CASE REPORT

A case of tracheal varices: an unusual but important cause of mural nodules in the trachea

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ABSTRACT. Tracheal varices (TV) are uncommon but can be an important source of massive or recurrent haemoptysis. We present a case of TV in a 32-year-old patient with a history of Glenn–Fontan surgery, for congenital tricuspid atresia, and portal hypertension owing to cardiac cirrhosis. We discuss TV presenting as tracheal nodules in the presence of extensive mediastinal collateral circulation.

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There are many well-known benign and malignant conditions that can cause tracheal nodules [1–3]. Tracheal varices (TV) are an uncommon cause [4–6]. There are multiple possible aetiologies that have been proposed for TV including increase in the pulmonary vein pressure and portal hypertension [4–6]. This uncommon diagnosis should be suspected in patients with the appropriate medical history and haemoptysis who have tracheal nodules associated with mediastinal collateral vessels seen on CT.

Case report

A 32-year-old male presented to the emergency department with a history of mild haemoptysis and shortness of breath. He was afebrile. His vital signs showed blood pressure of 100/70 mmHg, heart rate of 90 bpm and respiratory rate of 19. He had a history of congenital tricuspid atresia for which he underwent Fontan's surgery with 2 revisions; the last one was 6 years prior to presentation. Other cardiac problems included mitral valve insufficiency, congestive heart failure and atrial arrhythmias. Non-cardiac medical history was significant for chronic liver congestion, which was thought to be secondary to his failing Fontan procedure. He also had oesophageal varices with history of haematemesis. A transjugular biopsy showed Stage 3 hepatic fibrosis.

On admission the physical evaluation was unremarkable. To further evaluate the haemoptysis, intravenous (iv) contrast material-enhanced CT angiography (CTA) of the chest was performed. 1.25 mm thick axial images were helically obtained following injection of 85 ml iohexol (Omnipaque 350 GE Healthcare, Milwaukee, WI) intravenously after timing the bolus for pulmonary arteries.

A 20 ml injection of saline chaser was followed. CT of the trachea (0.625 mm axial images) and a bronchoscopy were then performed.

Imaging findings

CT depicted the surgical changes related to modified Fontan surgery, which includes direct anastomosis of the superior vena cava to the right pulmonary artery and the use of an extracardiac interposition graft between the transected inferior vena cava and the right pulmonary artery. Extensive collateral vessels were identified in the mediastinum with prominent intercostal arteries and chest wall vessels (Figure 1). There were multiple subtle nodules in the mid and distal portion of the trachea predominantly along the posterior wall (Figure 2). There was no tracheal narrowing, wall thickening or calcification. Diagnostic considerations included secretions or papillomatosis. In view of extensive collaterals in the mediastinum, vascular malformation was suggested. Virtual bronchoscopy showed subtle nodulations in the tracheal wall (Figure 3). Bronchoscopy revealed purplish prominent submucosal tortuous varices with active bleeding (Figure 4).

Clinical outcome

The patient was conservatively managed for episodic haemoptysis, which did not immediately recur. There were multiple other comorbidities, which complicated management. The patient suffered worsening left heart failure and multiple organ failure and died 14 months later.

Discussion

Nodularity or multiple small mural nodules in trachea may be secondary to several conditions [1–3]. Mucus secretions are by far the most commonly encountered abnormality on CT [1, 2]. Relapsing polychondritis is an autoimmune condition characterised by long segment

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Figure 1. Axial 1.25 mm thick CT images at the level of the main bronchi during the arterial phase. This image shows collateral circulation adjacent to the carina (arrow) and prominent intercostal arteries (arrow head).

tracheobronchial stricture formation, mural nodular thickening, calcification and collapse on expiration [2]. Tracheobronchial amyloidosis is an uncommon manifestation of amyloidosis characterised by luminal narrowing and mural thickening of the trachea and the bronchi. Visible tracheobronchial nodules are an uncommon manifestation of sarcoidosis. Tracheobronchial papillomatosis is caused by human papilloma virus infection

Figure 2. Axial 1.25 mm thick CT of the upper trachea at lung window setting shows a cluster of nodules in the posterior tracheal wall which represent tracheal varices (arrows).

Figure 3. Virtual bronchoscopy image of the mid and distal portions of the trachea demonstrates tracheal varices in the posterior wall (asterix).

acquired at birth and causes multiple nodules throughout the airways and lung parenchyma [2, 3]. Other benign, but uncommon, conditions of tracheal nodules include ulcerative colitis, Crohn's disease and tracheopathia osteochondroplastica. Endobronchial and tracheal metastases are a very unusual cause of tracheal nodules. A wide variety of tumours can metastasise to the trachea including many types of carcinomas, sarcomas and melanomas [7].

TVs are another rare, but important, cause of mural nodularity in trachea. These submucosal nodules appear on an otherwise normal trachea. Extensive adjacent collateral vessels should raise suspicion.

The blood supply to the cervical trachea is different from the thoracic trachea and bronchi. Thin arteries branching from the inferior thyroid artery are responsible for the cervical trachea while bronchial arteries supply the thoracic trachea and bronchi. Veins draining

Figure 4. Bronchoscopy at the level of the carina and origin of the left bronchus confirm the presence of varices.

the trachea end in the inferior thyroid venous plexus. Bronchial veins drain blood from larger bronchi and from hilar structures [8]. As a result there are different mechanisms for TV. Varices limited to the cervical trachea have been reported [4].

There are two principal causes of tracheal and bronchial varices: portal hypertension and pulmonary venous hypertension. Both can lead to mediastinal collateral circulation [5, 6]. Anecdotal cases of bronchial varices in portal hypertension with and without cirrhosis have been reported in the literature [9, 10].

Pulmonary venous hypertension can be caused by mitral valve disease, pulmonary venous occlusive disease, Fontan shunt and many other causes. The gradient pressure created in Fontan surgery promotes communications between the higher pressure superior caval system and lower pressure veins. Collaterals can also develop between the aorta and its branches [9, 11]. In accordance with previous published cases of patients with Fontan surgery, haemoptysis can result from aortopulmonary vessels [9, 12, 13]. Pathological evaluation of these patients has shown the presence of submucosal varices in small cartilaginous and non-cartilaginous bronchi [6].

Angiographic intervention can be helpful in these patients since the identification of the bleeding bronchial collateral vessels can be treated with embolisation with coils or ethanol. The combination of transjugular intrahepatic portosystemic shunt (TIPS) and embolisation has been reported in patients with portal hypertension and bronchial varices [9, 10, 13].

The precise mechanism responsible for tracheal varices and haemoptysis in our patient is difficult to elucidate, it may be a combination of the above described mechanisms. However, it is important to note that tracheal varices should be considered in addition to other known tracheal pathologies in a case of multiple endotracheal mural nodules and mediastinal collaterals to prevent inadvertent biopsy.

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