Case report

Unilateral pulmonary artery agenesis: An unusual cause of unilateral ARDS

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ABSTRACT

Unilateral pulmonary artery agenesis (UPAA) is a rare malformation that can present as an isolated anomaly or may be associated with certain congenital cardiac anomalies, such as tetralogy of Fallot, atrial septal defect, coarctation of aorta, right aortic arch, truncus arteriosus and pulmonary atresia. Clinical presentation is non-specific which makes the diagnosis elusive; chronic dyspnea, hemoptysis or recurrent infections are the most common manifestations. Patients may remain asymptomatic until adulthood. There is no definitive treatment for patients with UPAA. Acute respiratory distress syndrome (ARDS) is usually a bilateral disease, unilateral ARDS has been described after lung resection or trauma. We present a case of a 39-year-old woman who developed unilateral ARDS and was later diagnosed with isolated UPAA.

1. Case presentation

A 39-year-old woman presented for elective right robotic nephrectomy prior to renal transplant. Her medical history was significant for end stage renal disease on hemodialysis, atrophic left kidney and deformed right kidney. Her deceased brother had unknown congenital malformations. She had no toxic habits. Immediately after induction of general anesthesia, she developed severe laryngospasm and attempts for intubation were unsuccessful. She was placed on non-invasive positive pressure ventilation with some improvement, however 4 to 6 hours later she developed progressive hypoxic respiratory failure associated with massive hemoptysis and shock. She required intubation and transient use of vasopressors. Pertinent findings on examination were decreased breath sounds in left hemithorax and a grade IV/VI systolic murmur along left sternal border. No jugular venous distention or leg edema was present. She was dialyzed the day prior to planned surgery.

Chest computed tomogram (CT) revealed absent left pulmonary artery and extensive right lung consolidation (Fig. 2).

Echocardiogram revealed severe pulmonary hypertension (pulmonary artery systolic pressure: 62 mmHg) with right ventricular dysfunction and normal left ventricular function. Ventilation/perfusion scan confirmed absence of perfusion to the left lung (Fig. 3).

She was managed with lung protective strategy and later liberated from mechanical ventilation and discharged home.

2. Discussion

The first reported case of UPAA was published in 1868 by Frantzel O. Angeborener and was demonstrated angiographically in 1952 by Madoff and colleagues [1,2] UPAA is rare, with a prevalence of 1:200,000 in young adults and usually occurs in conjunction with cardiovascular anomalies. This condition commonly affects the right lung and occurs on the side opposite the aortic arch [3].

Pulmonary artery branches are formed from the sixth aortic arches in embryos during the fourth week of gestation. During normal development, the proximal portions of the sixth arch persist, forming the right and left main branches of the pulmonary trunk. The pulmonary vascular bed may form normally even when a main branch of the pulmonary artery is absent because it develops from ventral branches of the dorsal aorta [4].

This condition can be under diagnosed as 30% of patients are...
asymptomatic [5]. It can present with dyspnea on exertion, hemoptysis and frequent respiratory infections. Cases of severe recurrent pneumonia due to impaired mucociliary clearance leading to bronchiectasis have been reported [3].

Pulmonary hypertension is seen in 44% of isolated UPAA and results from an imbalance between decreased pulmonary vascular bed due to absent artery and increase in blood flow to the normal pulmonary artery as blood is directed away from the absent pulmonary artery that further leads to endothelial stress and release of vasoconstrictor mediators [6]. Hemoptysis occurs in about 20% of these cases as a result of excessive collateral circulation in bronchial, intercostal, subclavian arteries on ipsilateral side and hyperperfused vessels on the contralateral side.

Common findings on CXR and CT scans include decreased pulmonary vascular markings, absent hilar shadow, mediastinal shift to the affected side, a mosaic attenuation pattern, interruption of pulmonary artery and hyperinflated contralateral lung [7]. High resolution chest CT scan may be required to diagnose bronchiectasis. Echocardiogram is useful to rule out associated cardiac defects and diagnose pulmonary hypertension. Ventilation perfusion scan elucidates anatomy of the affected side. Cardiac catheterization is necessary when revascularization is planned [8].

Swyer-James syndrome or unilateral hyperlucent lung syndrome is an important differential in these patients; it is a rare entity associated with postinfectious bronchiolitis obliterans occurring in childhood. Peripheral pulmonary vascularization is decreased as a result of inflammation. It is characterized by a delay in the washout phase of the ventilation lung scan in the hyperlucent lungs due to obstructive airways [9].

There is no consensus regarding treatment of UPAA. Several case reports of successful outcomes following revascularization have been described in the pediatric population [10]. Patients who develop pulmonary hypertension can be treated medically with vasodilator therapy [11,12]. Hemoptysis may be self-limiting, selective embolization of the systemic collaterals and in selected cases pneumonectomy of the affected side may be required for massive hemoptysis [13,14].

Outcomes of patients with UPAA have not been well described, likely due to the rarity of the condition. Death can result from massive hemoptysis, respiratory failure, pulmonary hypertension leading to right heart failure and high-altitude pulmonary edema [15].

Asymptomatic patients with UPAA should undergo serial echocardiography to monitor for development of pulmonary hypertension [15].

There has been limited literature describing complications of anesthesia in patients with UPAA. Jiang et al. [16] studied the unusual kinetics of sevoflurane uptake and also differences in end tidal CO2 levels between the two lungs in patients with UPAA. The affected lung which has collaterals from systemic circulation has a slower uptake rate of inhaled anesthetic and produces lower ETCO2 levels compared to the normal lung which has mixed venous blood from both pulmonary and bronchial circulations. They concluded that UPAA greatly affects the kinetics of volatile anesthetic uptake in both lungs individually and suggest close ETCO2 monitoring especially during surgery. Barkshire et al. [17] described a patient that underwent non-cardiac surgery using laryngeal mask airway and isoflurane as suitable anesthetic technique. They suggested two important considerations in patients with isolated UPAA - high risk of pulmonary hypertension and serious consequences in patients where the normal function of lung is disrupted. Isoflurane has been shown to inhibit hypoxic pulmonary vasoconstriction more effectively than other volatile agents [18].

Our patient presented with a very rare condition, she had isolated UPAA without cardiac malformation that was asymptomatic until...
Fig. 3. Ventilation perfusion scan with no ventilation perfusion mismatch in normal right lung. The left lung shows almost no perfusion and minimal ventilation.
adulthood. In view of family history, we suspected an associated congenital renal disorder. We hypothesize that she developed severe laryngospasm with upper airway obstruction during induction of anesthesia and attempts of intubation. She most likely had ARDS resulting from negative pressure pulmonary edema due to upper airway obstruction and UPAA. The hemodynamic instability could be from unrecognized severe pulmonary hypertension. ARDS is characterized by inflammatory injury of pulmonary endothelium and epithelium leading to increased vascular and epithelial permeability and leakage of protein-rich edema fluid into the airspaces. Unilateral ARDS and hemoptysis in our patient can be explained by UPAA as perfusion plays a key role in pathogenesis of ARDS [19].

3. Conclusion

In patients presenting with unilateral ARDS, UPAA should be included in the differential diagnosis and awareness of this condition can lead to earlier diagnosis. Early diagnosis with chest CT and perfusion scan can improve the management and potentially the outcome of these patients. Echocardiogram is advised to evaluate for pulmonary hypertension. Larger studies are required to establish optimum airway management and anesthesia for patients with this condition undergoing surgery.

Conflict of interest

The authors have no conflict of interests to report.

Author disclosure

None of the authors have a financial relationship with a commercial entity that has an interest in the subject of the manuscript. No financial support was used for the study.

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