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Case Report

Right pulmonary artery agenesis with transient hypothyroidism in a newborn

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ABSTRACT

Unilateral absence of pulmonary artery (UAPA) is a rare congenital anomaly that can present as an isolated lesion or may be associated with other Congenital heart diseases. It is usually detected in infancy or incidentally found on X ray chest in adulthood. The diagnosis of UAPA has rarely been made in a newborn. Only one case has been reported in newborn period from Belgium by F. Marchau who reported the rare occurrence of this anomaly in a 2-day old male baby. We report the rare occurrence of UAPA in a newborn female from India who presented with severe respiratory distress at birth with high cord blood TSH levels. Our case also reflects the importance of a high index of suspicion along with the value of X-ray chest and echocardiogram in the neonatal period in detection of this rare anomaly.

Keywords: Newborn, Pulmonary artery agenesis, Transient hypothyroidism

INTRODUCTION

Congenital "unilateral absence of a pulmonary artery" is a rare malformation. The first case of UAPA was reported by Fraentzel in 1868.[1] "Absent'' right pulmonary artery is mostly found as an isolated lesion. In contrast, 75% of patients with "absent" left pulmonary artery have associated congenital cardiac anomalies.[2] The most frequent accompanying cardiovascular anomalies include Tetralogy of Fallot or septal defects. Other congenital cardiac defects associated with UAPA are: coarctation of the aorta (either isolated or in combination with a ventricular septal defect), subvalvular aortic stenosis, transposition of the great arteries (either isolated or in combination with ventricular septal defect or pulmonary stenosis), Taussig-Bing malformation and coarctation, congenitally corrected transposition and pulmonary stenosis, and Scimitar syndrome.[3] Aortopulmonary window. Patients with isolated unilateral ''absence'' of a pulmonary artery are mostly asymptomatic, this being an incidental finding, e.g. on a chest X-ray film.[4-8] Some patients develop recurrent respiratory infections, dyspnea, exercise intolerance, hemoptysis and pulmonary hypertension in the contralateral pulmonary artery.[9]

Transient Hypothyroidism in a new born is either due to transplacentally acquired TSH binding inhibitory immunoglobulins, maternal exposure to goitrogens (iodides or anti-thyroid drugs), transient hypothyroxinemia of Prematurity or sick euthyroid syndrome. As far as congenital hypothyroidism is concerned, the worldwide incidence is 1:3000-4000 live births and the estimated incidence in India is 1:2500-2800 live births.

Only few cases of unilateral pulmonary artery agenesis have been reported from India.[4,10-13] We report for the first time from India the case of a new born baby with

Absent Right Pulmonary Artery and Bronchiectasis of Right Lower Lobe with Transient Hypothyroidism.

CASE REPORT

A Female baby, 2nd in birth order, 37 weeks+5days by gestational age, was born by LSCS. The indication of LSCS was Previous LSCS with impending rupture. There was H/O nephrolithiasis and UTI in mother detected few days prior to delivery (with heavy growth of Staphylococcus aureus in Urine culture) for which she had received intravenous antibiotics. The mother had subclinical hyperthyroidism which was detected in the last trimester. She was on Propylthiouracil 100ug Once daily orally, started one week prior to delivery. Her anti-TPO antibodies were negative, TSH was low with a value 0.038uIU/ml, FT3 and FT4 were normal (3.15pg/ml and 1.15ng/dl respectively). There was a history of thyroid disturbance in previous pregnancy as well but the exact nature was neither known to mother nor were any previous records available. The mother was not fully evaluated in the pre-pregnancy period during the present gestation.

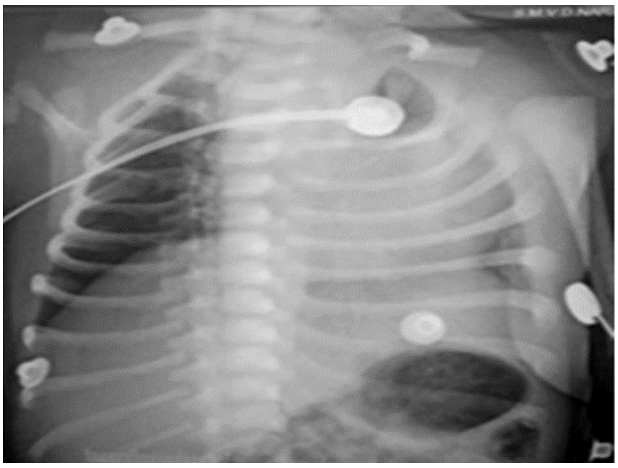

Figure 1: Chest x-ray showing oligemic right lung, elevated hemidiaphragm, hyperinflated and hyperlucent left lung.

Birth weight of the baby was 3.52 kg with 1 min and 5 min APGAR score of 8/10 and 8/10. The baby had grunting, respiratory distress with a Silverman's score of 6/10. On examination the chest was clear, heart sounds were normal with no murmurs, no choanal atresia or any other apparent congenital anomaly was observed. The baby had a saturation of 70-75% on head box. Samples were drawn for sepsis screen and X ray chest was obtained. The initial and repeat sepsis screen at 12 hrs were both negative. X ray chest was suggestive of contracted right hemi thorax, oligemic right lung, mild elevation of the right hemi diaphragm and a hyper-lucent and hyperinflated left lung. Echocardiography was done which showed Absent right pulmonary artery, small PDA with L to R shunt with peak systolic gradient of 10mm Hg, High flow in left lung, severe PAH, PFO with L to R shunt. CT angio-chest was done to delineate branch pulmonary anatomy and it was suggestive of Absent/Aplasia - Right pulmonary artery, Multiple collaterals supplying the right lung from ascending and descending aorta, subclavian arteries and bronchial arteries, Aorto -left atrial shunt, Oligemic right lung with cystic changes in anterobasal segment of right lower lobe. The baby was managed conservatively with supportive treatment that included CPAP ventilation at 5 cms of water pressure, intravenous fluids, intravenous antibiotics and diuretics. Her clinical status improved and she was roomed in with the mother on Day 8. Her cord blood TSH was >150uIU/ml, repeat TSH done on Day 4 and Day 7 were 19.05 and 23.83 respectively. X-ray Knee confirmed the presence of both ossification centres and USG neck showed normal size and normal echo pattern of both lobes of thyroid gland. She was discharged on day 10. Follow up TSH at 2 weeks was 4.382 uIU/ml. The baby is on regular follow up and doing well. A repeat echo has been planned at 3 months of age.

