Early Intrauterine Diagnosis of Scimitar Syndrome and Postnatal Diagnosis of Pulmonary Sequestration in a Private Maternity In Salvador-Ba/Brazil: A Case Report

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Abstract:

Scimitar Syndrome (SS) is a rare congenital cardiopulmonary malformation most defined by partial anomalous pulmonary venous return (PAPVR) from the right lung into the inferior vena cava. We report a case where fetal echocardiogram raised suspicion of SS at 27 weeks and confirmed the diagnosis at 31 weeks of gestation, demonstrating non-obstructive partial anomalous drainage of the right pulmonary veins and right pulmonary artery hypoplasia, without classical signs of rightward mediastinal shift or cardiac dextroposition. The fetus exhibited situs solitus with mesocardia, underscoring the diagnostic challenge. A male infant was delivered at 36+5 weeks by cesarean section. Postnatal pediatric echocardiography and computed tomography angiography confirmed SS and identified associated pulmonary sequestration (PS), a frequent comorbity contributing to left-to-right shunting through anomalous systemic arterial supply. The patient underwent successful transcatheter embolization of the sequestered lung segment and was scheduled for elective surgical repair of the SS. This case highlights the importance of detailed fetal cardiac assessment for early SS diagnosis even when classical ultrasonographic signs are absent. It challenges current fetal echocardiography paradigms and emphasizes the role of multimodal postnatal imaging in diagnosis confirmation and therapeutic planning.

Keywords:

Scimitar syndrome, intrauterine diagnosis, pre-natal diagnosis, echocardiogram findings, pulmonary sequestration.