ORIGINAL RESEARCH PAPER

Neonatology

SCIMITAR SYNDROME: RARE CASE REPORT OF NEONATAL SCIMITAR SYNDROME IN MONOCHORIONIC MONOAMNIOTIC TWIN

KEY WORDS:

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ABSTRACT

Scimitar syndrome is a rare congenital heart disease characterised by anomalies that include hypoplasia of the right lung, total or partial anomalous pulmonary venous return of the right lung, dextroposition of the heart, right pulmonary artery hypoplasia(1). Curvilinear pattern visible on chest radiography resembling a curved Turkish sword (scimitar) is created due to abnormal drainage of the pulmonary veins into the inferior vena cava instead of the heart(2). Here we report a case of neonatal Scimitar syndrome in a monochorionic monoamniotic twin gestation with significant clinical improvement after successful coil embolization therapy. Incidence ranges from 1 to 3 per 100,000(3), true incidence may be higher as many patients are asymptomatic.

CASE:

A late preterm (35weeks) female child delivered by caesarean section to para2 mother with monoamniotic monochorionic twin gestation, with an uneventful antenatal history.ANC ultrasound scans of both twins were normal. Twin A weighed 2.49 kg at birth was given normal new born care while Twin B had a birth weight of 2.25kg was shifted to NICU soon after birth in view of moderate respiratory distress.

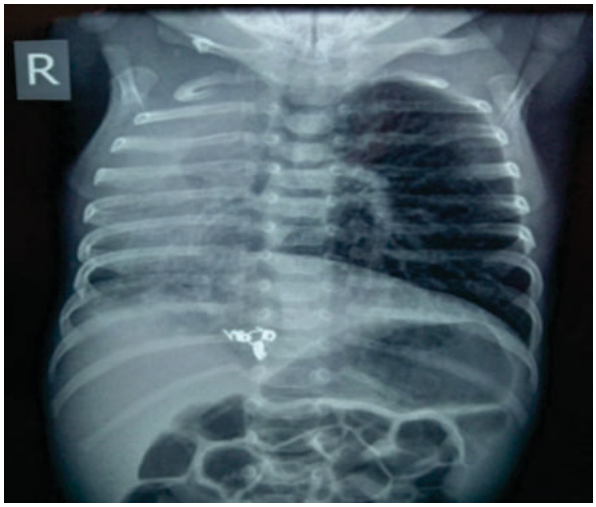
Course in NICU - Physical examination revealed a respiratory rate of 70/min, heart rate of 160/min, peripheral pulses well felt with normal capillary refill time. Respiratory system examination revealed decreased air entry over right side of chest. Cardiovascular system examination revealed shifting of apex beat to right side with ejection systolic murmur of Grade II/VI over the left parasternal border with normal S1 and loud S2 on auscultation. Neonate was kept on respiratory support in the form of oxygen by bubble CPAP.

Other system examination was unremarkable.

Routine lab investigations were within normal limit except CRP which was found to be positive.

USG abdomen and skull had no detected abnormality.

Chest X-ray showed radiopaque right hemithorax suggestive of atelectasis of right lung with mediastinal shift to the right and dextroposition of the heart.



Echocardiography was done on Day 5 of life which showed small ASD with left to right shunt and PDA, right sided pulmonary veins draining into inferior vena cava (PAPVC), left pulmonary veins draining into left atrium, hypoplastic right pulmonary artery with severe pulmonary hypertension.

CT Angiography was needed in order to confirm the diagnosis. It revealed obliteration of the right bronchus with right lung consolidation and volume loss, infra-cardiac PAPVC of right lung draining into hepatic inferior vena cava, arterial supply to right lower lobe from aorta suggests sequestration. Large PDA, hypoplastic right pulmonary artery and dilated main and left pulmonary artery, aberrant right subclavian artery with no tracheal narrowing.

Hypogenetic Right Lung With Mediastinal Shift To Right

Baby was treated with furosemide, sildenafil and lasilactone and given respiratory support in the form of oxygen by bubble CPAP. Baby also had CRP positive sepsis for which baby was treated and after medical stabilization was shifted for surgery.

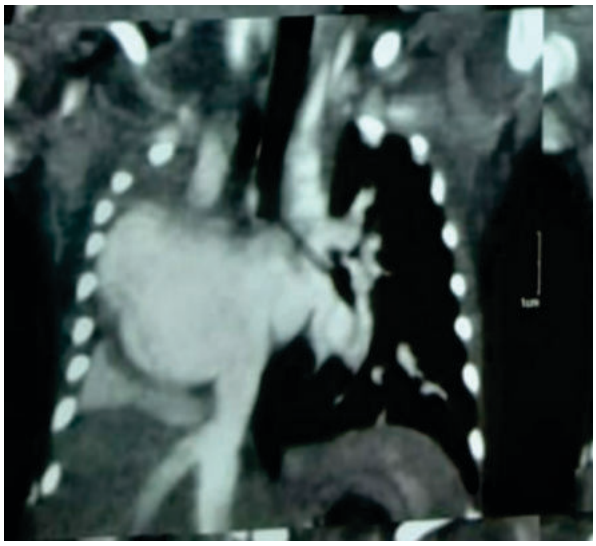
On Day 42 of life, after elective intubation and ventilation, coil embolization of multiple collaterals from aorta to right lower lobe of lung was done. The procedure was performed successfully without intraoperative or postoperative complications. Baby was shifted post procedure to the PICU and was started on sildenafil infusion which was continued and tapered over the next two days. Feeds were started with supportive management and the baby was continued to be monitored. Follow up echocardiography was done post procedure which was found to be normal. Pulmonary hypertension improved after taking sildenafil.The baby started taking feeds well and improve post procedure. Baby was discharged on oral sildenafil, furoped and Aldactone.

DISCUSSION:

Scimitar syndrome has a varied presentation from asymptomatic to severe respiratory distress due to right lung hypoplasia, pulmonary hypertension, left to right shunt, atrial or ventricular septal defect and coarctation of aorta(3). Infantile form is almost always symptomatic and needs surgical correction. Embolization of anomalous systemic arteries have been performed using detachable balloons, coils, or tissue adhesive. Coil occlusion of anomalous systemic arteries can improve symptoms of heart failure and pulmonary hypertension in infants and may bring about good surgical results(4). Course of infantile form is often complicated by severe pulmonary hypertension and cardiac

failure leading to poor outcome(5) Main pathophysiological mechanism is abnormal return of one or more pulmonary vein(s) leading to development of left to right shunt. This shunt eventually causes volume overload on right sided heart causing heart failure.

This is the first reported case of scimitar syndrome presented at birth in monochorionic monoamniotic twin and underwent successful coil embolization and discharged home with an uneventful follow up. One case of neonatal Scimitar syndrome is reported in dichorionic diamniotic twin(6).



CONCLUSION:

Although Scimitar syndrome is now more widely recognised, it needs a high index of suspicion in neonate with respiratory distress with shift of apex beat and typical chest radiograph findings even if the neonate has other cardiac lesions sufficient to explain the signs and symptoms of heart failure. Classic curvilinear scimitar vein may or may not be apparent on chest radiograph. Mortality remains high even with surgical intervention, especially in those with cardiac malformations. Early recognition and treatment are keys to survival. Echocardiography, CT angiography and cardiac catheterization is necessary for confirmation of diagnosis(7). Once confirmed definitive medical and surgical intervention is needed before development of irreversible pulmonary hypertension.

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