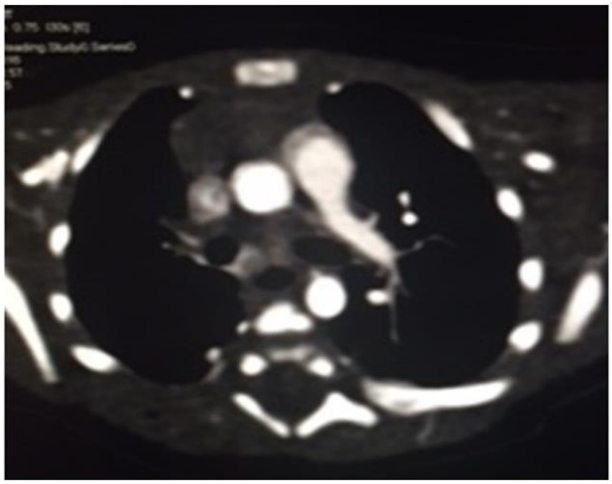

Figure 2: Computed Tomogram showing absent right pulmonary artery and low vascularity in the right lung.

DISCUSSION

Unilateral absence of pulmonary artery is a rare disease. This developmental anomaly is the result of a failure in the connection of the sixth aortic arch with the pulmonary trunk. The affected lung is perfused by persisting embryonic arteries from the aorta or its branches, abnormal collaterals arising from the bronchial, subclavian, intercostal or diaphragmatic arteries, or hyperplastic normal bronchial arteries. Bouros et al. described the prevalence by analysing the chest radiograph of 600,000 army recruits and found a prevalence of 1 in 200,000.[2] This study was carried out in the healthy, adult population. The true prevalence of the disease in children is not known.

The entity of unilateral absence of pulmonary artery was reviewed in four publications covering different time era

in the last ~60 years. Pool et al reviewed 32 cases before 1962 whereas Shakibi et al studied 47 cases from 1962 to 1976.[5,7] Harkelet al added a review of 107 cases from 1978 to 2000.[8] RPA was absent in 56-63% of the cases in these three case reviews. Singhi et al reported Absent RPA in 5 infants with a median age of 80 days (range 40-120 days).[11] Cucciet al opined that absence of RPA was more frequently seen in an otherwise normal heart and absence of left pulmonary artery was accompanied by additional cardiac lesions.[14] F. Marchau et al reported the rare occurrence of this anomaly in a 2-day old male baby.[9]

Two types of presentations are described. The first presentation is the one seen in infants, where they usually present with congestive cardiac failure and pulmonary hypertension (PHT). The other presentation is in older patients, who usually do not develop pulmonary hypertension and do not have manifest heart failure. They present with exercise intolerance (18-40%), haemoptysis (20%) or are incidentally detected during chest radiography.[8,15] Our patient falls in the first category.

The incidence of PHT among the patients with unilateral absence of pulmonary artery is reported to be between 19 and 44% in different case series.[6,8,9] PAH may result from blood flow directed solely towards the contralateral PA. Increased blood flow in the contralateral PA leads to shear stress within the endothelium, with subsequent release of vaso-constrictive compounds, such as endothelin. Chronic vasoconstriction of the pulmonary arterioles may lead to remodelling, resulting in increased resistance of the pulmonary vasculature and PHT.

The diagnosis of isolated absence of RPA is based on history, clinical evaluation and imaging. The diagnosis can easily be missed in infancy and a high index of suspicion is needed.[6-8] Chest X-ray may show an absence of right PA, reduction of pulmonary vascular markings on the right side (~60% cases), a small hemi thorax and reduced intercostal bone space, shrunken right lung and a shift of the mediastinal structures to the affected side with contralateral lung hyperinfation. Though the initial suspicion in our case was Respiratory Distress Syndrome (RDS) due to congenital pneumonia, meconium aspiration syndrome or transient tachypnoea of newborn but the differential vascularity and a small hemi thorax on X ray chest played a key role in suspecting this entity which was confirmed by echocardiography and CT angiogram.

A cardiac MDCT scan and magnetic resonance imaging (MRI) can confirm the diagnosis and delineate the pulmonary artery anatomy. Any infant with unexplained Pulmonary artery hypertension should be thoroughly evaluated for the possibility of isolated unilateral absence of pulmonary artery (UAPA). Differential vascularity on the chest X-ray is a good clue for the diagnosis.

High cord blood TSH could be explained by history of ingestion of anti-thyroid drugs in the mother supported by the presence of both the ossification centres on X-ray knee and a normal thyroid gland on USG. Infants with transient hypothyroidism due to maternal goitrogenic drugs need not be treated unless they have low T4 and elevated TSH values persisting beyond 2 weeks. The supplemental therapy can be discontinued after 8-12 weeks. Intake of anti - thyroid drugs can be continued by the hyperthyroid mothers during breast feeding as concentration of these drugs is very low in breast milk. But the need of anti- thyroid drugs in mother in our case is questionable as the mother had sub-clinical hyperthyroidism and needed further evaluation.

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