Case Report

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Gynecology & Obstetrics Case report
ISSN 2471-8165

2016
Vol.2 No.1:14

Prenatal Diagnosis of Right Pulmonary Agenesis with Left Pulmonary Artery Sling and Total Anomalous Venous Return

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Received date: Feb 29, 2016; **Accepted date:** Mar 14, 2016; **Published date:** Mar 17, 2016

Abstract

Right pulmonary agenesis (PA) with left pulmonary artery sling (LPAS) is a very rare vascular anomaly commonly associated with tracheobronchial anomalies. It is also frequently linked with cardiac and extra cardiac defects. Associated major cardiac and tracheobronchial anomalies determine patients' surgical prognosis. Fetuses with PA and LPAS prenatal diagnosis require echocardiograhic screening to rule out intra and extra cardiac lesions which worsen prognosis. We report a case of prenatal diagnosis of right pulmonary agenesis with LPAS and total pulmonary venous return. To our knowledge, this malformation has never been reported as an associated cardiac defect.

Keywords: Foetus; Pulmonary agenesis; Left pulmonary artery sling; Total anomaly of pulmonary venus return

Case Report

A 32 year-old woman, gravida 3, para 2 was referred for fetal echocardiographic investigation of dextrocardia at 23 weeks' of gestation. No extrathoracic anomalies were observed except for bilateral preauricular tags.

Cardiac ultrasound examination (ALOKA alpha 10 echograph) demonstrated that dextrocardia was caused by right pulmonary agenesis (PA). There was no left pulmonary artery (LPA) in the expected location (**Figure 1a**).

A small pulmonary artery araised from the right side of the pulmonary trunc, ran posteriorly between the trachea and oesophagus and reached the left hilum (Figure 1c and 1d). The trachea seemed narrow (**Figure 1d**).

Cardiothoracic index was higher than normal with no hemodynamic anomaly. Pulmonary veins were not visible in the left atria and an abnormal vein was seen running along the left atrial roof, from the left lung to the superior vena cava (**Figure 2**). There were no other intracardiac defects. Genetic investagation showed a normal karyotype 46XX and absence of 22Q1.1. microdeletion. The patient opted for pregnancy termination.

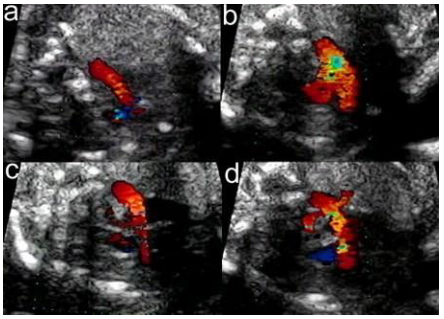

Figure 1 Three vessel view (a): Absence of LPA in the expected location. (b): Aortic arch and ductus arteriosus on the left side of the trachea. (c) and (d): LPAS: left pulmonary artery arising on the right side of the pulmonary trunk, running from right to left behind trachea

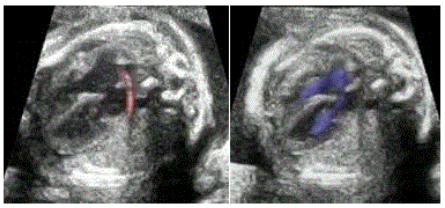

Figure 2 TAVR of left lung. Left atria filled with foramen ovale shunt alone

Post-Mortem Examination

Post-mortem analysis confirmed agenesis of the right lung with no parenchyma, right pulmonary artery or veins and no right bronchus. The thymus filled the upper right chest, and the right hemidiaphragm was elevated. The left lung had only one lobe. A single left pulmonary vein was connected to superior vena cava. The left lung was vascularised by the LPAS, arising from the right side of the pulmonary trunk and circumventing the trachea. The LPAS compressed the lower part of the trachea but there was no structural anomaly.

No intracardiac anomaly was observed, except for a large ostium secundum atrial septal defect (**Figure 3**).

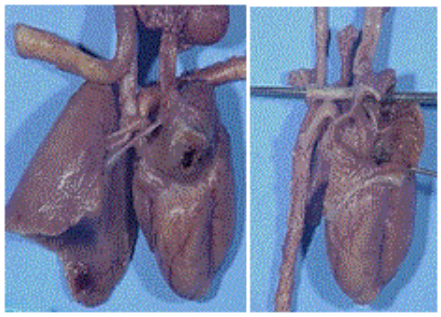

Figure 3 Posterior view. LPAS and APVR in superior vena cava

Discussion

PA is defined by a total absence of the pulmonary parenchyma, vascular structures and bronchi. It is an uncommon anomaly in which there are frequent pulmonary, cardiovascular, tracheal, renal, facial, skeletal and urogenital associated anomalies. Association of PA with syndromes (DiGeorge, Goldenhar, VACTERL) is frequent [1]. In our case, the presence of bilateral preauricular tags raised the question of a genetic syndrome.

The clinical presentation of unilateral PA is variable. Symptoms may appear soon after delivery, or the patient may remain asymptomatic, and be diagnosed during routine examination.

LPAS is a rare vascular anomaly in which the left pulmonary branch originates from the right pulmonary artery, passes over the right main stem bronchus, turns to the left, and courses between the trachea and oesophagus to reach the left lung. One case of prenatal diagnosis has been reported [2]. Neonatal presentation depends on associated tracheal and cardiac anomalies [3,4]. Tracheal narrowing is constant in LPAS with right pulmonary agenesis. Prenataly, it is difficult, to differentiate intrinsic tracheal stenosis with cartilaginous rings, from vascular compression [4]. Cardiac malformation is a major pronostic factor when early hemodynamic changes occur. In our case, early congestive heart failure and severe pulmonary hypertension could have been expected because of total TAVR. Left pulmonary hypoplasia was suspected due to enlarged cardio thoracic ratio without hemodynamic disease. Left pulmonary hypoplasia would have worsened prognosis.

Despite surgical progress, the management of very young infants with LPAS and major associated malformations remains of very poor prognosis [5-9]. Accurated expertise is required in prenatal diagnosis to assess the risk of surgical correction.

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