Case Report

Rare cause of pulmonary hypertension and elevated lactate in a neonate: Scimitar Syndrome- Case Report

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ABSTRACT

Scimitar syndrome, a rare congenital anomaly with an incidence of 1-3/100,000 live births, is commonly associated with hypoplasia of the right lung, pulmonary sequestration, and persisting superior vena cava (SVC). The other features are a partial anomalous pulmonary venous connection (PAPVC) with a curvilinear pattern on chest radiograph, with the pulmonary veins draining to the Inferior Vena Cava (IVC). This case report presents a two-week-old female, presented with escalating respiratory distress. Serum lactate gradually increased despite sildenafil and diuretics. Scimitar syndrome with large aortopulmonary collaterals from the celiac trunk was confirmed by contrast CT. Multiple abnormal systemic arteries (4-6 mm in diameter) were discovered during a one-month cardiac catheterization. Transcatheter embolization with Piccolo coils, PDA occlusion devices, and microvascular plugs worked well. Transcatheter embolization of aortopulmonary collateral vessels in neonatal Scimitar syndrome leads to a rapid resolution of high-output cardiac failure and pulmonary hypertension. Transient persistent hyperlactemia (resolving by 6 months of age) is likely due to ischemic changes in the embolized lung. Neonates with hyperlactemia should be screened for Scimitar syndrome.

Key words: Scimitar syndrome, neonatal pulmonary hypertension, transcatheter embolization, hyperlactemia, aortopulmonary collaterals.

Scimitar syndrome, a rare congenital anomaly with an incidence of 1-3/100,000 live births, is commonly associated with hypoplasia of the right lung, pulmonary sequestration, and persisting superior vena cava (SVC) [1, 2]. The other features are partial anomalous pulmonary venous connection (PAPVC) with a curvilinear pattern on chest radiograph, by the pulmonary veins that drain to the Inferior Vena Cava (IVC) [3]. Scimitar syndrome may be present in the neonatal period with heart failure, pulmonary hypertension, recurrent respiratory infections, or may remain asymptomatic till adulthood, presenting mild exertional dyspnea [3,4]. This case report presents the clinical course, diagnostic evaluation, and interventional management of a neonate with Scimitar syndrome presenting as severe pulmonary hypertension and persistent hyperlactatemia.

CASE PRESENTATION

A two-week-old female, born via elective cesarean section with a birth weight of 2248 gm, was presented with a history of second-degree consanguinity with an elder sibling with Williams Syndrome. The antenatal period was uneventful. She developed respiratory distress six hours after birth and was immediately shifted to the neonatal intensive care unit (NICU), initially supported with high-flow nasal cannula oxygen along with intravenous (IV) fluid and antibiotics; however, she required mechanical ventilation on day 3 because of increasing respiratory distress and elevated CO2. The routine laboratory investigations were normal.

Echocardiogram (ECG) on day 3 of life showed a dilated right atrium (RA), a patent ductus arteriosus (PDA), and a patent foramen ovale (PFO with a bidirectional shunt), and severe pulmonary hypertension (PHTN) with suprasystemic right ventricular (RV) pressure (peak gradient: 70 mm-Hg). PHTN was managed with diuretics and sildenafil 0.5 mg 6 hourly. The elevated pulmonary pressure responded partially to sildenafil over the next 2 weeks. However, the lactate slowly increased, ranging from 4 to 5.6 mmol/L.

The ventilation continued with minimal pressure settings, and she had normal Blood Pressure (BP) and heart rate, without inotropes. After one week of ventilation, a repeat chest X-ray was clear, and viral and bacterial cultures of endotracheal (ET) tube secretions were negative. On day 13, she was extubated to nasal Continuous Positive Airway Pressure (CPAP); however, she developed respiratory distress

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and was reintubated. The lactate levels progressively increased without metabolic acidosis.

