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Case Report

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Unilateral Absence of Left Pulmonary Artery, Left Main Bronchus and Left Lung

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Abstract

Our patient is a 2 year old girl, born through in vitro fertilization (IVF), who immediately after birth experienced episodes of desaturation. Screening transthoracic echocardiography was done urgently and showed absent left pulmonary artery (LPA). A computed tomography (**CT**) scan and angiography of the chest confirmed the absence of LPA, in addition to the absence of the left main bronchus and left lung agenesis.

Keywords: Left pulmonary artery; Lung agenesis; Left bronchus

Introduction

Absent LPA is a known entity associated with absent pulmonary valve syndrome, and at times with RVOT (Right Ventricle Outflow Tract) obstruction [1]. It is frequently referred to as the "Tetralogy-absent pulmonary valve syndrome". It has also been reported as an isolated anomaly; however the left lung and bronchus were present usually.

We are unaware of any reported case of an absent left pulmonary artery with complete absence of the left main bronchus and left lung.

Case Report

A 2 year old girl, product of a twin IVF pregnancy, was delivered by cesarean section; her birth weight was 2.5 kg. After birth, she was admitted to the neonatal intensive care unit because of desaturation and respiratory distress. She was intubated for several days, extubated successfully to oxygen via nasal cannula, finally weaned off O_2 and discharged home on room air with O_2 saturation above 92%.

This patient has dysmorphic features including an extra digit on the left hand, fusion of the first and second left ribs, and bifid T7 vertebra discovered on chest x-ray and confirmed by CT scan of the chest.

Immediately after birth she had desaturation but no murmur on clinical exam. A screening transthoracic echocardiography showed absent left pulmonary artery with no associated intracardiac anomalies and right aortic arch. A CT scan of the chest was done and confirmed the absence of the left pulmonary artery, left main bronchus and the left lung (Figures 1 and 2).

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The patient was evaluated by our genetic service who found that the patient does not have a specific genetic syndrome. Chromosomal analysis was normal.

The patient now has normal oxygen saturation and is in regular follow-up with the pulmonologist due to intermittent mild asthma.

The other twin is a boy who has no dysmorphism, however, a transthoracic echocardiography showed a small secondum atrial septal defect and a small ventricular septal defect.

Discussion

Although not common, absent or isolated LPA is a well known entity. It has been associated with "absent pulmonary valve syndrome" [2] where the LPA is isolated and supplied by a ductus or collaterals. In some reported cases it was found to be confluent but very hypoplastic [3]. Absent unilateral pulmonary arteries, but associated with collaterals to the same side lung, has been described in medical literature [4]. As well, there is a reported case of a child with a right aortic arch and right ligamentum arteriosus presenting with respiratory symptoms due to vascular ring and found to have absent left pulmonary artery [5]. Farghly et al. reported absent LPA in a patient presented with hemoptysis due to bleeding from peribronchial vessels [6].

We found it unusual to have a combination of an absent LPA, left main bronchus, and left lung in the absence of other cardiac

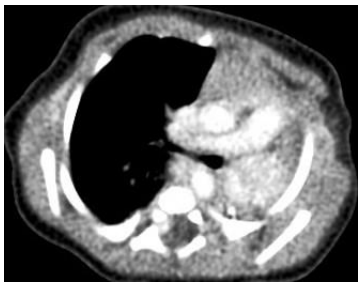

Figure 1: Contrast enhanced cardiac CT showing main and right pulmonary arteries and absent LPA, Left main bronchus and left lung.

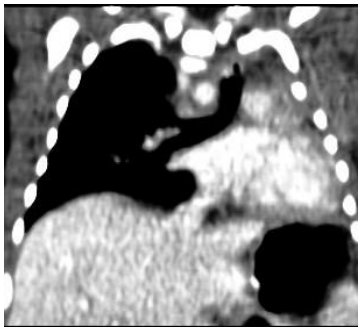

Figure 2: Coronal contrast enhanced cardiac CT showing absent left main bronchus and left lung.

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anomalies except the presence of a right aortic arch. To the best of our knowledge this is the first reported case of this condition.

Conclusion

Based on our literature search and our clinical experience, this is the first reported case of a unilateral combined absence of LPA, left main bronchus and left lung in association with right aortic arch in the absence of other cardiac anomalies.

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