A Case of Tracheal Varices in an Adolescent Patient with Cyanotic Heart Disease

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Abstract

Tracheal varices and bronchial varices are infrequently reported in adults as a complication of an underlying vascular obstruction, including portal hypertension, pulmonary arterial hypertension, or pulmonary venous hypertension. Tracheal varices and bronchial varices have been reported in adults with failing Fontan physiology, but this occurrence is rare in children. We report the unusual

presentation of tracheal-bronchial varices due to veno-venous collaterals in an adolescent patient with Glenn physiology for double-inlet left ventricle and portal hypertension secondary to cardiac cirrhosis. We document complete resolution of these varices after heart and liver transplantation.

Keywords: bidirectional glenn; single ventricle; tracheal varices; bronchial varices

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Case Report

A 14-year-old adolescent male patient with palliated single ventricle heart disease presented with several days of worsening hemoptysis without fever or difficulty breathing. His past medical history included double-inlet left ventricle with a large ventricular septal defect and a leftward and anterior aorta. He was initially palliated with a pulmonary artery band and underwent an unsuccessful attempt at two-ventricle repair with septation of his heart. As a result, his VSD patch was reopened, and a bidirectional Glenn was performed (anastomosis of the superior vena cava to the pulmonary artery) at 11 months of age (Figure 1).

The patient had presented 6 months earlier with similar symptoms. At that time, flexible bronchoscopy revealed bleeding from the right main stem bronchus, and he underwent embolization of a tortuous right bronchial artery. Additionally, esophageal

varices resulting from cardiac cirrhosis with portal hypertension required banding.

Physical examination revealed an otherwise pleasant young man in no distress. Vital signs included a blood pressure of 105/64 mm Hg, heart rate of 88 bpm, respiratory rate of 20 bpm, and SpO₂ of 85% (his baseline). He had occasional coughing productive of blood-streaked sputum. His lungs were clear to auscultation. Cardiac examination revealed a single S2 with a 2/6 systolic murmur. His liver was palpable 4 cm below the right costal margin.

Imaging Findings

Repeat bronchoscopy demonstrated new findings of multiple nonpulsatile, dilated vessels covering most of the distal trachea and carina and extending into both main stem bronchi with fresh blood covering the area (Figure 2).

Cardiac catheterization and innominate venography demonstrated a large veno-venous collateral vessel arising from the base of the innominate vein and extending in unobstructed fashion to a venous varicosity. The varicosity appeared to surround the left bronchus and extended proximally to the carina and left lateral tracheal wall with subsequent filling to the left upper pulmonary vein and left atrium (Figure 3A). There was no evidence of significant arterial collateral channels. His pulmonary artery and wedge pressures were significantly elevated. The liver was markedly enlarged with hepatic venous wedge pressures approaching right atrial pressures.

Amplatzer vascular plugs were inserted with effective occlusion of the vascular channel and virtual occlusion of the hemiazygos system (Figure 3B). Occlusion of this vascular channel did not significantly alter the innominate venous pressure acutely.

Figure 1. Our patient's cardiac anatomy. The arrow demonstrates anastomosis of the superior vena cava (SVC) to the right pulmonary artery (RPA) in a bi-directional Glenn. Elevated mean pressures are demonstrated in the pulmonary arteries, right atrium (RA), and right ventricle (RV). LA = left atrium; LV = left ventricle; RPV = right pulmonary vein; LPV = left pulmonary vein.

Clinical Outcome

Upon completion of catheterization, the patient had an episode of massive hemoptysis resulting in occlusion of

his endotracheal tube and acute decompensation requiring chest compressions and escalation of hemodynamic support. The patient was noted to be acutely bleeding from the right

mainstem bronchus on bronchoscopy with blood clots filling the segmental bronchi. He underwent a tube exchange with a dual-lumen endotracheal tube, allowing for adequate single-lung ventilation while effective tamponade was achieved. He required several follow-up bronchoscopies to remove clotted blood from his airway. He was extubated after 2 weeks but remained on vasopressors until undergoing a cardiac and liver transplantation 6 months later. He was discharged home in good condition. Upon return for surveillance catheterization 3 months after transplantation, flexible bronchoscopy showed complete resolution of previously noted TV and BV.

Discussion

Hemoptysis is defined as expectoration of blood or blood tinged sputum from the lower respiratory tract. Massive hemoptysis is defined as bleeding that exceeds 8 ml/kg per 24 hours (or >200 ml over a 24-h period) (1, 2). Hemoptysis is a rare phenomenon in children; it occurs most commonly in the setting of acute lower respiratory infections (e.g., pneumonia, pulmonary abscess). Other causes include upper airway disease, foreign body aspiration, congenital abnormalities (including cardiovascular and arteriovenous abnormalities), inflammatory/autoimmunity disorders, bronchiectasis, trauma, and tumor (3). Children usually swallow their sputum unless the bleeding is substantial, and this may underestimate the incidence of this phenomenon (2, 4). In one tertiary pediatric center, hemoptysis was the primary indication for bronchoscopy in only 0.8% of the cases over a 6.5-year period (4).

