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Case Study

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IVEMARK SYNDROME IN A NEONATE- CASE REPORT

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ABSTRACT

Ivemark syndrome is an uncommon congenital disorder which is characterized by involvement of multiple organ systems. Patients with this condition may have cardiac malformations, hypoplasia or aplasia of the spleen and the abnormal anatomy of abdominal and thoracic viscera. It is often associated with severe cardiac abnormalities, which are the usual causes of death in early neonatal life. To our knowledge only a few cases of Ivermark syndrome has been described in Indian literature. Here we report a case of a neonate who was diagnosed as Ivemark syndrome.

KEYWORDS: Ivemark syndrome, Asplenia syndrome, Heterotaxy disorders.

INTRODUCTION

Ivemark syndrome is also called as syndrome of right isomerism or asplenia syndrome. This syndrome occurs when axes of the body fails to rotate correctly in utero. This syndrome is characterized by aplasia or hypoplasia of spleen, abnormal arrangement of the internal organ of the chest & abdomen and congenital heart lesions. Mortality in these patients are very high due to severe cyanotic cardiac defects compounded by absent spleen resulting in fatal infections from encapsulated organisms such as Meningococcus, Haemophilus influenza and Streptococcus pneumonia.

CASE REPORT

A preterm (32 weeks period of gestation), 1370 gm female baby was born to a 23 years old, third gravida mother by vaginal delivery. There was history of polyhydraminos in mother and antenatal steroid cover was incomplete. Antenatal history did not reveal any drug intake or radiation exposure in mother. On admission in neonatal intensive care unit, baby heart rate was 150/min, spO2 96%, RR 75/min and Silverman Anderson score was 5/10. Cardiac examination revealed pansystolic grade III murmur. On abdominal examination liver was palpable continuously on left side. Patient was given surfactant at second hour of life. In view of severe respiratory distress baby was mechanically ventilated. Chest radiograph with abdomen revealed a midline cardiac shadow and right sided gastric fundal shadow (Figure 1).

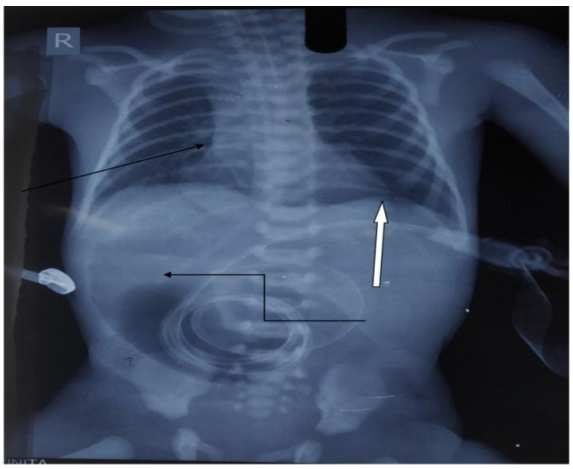

Figure 1: Chest & abdomen radiograph of the neonate showing mesocardia (black straight arrow), gastric fundal shadow with orogastric tube visible on right side (black curved arrow) and liver shadow on left side (white arrow).

Ultrasound abdomen was done which revealed liver on left side with left lobe of liver on right side and spleen was not visualized (Figure 2).

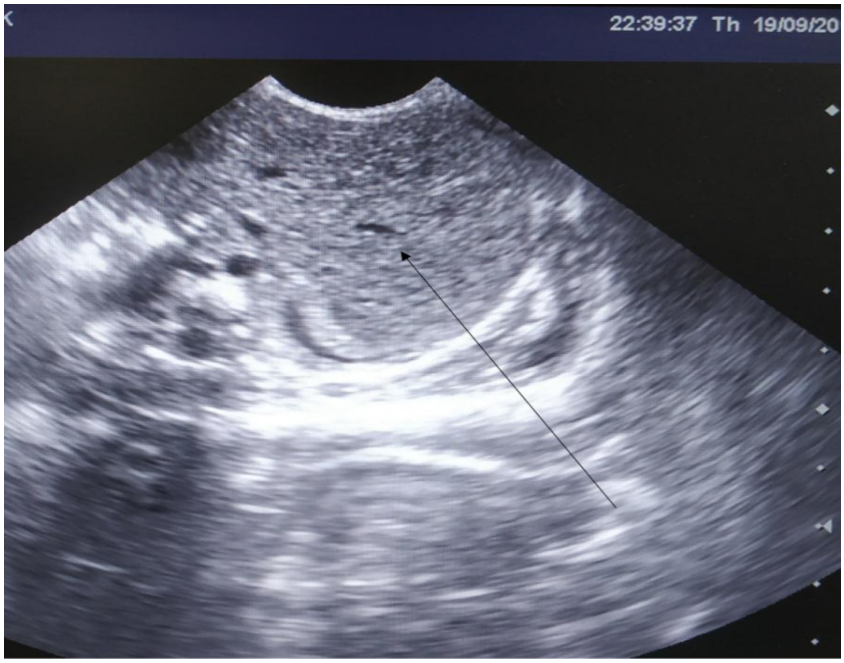

Figure 2: USG abdomen showing liver on left side (black arrow).

