Unveiling the Mysteries: Exploring a Case of Scimitar Syndrome

Fonseca R1, Nunes R1, Reis-Santos M1, Araújo A1, Ferreira JC1, Loureiro T1
1 Department of Obstetrics, Gynecology and Reproductive Medicine, Local Health Unit of Santa Maria;
2 Department of Pediatric Cardiology, Local Health Unit of Santa Maria;

INTRODUCTION

Scimitar syndrome is a rare and complex congenital heart defect. The main feature is an anomalous vein draining from the right lung into the systemic venous circulation, usually the inferior vena cava. Associated features include hypoplasia of the right lung, secondary dextroposition of the heart, anomalous systemic arterial supply to the right lung and other cardiac abnormalities. This report aims to address important aspects of the prenatal diagnosis of Scimitar Syndrome, emphasizing the significant complexity of its diagnosis.

METHODS


CASE REPORT

A 35-year-old pregnant woman was referred to our prenatal diagnostic unit due to suspicion of cardiomegaly and deviation of the cardiac axis detected during routine ultrasound in the second trimester. The patient had no significant personal history or family history of congenital anomalies or genetic disorders. The pregnancy progressed without complications. Serial fetal echocardiograms were performed, which did not demonstrate the previously observed alterations and were found to be normal, identifying two pulmonary veins draining directly into the left atrium.

Spontaneous premature labor occurred at 34 weeks and 6 days of gestation, and the delivery was by urgent c-section due to breech presentation. The male newborn weighed 2005 grams with an Apgar score of 7/8/8. During the first hour of life, the newborn developed a progressive respiratory distress requiring endotracheal intubation and was transferred to the neonatal intensive care unit. The postnatal echocardiogram, performed on the 1st day of life, detected dextrocardia with levoapex and structurally normal heart with apparently normal pulmonary venous return. Chest X-rays showed opacity in the right hemithorax. Due to persistent respiratory distress, a chest CT-scan was performed, providing evidence of Scimitar Syndrome.

The newborn was discharged 20 days after birth, clinically stable. Since then, the patient has been followed at the pediatric cardiology clinic, maintaining normal peripheral oxygen saturation and no signs suggestive of heart failure or pulmonary hypertension.

CONCLUSION

Scimitar Syndrome is often underdiagnosed prenatally. The occurrence of dextrocardia in the prenatal period should raise the suspicion when no external cardiac compression is seen, as it may indicate right lung hypoplasia. However, the definitive diagnosis remains difficult due to the significant challenge in visualizing the anomalous vein. Still, its consideration in the differential diagnosis may contribute to the stabilization and recovery of the neonate in the cases where early respiratory distress occurs.