Tracheal-bronchial varices have been attributed to underlying vascular obstruction, including portal hypertension, pulmonary arterial hypertension, and pulmonary venous hypertension. It can be seen with single ventricle heart disease with hepatic congestion (5), pulmonary vein stenosis (6), pulmonary vein atresia (7), extrahepatic portal vein stenosis (8), cirrhosis (9, 10), and portal vein thrombosis (11).

The nomenclature of the venous drainage of the tracheobronchial tree does not precisely correlate to the structures with which they are associated, and the venous

Figure 2. (A) Varices extending from the distal trachea, carina (black arrow) and into both main stem bronchi. Left mainstem bronchus (L). (B) Left main stem bronchus with varices extending into the segmental bronchi. LUL = left upper lobe; LLL = left lower lobe.

Figure 3. (A) Varicosity (arrow 2) filling with contrast from the veno-venous collateral arising from the base of the innominate vein (arrow 1). The innominate vein drains into the superior vena cava that drains into the right atrium. (B) Amplatzer vascular plugs (arrow 3) inserted with effective occlusion of the vascular channel.

anatomy can be just as variable among individuals as the arterial system. Venous drainage of the extrathoracic and intrathoracic trachea is via the tracheal veins, which primarily drain into the inferior thyroid plexus. The drainage of the proximal bronchi is via the bronchial veins, and the peripheral bronchi is via the pulmonary veins (12). The right bronchial veins usually drain into the azygos vein, and the left bronchial veins usually drain into the hemiazygos or the left superior intercostal vein. However, the bronchial veins may also drain into the intercostal, pericardiophrenic, or internal mammary veins (13, 14). Anatomical communication between the bronchial and pulmonary venous systems occurs in multiple locations, including the hilum and the subpleural regions (13, 15), although this communication is not usually functional in nonpathologic systems.

In general, varices are most commonly found at the site of anastomotic connections between the portal and systemic venous systems, such as the lower esophagus and the rectum. When portal pressures are increased, such as in cirrhosis of the liver, portal venous drainage decompresses through these anastomoses to the systemic venous circulation. This results in massive dilation of these thin-walled venous structures, increasing the risk of bleeding. The venous drainage of the hilum, main stem bronchi, and distal intrathoracic trachea is via the azygos system, but, because these veins traverse the hilum and run along the intrapulmonary bronchi,

a microscopic network of anastomoses with the pulmonary venous system exists. In the setting of heart failure, these connections may become engorged and dilated, resulting in a potential source of intrabronchial variceal bleeding and pulmonary hemorrhage (15). This is most frequently described in cases of mitral stenosis (16, 17) where increased pulmonary venous pressure results in reversal of flow through the bronchial and pulmonary vein collaterals, causing bronchial vein dilation (14). Rupture of the dilated bronchial veins is considered the mechanism of hemoptysis in these patients. A similar mechanism also explains the common occurrence of hemoptysis in patients with other causes of pulmonary venous obstruction. Most cases are preceded by upper or lower respiratory infections with associated cough.

The venous drainage of the airway may also communicate with vessels running along the alimentary tract. Therefore, portal hypertension has been proposed as a mechanism for the development of TV and BV (5). Some researchers claim that these varices arise from collaterals between the esophageal and tracheal venous systems and that the TV and BV are worsened after esophageal varices sclerotherapy or band ligation (8, 10, 11). Other researchers assert that increased systemic venous pressure is transmitted indirectly through inferior thyroid venous plexus to the tracheal venous system (9, 11).

Arterial blood supply to the tracheobronchial tree can have significant variation among individuals. The arterial

supply of the extrathoracic trachea is usually via the inferior thyroid artery but may also be via the subclavian arteries (14). Although the bronchial arteries almost always supply blood to the distal trachea, the arterial blood to the proximal intrathoracic trachea can be from branches of the bronchial, subclavian, innominate, internal thoracic, or intercostal arteries (14). In addition, the bronchial arterial system often has microscopic anastomoses with the pulmonary arterial system, although these are usually constricted to a point of nonfunctionality in healthy individuals (15, 18).

