Bronchial Artery to Pulmonary Artery Fistula Presenting with Massive Hemoptysis in a Pediatric Patient

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Abstract

Hemoptysis is a serious and potentially life threatening event. Mortality is estimated at 13% for this chief complaint with age, volume of hemoptysis and receipt of blood products as risk factors for mortality. Hemoptysis is mostly seen in those with underlying congenital cardiac conditions or Cystic Fibrosis. We describe a unique case of a previously healthy 10 year old male who presented to the ED by EMS with a moderate volume episode of hemoptysis. He was admitted to the PICU where a sudden episode of massive hemoptysis precipitated by forced respiratory effort occurred during his examination. He decompensated and was emergently brought to the OR for airway evaluation by ENT and pulmonology. A large clot was found in the RML segment with brisk bleeding following removal of the clot. A 5 Fr bronchial blocker was placed to achieve hemostasis. Bronchial artery angiogram by IR demonstrated extravasation of contrast from right bronchial artery to segmental right lower lobe pulmonary artery shunt. He underwent embolization of the right bronchial artery. He was extubated the following day after no recurrent bleeding was confirmed with bronchoscopy. BA-PA fistulas are rare vascular anomalies in which an anastomosis is formed between systemic and pulmonary arteries. They are most commonly acquired, often described secondary to chronic inflammatory lung diseases. BA-PA fistulas can also be congenital and have been seldom described in the literature. Our case highlights the importance of this rare diagnosis, which must remain on a pediatric pulmonologist's differential due to the significant associated mortality.

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Brief Title – Massive Hemoptysis in a Pediatric Patient

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Introduction

Hemoptysis or expectoration of blood from the airways is a serious and often life-threatening event that is, thankfully, rare in pediatric patients. Mortality associated with hemoptysis is caused by asphyxiation not exsanguination. Owing to its rarity, hemoptysis is often difficult to diagnose in pediatric patients. Children tend to swallow their sputum and thus, hemoptysis may go initially un-noticed (Pianosi P, Al-sadoon H 1996). Establishing that a child is experiencing true hemoptysis is essential as extra-pulmonary bleeding such as epistaxis or hematemesis may be incorrectly attributed to hemoptysis.

Massive hemoptysis has been defined as over 8ml/kg in 24 hours (Batra PS, Holinger LD, 2001) in the general population and over 240ml in a patient with cystic fibrosis (Flume PA et al, 2010). A 2017 retrospective cohort study by Moynihan et al, 2021 demonstrated high inpatient mortality with hemoptysis requiring ICU admission and found that mortality was independently related to hemoptysis onset location, underlying diagnosis and severity of clinical illness at the time of the event. They also crucially observed a paucity of current literature on pediatric hemoptysis which has led to a complete lack of contemporary evidence-based guidelines for the evaluation and treatment of hemoptysis in this population (Moynihan et al, 2021).

The etiology of hemoptysis in pediatric patients is numerous, ranging from the common, infection to the rare, vasculitides (Godfrey S, 2004, Shnayder R et al, 2018). The clear majority of hemoptysis occurs in patients with underlying pulmonary and/or cardiac disease states and is rare in previously healthy children. Hemoptysis secondary to vascular anomalies in pediatric patients has been detailed in the past 20 years, however it has been largely attributed to arteriovenous malformations (AVMs) (Chung Y et al, 1997, Sharifi, M et al, 1996 Sheikh S et al, 1999, Villardo RJM et al, 2011). Bronchial artery to pulmonary artery (BA-PA) fistula is an exceedingly rare condition seen in children. There is only a scarce number of cases reported in the current literature, which makes our case relevant to bring awareness about this severe entity and its potential to cause massive hemoptysis and death in otherwise healthy pediatric patients (Avdimiretz N et al, 2020, Gachelin E et al, 2014, Nugent Z et al, 2013, Sismanlar T et al, 2016).