Echocardiogram showed mesocardia, atrial septal defect, a ventriculat septal defect and atrioventricular canal defect (Figure 3).

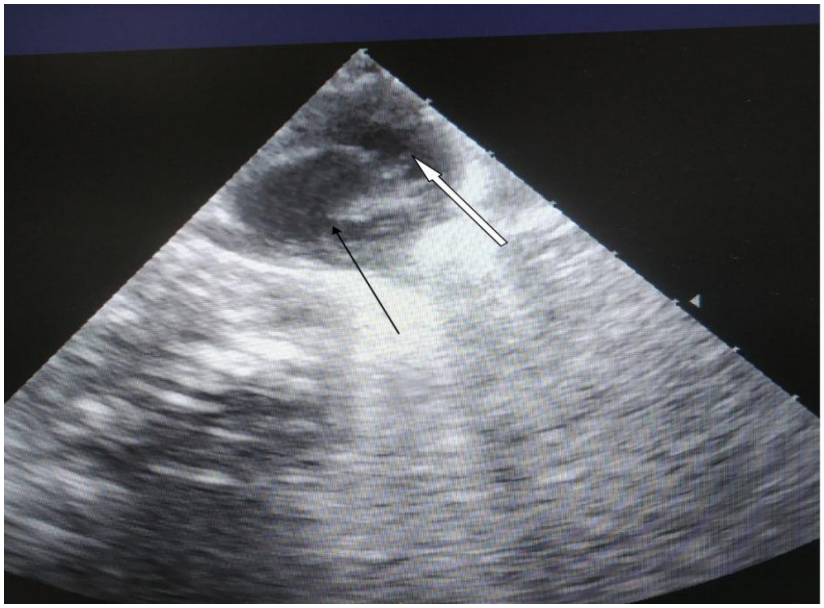

Figure 3: Echocardiogram showing atrial septal defect (black arrow) and ventricular septal defect (white arrow).

A diagnosis of Ivemark syndrome was made. Poor prognosis of the neonate and risks of surgical management was explained to the parents who refused further intervention. Despite our repeated counseling, parents took their child home against medical advice after giving written consent. The baby expired on day 4 of life.

DISCUSSION

Ivemark syndrome/Asplenia syndrome is a rare disorder that affects multiple organ systems of the body. Ivemark syndrome is classified as a heterotaxy disorder. Heterotaxy refer to the failure of the internal organs of the chest and abdomen to be arranged in the proper location within the body.[1] The exact etiology of Ivemark syndrome is unknown. Most cases occur randomly with unknown causes(sporadic cases). Ivemark syndrome has been reported in multiple members of the same family suggesting that a genetic predisposition may have been a factor in the development the disorder in these cases.[2] The exact incidence of this rare syndrome is unknown. The incidence of laterality disorders taken

together is estimated to be 1 in 15,000 children.[3] The symptoms of Ivemark syndrome are due to the abnormal arrangement and malformation of certain organs specially congenital heart defects. Heart defects commonly associated with Ivemark syndrome include total anomalous pulmonary venous connection, double outlet right ventricle, transposition of the great vessels, ventricular or atrial septal defects and atrioventricular canal defect.[4] In the index case, mesocardia and atrioventricular septal defect was present. Other associated conditions include intestinal malrotation, biliary atresia, splenic abnormalities, faulty gastric suspension mechanisms, displacement of abdominal viscera, and aberrant vascular structures and vascular connections.[5] In the index case there was absent spleen and left sided liver. Absence of spleen in these individuals predisposes them to repeated infections especially by capsulated organisms. Treatment of Ivemark syndrome require multidisciplinary approach with coordinated efforts of a team of specialists such as pediatricians, surgeons, pediatric cardiologists, pediatric gastroenterologists, pulmonologists, neurologists, immunologists and other healthcare professionals.

CONCLUSION

Ivemark syndrome is associated with high mortatity rate due congenital cardiac defects. Any newborn that has characteristic features of this syndrome, we should use various imaging modalities to reveal the anatomical features in these patients because being aware of them prior to surgery and invasive intervention prevents the possible risks and complications.

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