Journal of Pulmonary & Respiratory Medicine

Mikulic et al., J Pulm Respir Med 2014, 4:3
DOI: 10.4172/2161-105X.1000185

Case Report Open Access

Absence of Left Pulmonary Artery after Delivery, a Case Report and Review of Literature

Lucas A Mikulic* **and Haitham Nsour**

Department of Pulmonary and Critical Care Medicine, Fletcher Allen Healthcare, University of Vermont, USA

***Corresponding author:** Lucas Alejandro Mikulic, Clinical Instructor, Department of Pulmonary and Critical Care Medicine, Fletcher Allen Health Care, University of Vermont College of Medicine, 111 Colchester avenue, Burlington, Vermont, 05401, USA, Tel: 802-847-1158; E-mail: lucas.mikulic@vtmednet.org

Received date: Mar 15, 2014, **Accepted date:** Apr 29, 2014, **Published date:** May 01, 2014

Introduction

Congenital unilateral absence of a pulmonary artery (UAPA) is a rare condition that has seldom been described in pregnancy. We report the case of 22 year old women who remained asymptomatic until she was diagnosed with unilateral absence of the left pulmonary artery after child delivery.

Case Presentation

A 22 year old female with no past medical history underwent an uneventful caesarian section and gave birth to a healthy newborn. Three days after she was discharged home the patient was readmitted with acute respiratory failure requiring intubation and mechanical ventilation.

Her physical exam was significant for tachypnea with a respiratory rate 30 breaths/minute, pulse 120 beats per minute, blood pressure 150/90 mmHg, temperature 100.6 F. Lungs revealed bilateral bronchial breath sounds. Her cardiovascular examination was significant for tachycardia with normal s1 and s2 and no murmurs or gallops noted. Abdominal exam was benign. Her extremities showed normal pulses with no clubbing or cyanosis.

Her initial blood work showed mild leukocytosis (white blood cell count of 11.9 mg/dL), chemistries within normal limits, brain natriuretic peptide 337 ng/dL, and arterial blood gasses significant for a pH 7.44, pCO2 33, pO2 145. Influenza swab was negative. Chest X ray revealed decreased size of the left hemithorax, compensatory hyperinflation of the contralateral hemithorax, elevation of the ipsilateral hemidiaphragm, enlarged contralateral pulmonary artery shadow, a right-sided aortic arch and an ipsilateral shift of the mediastinum (Figure 1).

A chest computed tomography with intravenous contrast revealed absence of the left pulmonary artery, dilation of the right pulmonary artery, and no acute pulmonary emboli. No evidence of ductus arteriosus or ductal stump at the level of the brachiocephalic artery was seen (Figure 2).

A ventilation/perfusion scan was performed showing moderately decreased ventilation with no perfusion on the left lung, and normal ventilation and perfusion of the right lung (Figure 3). An echocardiogram revealed a normal ejection fraction with no ventricular septal defect. Pulmonary pressures were not obtained.

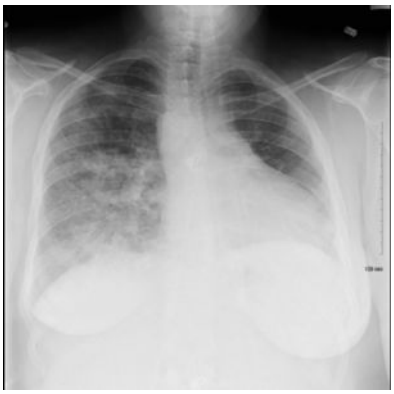

Figure 1: Chest x ray

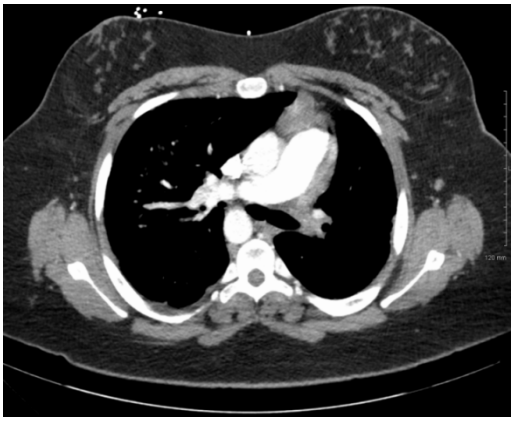

Figure 2: Computed tomography angiography of the chest

The patient was treated with broad spectrum antibiotics for presumed multilobar pneumonia, and eventually she was weaned off the ventilator and discharged home on oxygen therapy.