Case Report

A previously healthy 10-year-old boy presented to our pediatric emergency department (ED) with complaints of large volume hemoptysis one hour prior to arrival in the setting of scant hemoptysis for the past 3 days. He reported initially coughing up small amounts of blood-tinged sputum that steadily increased in volume over the preceding 72 hours. Coughing was accompanied by mild left-sided chest wall pain but no pleuritic chest pain was reported. Our patient reported being otherwise well over the past 3 days with no fever, nasal congestion, headaches or GI symptoms noted. He had been playing football all week and was involved in

multiple significant tackles. He attended the ED 10 days prior for epistaxis following a helmet to helmet collision which was treated with oxymetolazine hydrochloride nasal spray with no recurrence of epistaxis.

Upon arrival in the ED, he was awake, alert and in no acute distress with heart rate of 90, blood pressure of 114/64, oral temperature of 98.4, respiration rate of 18 and SpO2 of 97% on room air. He was noted to have dried blood in and around his mouth and nose. No other focal findings noted on initial exam, with clear air entry bilaterally noted on respiratory exam. Laboratory work did not reveal any evidence of anemia (haemoglobin of 14.7), electrolyte dysfunction, liver function abnormalities, infection or coagulopathy. No hematuria noted on urine analysis. Chest x-ray demonstrated patchy consolidation in the right lower lobe which was further characterised on CT chest as abnormal patchy ground-glass opacities in the right middle and lower lobes suggestive of pulmonary haemorrhage (figure 1). He remained hemodynamically stable on room air while in the ED and was transferred to our pediatric ICU (PICU).

Upon arrival to our PICU, while transferring from stretcher to the bed, our patient experienced another episode of hemoptysis of approximately 20ml with desaturation to 86% which resolved with 2 litres of oxygen FiO2 100% via nasal cannula. He remained stable on 2L NC overnight until he experienced another 2 episodes of hemoptysis of approximately 200ml the following morning during examination. The episodes occurred in quick succession with desaturation to the low 80s, tachypnea to the 30s with diminished air entry in the right mid and lower zones with late inspiratory crackles. Oxygen was increased to 4L via NC, 500ml normal saline bolus and 1 unit of packed red blood cells were administered prior to emergent transfer to the operating room for flexible nasal endoscopy, microlaryngoscopy and bronchoscopy by pediatric otolaryngology, pediatric pulmonology and adult interventional pulmonology. Later, haemoglobin drawn at the time of decompensation was noted to have decreased to 13.1.

Upon arrival to the OR, patient was placed under general anesthesia with easy intubation. Evaluation by ENT revealed no bleeding in the nasal cavity, nasopharynx, oral cavity, supra-glottis, glottis or sub-glottis but did note streaks of blood in the trachea with no obvious bleeding source in the trachea. Flexible bronchoscopy revealed the normal anatomy and normal mucosa of the left lung to the first sub-segmental level. Evaluation of the tracheobronchial tree in the right lung revealed scant fresh blood and small blood clots over the trachea and right mainstem bronchus. A large blot clot that was partially obstructing the airway was found in the right middle lobe (RML) and in the right lower lobe (RLL) (figure 2). Upon removal of the clot, brisk hemorrhage occurred from the RML. Bronchial washings with bloody return were obtained from the RML and sent for microbiology analysis. A Fogarty balloon (#4) was placed in the RML by interventional pulmonology until hemostasis was achieved. A 5F Arndt^{TM} bronchial blocker was placed, driven to the RML and inflated to occlude this bronchus.

Our patient was then transferred to the interventional radiology suite for emergent angiography and embolization. Thoracic aortogram via the right common femoral artery demonstrated a hairpin-like artery corresponding to Adamkiewicz artery supplying the right mainstem bronchus. Angiogram of the Adamkiewicz artery revealed a fistula between the right bronchial artery and segmental pulmonary artery to the right lower lobe (figure 3). A decision was made to embolize the right bronchial artery with final angiogram demonstrating patency of the proximal Adamkiewicz artery with occlusion of the distal vessel including right bronchial artery and bronchial to pulmonary artery fistula (figure 4). A right pulmonary artery angiogram was performed post embolization and demonstrated no thrombosis, AVM, aneurysm or pseudoaneurysm (figure 5). Our patient remained intubated with bronchial blocker in place for a further 24 hours with repeat bronchoscopy on post-op day (POD) 1 showing no evidence of further bleeding. Bronchial blocker was deflated and removed and patient extubated on POD 1. Full evaluation for bleeding diathesis and Echocardiogram found no additional underlying cause for his hemoptysis. Our patient was discharged on POD 3 with stable hemoglobin and hematocrit and no further episodes of hemoptysis have occurred during 2 year-follow up. CT angiogram of the chest 1 month later demonstrated no aberrancy of pulmonary vessels or AVMs.

