Case report

Pulmonary artery agenesis and Kommerell’s diverticulum presenting with hemoptysis

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Abstract

Unilateral pulmonary artery agenesis is a rare congenital anomaly that is commonly associated with additional cardiovascular abnormalities. We report a case of intrapulmonary hemorrhage and hemoptysis in a patient with left-sided pulmonary artery agenesis as well as the first description of this anomaly in a patient with a diverticulum of Kommerell. The patient was successfully treated by performing a left pneumonectomy.

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1. Introduction

Unilateral pulmonary artery agenesis (UPAA) is a rare condition typically diagnosed as the affected infant or child is being evaluated for one of the commonly associated cardiovascular abnormalities (tetralogy of Fallot, right aortic arch, septal defects, and patent ductus arteriosus) [1]. In adults, on the other hand, UPAA is more likely to be an isolated finding and often presents with recurrent respiratory tract infections, dyspnea, or on rare occasions, hemoptysis [2].

A Kommerell’s diverticulum, or the broad-based origin of an aberrant right subclavian artery from the descending aorta is a similarly rare cardiovascular anomaly to be diagnosed in an adult [3]. The resultant partial vascular ring may lead to symptomatic compression of the underlying trachea and esophagus; however the majority of patients are asymptomatic [4]. The following case represents an unusual presentation of UPAA and the first report of an adult patient with both of these rare vascular anomalies.

2. Case report

A 34-year-old previously healthy man was admitted after his second episode of hemoptysis (200 cc of blood and clots) in a two-week period. He was otherwise without symptoms and denied similar episodes outside of this two-week presentation. He reported being a full term infant without additional medical problems. He was an active smoker with a 30 pack-year smoking history.

Bronchoscopy revealed blood clots within the left-sided airways, but no source of active bleeding. A computed tomography (CT) angiogram demonstrated left-sided pulmonary artery agenesis (Fig. 1A) and an aberrant right subclavian artery emerging from a diverticulum of Kommerell (Fig. 1B). Formal angiography identified a hypertrophied left-sided bronchial artery with multiple collateral vessels (Fig. 2). Transthoracic echocardiography revealed mildly elevated pulmonary systolic pressure but did not identify any structural cardiac anomalies.

The patient’s pulmonary function was consistent with mild airway obstruction with a forced expiratory volume in 1 s of 4.10 l (88% predicted), vital capacity of 5.5 l (96% predicted), and a diffusion capacity of 78% of predicted.

After discussion with a multidisciplinary care team, it was felt that embolization of this extensive network of collaterals was unlikely to provide a durable solution; therefore, the patient underwent a left pneumonectomy. At the time of surgery, the inferior and superior pulmonary veins were identified draining into the left atrium while an extensive network of large caliber arterial collaterals encased the left mainstem airway. The patient was re-explored on the second postoperative day in response to premature filling of his pneumonectomy space, however no active bleeding was identified. The remainder of the hospital course was unremarkable and the patient was discharged home seven
days after the pneumonectomy. The patient continues to do well, without symptoms or further episodes of hemoptysis eight months after his surgery.

3. Discussion

UPAA is thought to result from a failed development of the ventral bud of the ipsilateral 6th aortic arch [5]. Collateral circulation may allow relatively normal-appearing parenchyma to develop; however, a subset of patients develop complications in the affected lung. Ten Harkel et al. [6] performed a retrospective analysis of 108 cases of isolated UPA and found that most patients were symptomatic with recurrent pulmonary infections (37%), dyspnea or limited exercise tolerance (40%), or hemoptysis (20%).

The development of parenchymal hemorrhage and hemoptysis in the reported patient was thought to result from elevated perfusion pressure of lung parenchyma (secondary to systemic collaterals). In fact pulmonary hypertension was diagnosed in 44% of the patients in the Harkel series [6]. Endobronchial and intra-alveolar hemorrhages can result from the capillary turgidity resulting from the systemic blood-flow through enlarged bronchial arteries and other aberrant vessels that originated from persistent embryonic channels [5].

Treatment options for UPA vary depending on the age of the patient, associated conditions and symptoms. In pediatric patients with pulmonary hypertension, revascularization of the affected side is at times an option [6]. In cases of hemoptysis, options include selective embolization and/or pneumonectomy [2]. Long-term follow-up after embolization does show the development of alternate vasculature and collaterals ultimately putting the patient at significant risk for recurrence of hemoptysis [7]. In general, the contribution of the affected lung to gas exchange is minor in this population, making a pneumonectomy a more attractive option.

Whether, and when, to treat Kommerell’s diverticulum is a matter of some debate. Some authors advocate that all Kommerell’s diverticulum should be surgically addressed regardless of size to prevent rupture [8]. Other surgeons recommend interventions for symptomatic diverticuli larger than 5 cm [4]. The diverticulum in the current patient was felt to represent a low risk for rupture at its current size and was completely asymptomatic. Most patients with this anomaly are asymptomatic, although it can cause dysphagia, dyspnea, stridor, wheezing, cough, recurrent pneumonia, obstructive emphysema or chest pain secondary to compression of surrounding structures [4]. All of this patient’s symptoms were attributed to the UPA, and therefore no specific treatment for the diverticulum was undertaken. He will be followed closely in our aneurysm surveillance program. When a diverticulum of Kommerell needs treatment because of aneurysmal dilation or symptomatology, a wide range of surgical approaches has been employed ranging from simple ligation to total arch replacement. A standard approach is still lacking due to the rarity of this condition requiring treatment [4].

In summary, this case represents an unusual presentation of UPA in an adult and the first description of unilateral pulmonary artery agenesis in conjunction with an aberrant right subclavian artery and Kommerell’s diverticulum. The patient was successfully managed with a left pneumonectomy.
References