As an outpatient her pulmonary function tests showed an FEV1/FVC 96% predicted, an FVC 73% predicted and FEV1 70% predicted, consistent with muscle weakness, poor effort or restrictive disease.

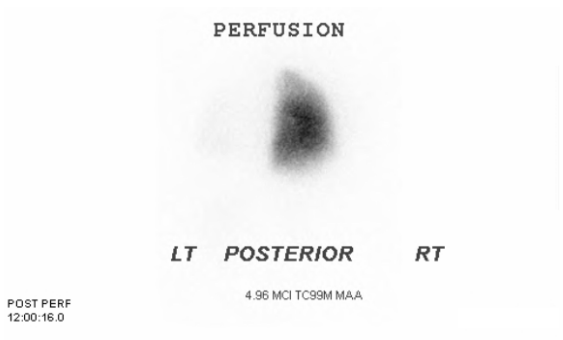

Figure 3: Perfusion scan

Discussion

Isolated unilateral pulmonary artery agenesis is an imprecise term that refers to the absence on the origin of the proximal pulmonary artery.

During embryogenesis, the proximal sixth aortic arch is destined to become the proximal pulmonary artery. The distal pulmonary artery arises from their respective lung buds, and the ductus arteriosus originates from the distal sixth aortic arch. When there is involution of the proximal sixth aortic arch, the proximal pulmonary arteries fail to develop, and the distal pulmonary arteries join the distal aortic arch (which is destined to become the ductus arteriosus) instead [1].

It has an estimated incidence of 1:200,000 [2,3] with right pulmonary artery agenesis being twice as common as left pulmonary artery agenesia [4]. The median age at presentation is 14 years [3] and no gender predilection or evidence of heredity has been identified [5].

The incidence of isolated UAPA in pregnancy is not well reported in literature. In a review of 108 cases reported between 1978 and 2000 performed by Tan Herkel and colleagues, only three cases were identified [3,6-9]. In one patient the diagnosis was made at 27 weeks gestation, while the other two were diagnosed the first and fourth day post-partum.

The clinical presentation of isolated UAPA in adults varies and it has been reported that up to 30% patients with isolated UAPA may remain asymptomatic [3,4]. The most common presentations are dyspnea (40%), infections (37%), and hemoptysis (20%) [3]. The etiology for recurrent infections has been associated with the lack of blood supply to the affected lung, which leads to secondary bronchoconstriction due to alveolar hypocapnia, decreased inflammatory response, decreased mucociliary clearance and mucous trapping [3,4]. Hemoptysis may occur due to increased intercostal and bronchial collateral circulation (the latter receiving 17% of the cardiac output instead of the usual 1%). This increase in blood flow leads to an increase arterial pressure in bronchial arteries and venules, making them more predisposed to bleed [10]. Pulmonary hypertension has been described as being present in 19% cases, and it plays an important role in the prognosis of these patients [3,11,12].

The diagnosis of UAPA requires a detailed medical and physical exam. Initial chest X rays shows the typical findings seen in our patient. To establish a definite diagnosis, a chest Computed Tomography (CT) or magnetic resonance imaging MRI can be performed, which would show the absence of the pulmonary artery. Ventilation/perfusion shows absent perfusion and hypoplastic ventilation of the homolateral lung. An echocardiogram is crucial in order to establish if the patient has any cardiac anomalies as well as to quantify and follow up patients with pulmonary hypertension. Angiography is currently reserved for cases where embolization is indicated [13].

Treatment is directed towards the clinical presentation. Patients with recurrent infections or massive hemoptysis may need pneumonectomy or embolization of the affected vessels [13]. The development of pulmonary hypertension during pregnancy imposes a high risk of morbidity and mortality, which requires termination of pregnancy. If this is not accepted by the patient, or the pregnancy is too advanced, adequate oxygenation (keeping maternal PaO2 >70 mmHg) is crucial to avoid hypoxemia, pulmonary vasoconstriction, and right sided heart failure [14]. It has been described in the literature that placing the patient on the lateral decubitus with the affected lung down improves oxygenation [7].