Discussion

Hemoptysis is a rare, albeit potentially life-threatening, condition in pediatric patients. Infectious processes

such as pneumonia, bronchitis, bronchiectasis and pulmonary tuberculosis are the underlying cause in the majority of cases (Batra PS, Holinger LD, 2001, Godfrey S, 2004, Simon DR et al, 2017). CF also remains an important predisposing factor with 9% of people with CF experiencing hemoptysis and 4.1% having at least one episode of massive hemoptysis in their lifetime (Flume PA et al, 2005, Martin LN et al, 2020, Moynihan et al, 2021). It is exceedingly rare to see massive hemoptysis in an otherwise healthy child. Regardless of the cause or lack thereof, early recognition and prompt efficient work-up is essential in any case of hemoptysis to avoid the potentially fatal sequelae of pulmonary hemorrhage.

Dual circulatory systems, bronchial and pulmonary, supply blood to the lungs. The bronchial circulation is high pressure arising from the systemic circulatory system but low volume while the pulmonary circulation is high volume but low pressure (Shnayder R, Needleman JP, 2018). Consequently, significant volume hemoptysis arises from the bronchial circulation (Colson DJ, Mortelli AJ, 2005). Massive hemoptysis has been attributed to vascular anomalies such as AVMs in recent literature but aberrant vessels have been a rarer cause (Chung Y et al, 1997, Sharifi, M et al, 1996 Sheikh S et al, 1999, Villardo RJM et al, 2011). BA-PA fistulae case reports, including our patient, account for only 5 total cases known to this author (Avdimiretz N et al, 2020, Gachelin E et al, 2014, Nugent Z et al, 2013, Sismanlar T et al, 2016). In adult patients, shunts involving the systemic and pulmonary circulations are typically acquired from chronic pulmonary disease states, malignancy or trauma (Yon JR, Ravenel JG, 2010). Congenital bronchopulmonary arterial connections are rarer and have been previously described as preferentially unilateral, right-sided and in males in the adult population (Uchiyama D et al, 2007, Hsieh CG et al, 2017). In our case, we suspect his BA-PA fistula was congenital in nature, as our patient did not have any underlying condition that would have predispose him to collateral formation, and no recurrence of hemoptysis has been observed during follow up.

Bronchial artery embolization (BAE) was first described by Remy and colleagues in 1974 (Remy J et al, 1974). It has been widely used as treatment for hemoptysis in patients with CF and has been shown to be safe and effective (Moynihan et al, 2021, Barben JU et al, 2002, Barben J et al 2003). High success rates have been reported, largely based on adult literature. Recurrence of hemoptysis after BAE has also been described ranging from 10-55% (Sopko DR, Smith TP, 2011). In the few known cases of BA-PA fistulae, our case included, recurrence of hemoptysis is detailed in 2 cases at 6 and 14 months respectively (Avdimiretz N et al, 2020, Gachelin E et al, 2014, Nugent Z et al, 2013, Sismanlar T et al, 2016). This highlights the importance of close post-op observation and long-term follow up of these patients.

Conclusion

Hemoptysis is a serious and potentially fatal event that is thankfully rare in pediatric patients. A high clinical suspicion should be maintained when assessing a pediatric patient presenting with hemoptysis and referral to the appropriate level of care should be considered. CT chest is superior to chest radiography in ability to detect hemorrhage, however a positive chest x-ray should add considerable weight to the clinical suspicion (Olsen KM et al, 2010)[26]. Infection is the most common cause of hemoptysis in pediatrics but clinicians should not overlook congenital vascular anomalies as a cause due to their risk of catastrophic haemorrhage. BAE is a relatively safe method to treat hemoptysis and has been also demonstrated to be effective in pediatric patients with CF (Roebuck DJ, Barnacle AM, 2008). However, BAE is not always available in children's hospitals.

Clinicians should be cognizant of the multidisciplinary care required to treat pediatric patients with hemoptysis and ensure that services are readily available to guarantee optimal outcomes.

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