After admission, the baby commenced on a ventilator with normal BP and heart. IV fluids, antibiotics, diuretics, and sildenafil were continued. Repeated investigations, including hemogram, RBS, serum electrolytes, Renal Function Tests (RFTs), and liver function tests (LFTs), were normal. Ultrasound of the chest and abdomen showed no fluid accumulation. Blood gas analysis showed a normal anion gap and no metabolic acidosis. Serum lactate was high (4.5 mmol/L). The sepsis screens (C-Reactive Protein, blood, urine, and ET cultures) were negative, and serum ammonia levels were normal; inborn errors of metabolism screening were negative.

Repeat ECG showed 3 pulmonary veins returning to the left atrium (LA), and the 4th pulmonary vein was not visualized. There was an atrial septal defect (ASD) (7 mm), left SVC draining to a dilated coronary sinus, dilated RA, and RV systolic pressure at 2/3 systemic pressure. Contrast Computer Tomography (CT) of the chest showed right lung hypoplasia, the right lower pulmonary vein draining directly into the IVC, and the right lower lobe of the lung receiving anomalous systemic arterial supply from the coeliac trunk. This confirmed the diagnosis of Scimitar syndrome.

At one month of age, she underwent cardiac catheterization, showing multiple massive collaterals from the abdominal aorta to the right lung with pulmonary venous return of the lower right lobe of the lung to the IVC and significant cardiomegaly. The massive collaterals were occluded using multiple microvascular plugs, PDA occlusion devices, and Piccolo devices. Within 4 days, lactate began to decrease with improvement in pulmonary pressure and right heart dilatation. She gradually weaned off the respiratory support for 5 weeks of life.

The baby was discharged at 2 months of age, weighing 2.625 kg, active, and accepting feeds well. She was on spironolactone, captopril, and hydrochlorothiazide. There was a plan for surgical correction after gaining weight. The parents were advised to monitor weight gain, clinical status, and lactate levels. Follow-up showed grumbling lactates, which normalized after 6 months, possibly reflecting chronic ischemic changes of embolized lung tissue. Cardiac catheterization showed a growing Right Pulmonary Artery (RPA). At 3 years of age, insignificant cardiology findings, but she had poor growth and weight gain, and chronic kidney disease stage 1 (Glomerular Filtration Rate (GFR) 104 ml/min/1.73m2) was diagnosed, and primary hyperoxaluria (AGXT mutation) for which she was on Lumasiran maintenance therapy. Written consent was obtained from the parents for the publication of this case report.

DISCUSSION

Scimitar syndrome is a rare congenital cardiac anomaly associated with 3-6% of PAPVC cases. PAPVC, this anomalous vein returns to the hepatic portion of the IVC, but may also return to the portal vein, hepatic vein, or right atrium [5]. The other associated cardiac conditions are ASD, tetralogy of Fallot, left-sided lesions like aortic arch hypoplasia, and hypoplastic left heart syndrome [6]. Three-dimensional (3-D) CT and cardiac-gated magnetic resonance imaging (MRI) can visualize the anomalous pulmonary vein [7]. Gadolinium-enhanced 3-D MR angiography provides concurrent anatomical and functional diagnosis [8].

Fetal ECG permits prenatal diagnosis, and color Doppler helps to locate the obstructed pulmonary venous pathway by showing confluence behind the RA and vertical vein [9]. PAPVC shows hemodynamic changes like ASD, and symptomatic patients commonly present with signs of heart failure and pulmonary hypertension [10]. The present case had signs of high-output cardiac failure and Aortopulmonary (AP) collaterals. The lactate levels transiently decreased after occlusion of the AP collaterals but increased again. High lactate can be a manifestation of Scimitar syndrome per se or due to lactate leakage from lung tissue following occlusion of the aberrant systemic circulation. However, this assumption needs confirmation in future studies.

In neonates, a trial of medical therapy allows for growth before repair of the defect. However, pulmonary hypertension or lack of response to medical therapy demands prompt surgical management [11].

CONCLUSION

This case shows that transcatheter embolization of AP collaterals can be performed safely in neonates with Scimitar syndrome, to improve the symptoms of heart failure and pulmonary hypertension, potentially allowing the growth of RPA. However, lactates can remain high for a long time.

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