In regards to the mode of delivery, of the 3 cases reviewed by Ted Harkel et al. [3] two patients underwent vaginal delivery, while the remaining one had a cesarean section. In a separate case report, Yoshihara et al. [9] reviewed the case of a 28 year old primigravid patient with a known diagnosis of right sided UAPA who underwent a successful vaginal delivery of a normal female infant.

Prognosis depends on the presentation and its association with cardiac anomalies. The overall mortality of UAPA is 7% [3]. In in a review performed by Harkel et al. [3] one of three pregnant patients with UAPA died after delivery due to complications of pulmonary hypertension and acute respiratory distress syndrome [3].

Our patient was eventually discharged home and followed up a few months later at the pulmonary clinic. She was still requiring supplemental oxygen. Unfortunately, she was lost to follow up after she was referred to the pulmonary hypertension clinic.

Conclusion

Unilateral absence of the pulmonary artery is a rare disease that usually occurs in conjunction with cardiac anomalies. When this does not occur, the patient may present with minimal symptoms until they reach adulthood. Its diagnosis requires a high level of suspicion and it may be unmasked by pregnancy or immediately after delivery.

References

1. Apostolopoulou SC, Kelekis NL, Brountzos EN, Rammos S, Kelekis DA (2002) "Absent" pulmonary artery in one adult and five pediatric patients: imaging, embryology, and therapeutic implications. AJR Am J Roentgenol 179: 1253-1260.
2. Fraentzel O. Angeborener Defect de Rechten Lungernarterie. Virchows Arch Pathol Anat 1868; 43:420
3. Ten Harkel AD, Blom NA, Ottenkamp J (2002) Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest 122: 1471-1477.
4. Griffin N, Mansfield L, Redmond KC, Dusmet M, Goldstraw P, et al. (2007) Imaging features of isolated unilateral pulmonary artery agenesis presenting in adulthood: a review of four cases. Clin Radiol 62: 238-244.
5. Kremer S, Fayemi AB, Fish BG, Rothe WA, Spindola-Franco H (1992) Congenital absence of the left pulmonary artery associated with double-outlet right ventricle. AJR Am J Roentgenol 158: 1309-1311.

6. Stiller RJ, Soberman S, Turetsky A, Lockwood C, Haddad R (1988) Agenesis of the pulmonary artery: an unusual cause of dyspnea in pregnancy. Am J Obstet Gynecol 158: 172-173.
7. Ferrari M, Karrazi R, Lampronti G, Biasin C, Zuccali V, et al. (1997) Effect of changing position on arterial oxygenation in a patient with agenesia of the left pulmonary artery. Respiration 64: 371-374.
8. Ko T, Gatz MG, Reisz GR (1990) Congenital unilateral absence of a pulmonary artery: a report of two adult cases. Am Rev Respir Dis 141: 795-798.
9. Yoshihara K, Kurabayashi T, Tsuchiya M, Terabayashi K, Serikawa T, et al. (2006) Pregnancy with congenital unilateral absence of a pulmonary artery. Acta Obstet Gynecol Scand 85: 755-757.
10. Kadir IS, Thekudan J, Dheodar A, Jones MT, Carroll KB (2002) Congenital unilateral pulmonary artery agenesis and aspergilloma. Ann Thorac Surg 74: 2169-2171.
11. Shakibi JG, Rastan H, Nazarian I, Paydar M, Aryanpour I, et al. (1978) Isolated unilateral absence of the pulmonary artery. Review of the world literature and guidelines for surgical repair. Jpn Heart J 19: 439-451.
12. POOL PE, VOGEL JH, BLOUNT SG Jr (1962) Congenital unilateral absence of a pulmonary artery. The importance of flow in pulmonary hypertension. Am J Cardiol 10: 706-732.
13. Kruzliak P, Syamasundar RP, Novak M, Pechanova O, Kovacova G (2013) Unilateral absence of pulmonary artery: pathophysiology, symptoms, diagnosis and current treatment. Arch Cardiovasc Dis 106: 448-454.
14. Madden BP (2009) Pulmonary hypertension and pregnancy. Int J Obstet Anesth 18: 156-164.