Life-threatening hemoptysis after a Fontan or bidirectional Glenn has been reported due to the development of aortopulmonary arterial collateral development. A history of one or more Blalock-Taussig shunt procedures has been associated with the presence of collateral vessels (19). Bedard and colleagues estimated an event rate of 3.1% in a 10-year follow-up of 65 survivors after the fontan procedure (19), and Triedman and colleagues reported that angiographically diagnosed aortopulmonary chest wall collateral vessels were present in 37% of catheterizations performed in patients who had undergone a Fontan or a bidirectional Glenn procedure (20). Veno-venous collateral channels are also a well described phenomenon after a bidirectional Glenn and most often are a reflection of elevated pulmonary arterial pressure and resistance (19, 21–24). It is therefore likely that hemoptysis due to aorto-pulmonary arterial

collateral development and that due to trachea-bronchial venous varices are distinct entities requiring different diagnostic and therapeutic considerations.

Diagnosis of TV and BV requires a high index of suspicion after presentation with hemoptysis in the setting of underlying congenital heart disease. Given the much more common etiology of aorto-pulmonary collaterals, angiographic or bronchoscopic evidence of venous varices should be investigated. In one case, tracheal varices were confirmed after suspicious multiple subtle nodules in the mid and distal portion of the trachea after intravenous contrast material-enhanced CT angiography (5). Definitive confirmation is usually via bronchoscopy.

Our patient had a number of reasons to develop venous collateral vessels and varices. His underlying anatomy consisting of a bidirectional Glenn connection (anastomosis of the superior vena cava to the pulmonary artery) and antegrade pulmonary blood flow exposed his superior vena cava to high pressures with some degree of pulsatility. His long-standing heart failure with elevated intracardiac filling pressures (Figure 1) resulted in cardiac cirrhosis and portal hypertension. Occlusion of the venous collateral arising from the origin of the innominate vein and decompressing through a tortuous vessel into the left atrium resulted in an acute change in flow patterns through the Glenn connection and pulmonary circulation. This was followed by massive hemoptysis, perhaps due to a significant increase in variceal congestion and dilation after this alteration in flow dynamics.

The principles of management of massive hemoptysis include maintenance of airway patency, localization of the source

of bleeding, and controlling the hemorrhage. Localization and treatment of hemoptysis demands a multifaceted evaluation involving medical, radiologic, and surgical disciplines (25). Asphyxiation is the most frequent complication of massive hemoptysis. Patients should be monitored in the ICU, and, if emergent intubation is needed, a larger endotracheal tube is preferred. If the bleeding site is known, the patient should be placed in the lateral decubitus position with the affected lung in the dependent position. Routine labs should be obtained to determine and rapidly reverse any coagulopathies that may be present. In general, early flexible bronchoscopy is the procedure of choice to localize bleeding (26). If bleeding is so rapid that it makes visualization difficult, then rigid bronchoscopy can be used for more effective suctioning. Bronchoscopic techniques include irrigation with cold saline, topical administration of vasoconstrictive agents, endobronchial tamponade, and unilateral lung ventilation. If bleeding is severe, a double-lumen endotracheal tube may be placed to permit ventilation of both lungs while preventing aspiration from one lung to another. Pulmonary angiography and embolization is used principally for bleeds involving high-pressure bronchial circulation, which accounts for most cases of massive hemoptysis (27, 28). Laser therapy, electrocautery, argon plasma coagulation, or cryotherapy may be able to stop the bleeding if bronchoscopy identifies limited bleeding mucosal lesions. In almost all reported cases, TV and BV presenting with hemoptysis are too diffuse to ablate topically, and to our knowledge there is no literature supporting such an approach in diffuse TV and BV. Surgery for lateralized

uncontrollable massive hemoptysis unresponsive to other measures or as a definitive therapy is considered as a last resort.

Flexible bronchoscopy 3 months after heart and liver transplantation in our patient demonstrated no evidence of his previously noted tracheal-bronchial varices. The complete resolution of the venous collaterals reflected a reduction in his pulmonary arterial and systemic venous pressures. Our patient's surgically altered anatomy made his pulmonary circulation dependent on elevated systemic venous pressures, which likely contributed to the collateral formation observed on bronchoscopy and cardiac catheterization. Secondary reduction in his cardiac filling pressures would have had benefit on his portal and pulmonary venous pressures. This further supports elevated pulmonary and portal venous pressures as contributors to the development of these varices.

Conclusions

Although infrequently documented as a cause of hemoptysis, especially in the pediatric population, this case demonstrates that tracheal-bronchial varices should be included in the differential diagnosis by pulmonologists, particularly in patients with cyanotic heart disease. Furthermore, this case should remind cardiologists and pulmonologists to consider surveillance bronchoscopy in patients with failing Glenn physiology for early detection and treatment of TV and BV to avoid fatal hemoptysis. ■

Author disclosures are available with the text of this article at www.atsjournals.org.

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