant expired at the 4th minute despite aggressive neonatal resuscitation. The infant weighed 1,700 grams and was 43 cm in length. On examination, he was unresponsive and displayed multiple congenital anomalies, including partial thoracoabdominal ectopia cordis, a large abdominal wall defect with omphalocele, and gross lower limb deformities, as demonstrated in *Figures* 2,3 and 4. The parents received debriefing and counselling before and after the delivery. Moreover, the couple was advised on the

significance of postpartum genetic testing in identifying potential genetic or syndromic causes, such as the Pentalogy of Cantrell or associated chromosomal abnormalities like trisomy 18.

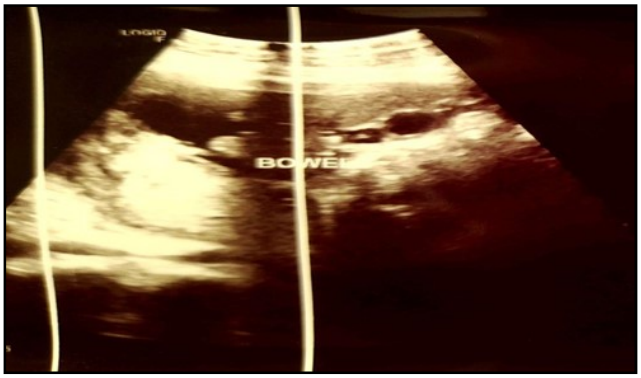

Figure 1. The ultrasound 2D image (a) at 16 weeks of gestation showing the fetal bowels lying outside the abdomen

Figure 2. The photograph of the neonate demonstrates a thoracoabdominal wall defect

Figure 3. The photograph of the neonate demonstrates partial thoracoabdominal ectopia cordis, extruded bowels, and fetal hydrops

Figure 4. The photograph (lateral view) of the neonate demonstrates gross lower limb defects; abdominal organs such as the liver can be appreciated.

DISCUSSION

EC is a rare congenital anomaly reported thousands of years ago. EC describes a condition in which the heart is either partially or completely extruded outside the thoracic cavity (1,3,6). Its etiology has not been fully explained. Failure of lateral mesoderm during the third week of gestation and midline fusion of the developing chest wall due to rupture of the yolk sac has been implicated (7). Despite being a sporadic malformation, EC has been associated with chromosomal abnormalities such as Turner syndrome and trisomy 18 (2).

The clinical management of ectopia cordis begins with a prenatal ultrasonographic diagnosis or fetal echocardiography between 16 and 20 weeks (8). However, in this case, the 20-week scan findings were delayed until 36 weeks due to poor ANC clinic attendance by the patient and failure of an active follow-up, affecting clinical decisions regarding prognosis and treatment. Timely prenatal diagnosis facilitates multidisciplinary planning, parental counselling, surgical intervention evaluation, and perinatal palliative care in scenarios where intervention remains unfeasible (8). Importantly, advanced cases with complications such as fetal hydrops have poor prognosis due to severe physiological compromise, as was evident in this case (10).

Conservative management allowed for prioritizing maternal well-being and minimizing neonatal suffering (11). The patient was counselled on the severity of fetal anomalies, outcomes, and management options, ensuring informed decision-making. Surgical management was unlikely to succeed given the severity of ectopia cordis, fetal hydrops,

and associated anomalies, with high risks and uncertain outcomes (12). Additionally, the lack of fetal specialist input due to delayed diagnosis and resource limitations at the peripheral facility limited a more comprehensive assessment of prognosis and surgical feasibility (13). Early referral to a multidisciplinary team could have facilitated detailed imaging, prognostic counselling, and broader consideration of management options (14). Ectopia cordis's co-occurrence with other abnormalities increases the complexity of care for such cases (5). Approximately 90% of ectopia cordis cases are associated with other anomalies (1,2,5). The prognosis of EC is worsened by its co-occurrence with other anomalies that constitute the Pentalogy of Cantrell. Moreover, the severe oligohydramnios and fetal hydrops increased the complexity of the management of this case. Severe oligohydramnios restricts fetal lung maturation, increasing morbidity (15,16). Inadequate ANC attendance is attributed to poor health outcomes post-delivery for both mothers and neonates (17). This case highlights the significance of adequate ANC attendance, timely diagnosis, and active maternal follow-up in ensuring pregnancy outcomes (18). Prompt identification of such congenital anomalies permits a thorough discussion with parents about options for termination or postnatal care (19,20). Moreover, conservative management prioritizes maternal well-being in poor prognosis, while operative delivery offers limited success and higher maternal risk. Additionally, psychosocial support for the family is critical in resource-limited settings often where the prognosis remains unfavorable (21).

CONCLUSION

This case explores a rare occurrence of ectopia cordis with gross congenital abnormalities complicated by delayed diagnosis, severe oligohydramnios, and fetal hydrops. It uncovers the clinical challenges and complexity of decision-making and emphasizes the significance of early anomaly scanning, accessible maternal-fetal health resources, and a multidisciplinary team. Despite the unfavorable outcome, similar cases show how healthcare systems can optimize prenatal care pathways.

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