Left main bronchus compression due to main pulmonary artery dilatation in pulmonary hypertension: two case reports

Shareen K. Jaijee,^{1,2} Ben Ariff,³ Luke Howard,⁴ Declan P. O'Regan,² Wendy Gin-Sing,⁴ Rachel Davies,⁴ J. Simon R. Gibbs⁴

¹University of Sydney, Camperdown, New South Wales, Australia; ²MRC Clinical Sciences Centre, Imperial College London, Hammersmith Hospital Campus, London, United Kingdom; ³Department of Imaging, Imperial College London, Hammersmith Hospital, London, United Kingdom; ⁴Pulmonary Hypertension Service, Imperial College London, Hammersmith Hospital, London, United Kingdom

Abstract: Pulmonary arterial dilatation associated with pulmonary hypertension may result in significant compression of local structures. Left main coronary artery and left recurrent laryngeal nerve compression have been described. Tracheobronchial compression from pulmonary arterial dilatation is rare in adults, and there are no reports in the literature of its occurrence in idiopathic pulmonary arterial hypertension. Compression in infants with congenital heart disease has been well described. We report 2 cases of tracheobronchial compression: first, an adult patient with idiopathic pulmonary arterial hypertension who presents with symptomatic left main bronchus compression, and second, an adult patient with Eisenmenger ventricular septal defect and right-sided aortic arch, with progressive intermedium and right middle lobe bronchi compression in association with enlarged pulmonary arteries.

Keywords: bronchus compression, pulmonary arterial dilatation, complications.

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Pulmonary arterial dilatation in the setting of pulmonary hypertension is a common finding.¹ Extrinsic compression of the left main coronary artery and the left recurrent laryngeal nerve due to pulmonary arterial dilatation has been reported in the literature.²⁻⁵ While uncommon, tracheobronchial compression due to pulmonary arterial dilatation is a well-known complication in infants with complex congenital heart disease and vascular rings.⁶ In absent pulmonary valve syndrome, significant symptoms in early infancy are commonly due to bronchial compression from dilated pulmonary arteries and an enlarged right atrium.⁷ In adults, respiratory symptoms caused by tracheobronchial compression are most commonly due to aortic aneurysms in the setting of atherosclerosis.⁸

To our knowledge, there have been no reports of tracheobronchial compression in patients with idiopathic pulmonary arterial hypertension and pulmonary arterial dilatation. We present the first report of an adult patient with severe idiopathic pulmonary arterial hypertension and pulmonary arterial dilatation associated with symptomatic left main bronchus compression. In addition, we present a case of adult congenital heart disease associated with pulmonary arterial dilatation, with progressive, symptomatic bronchus intermedium and right middle lobe bronchus compression.

CASE DESCRIPTION

Patient A is a 73-year-old female who was diagnosed and commenced on treatment for idiopathic pulmonary arterial hyperten-

sion 11 years earlier. On diagnosis, she had a pulmonary arterial systolic pressure of 98 mmHg, diastolic pressure of 40 mmHg, mean pulmonary artery pressure of 67 mmHg, pulmonary capillary wedge pressure of 8 mmHg, mean right atrial pressure of 10 mmHg, cardiac index of 1.92 L/min/m², and pulmonary vascular resistance of 1,181 dyn-s/cm⁵ on right heart catheterization. She was noted to have enlarged pulmonary arteries, 1.5 times the size of the aorta on computed tomography (CT). Over time, she progressed from New York Heart Association (NYHA) class II to class IV symptoms.

CT 10 years after diagnosis showed that the main pulmonary artery abutted but did not compress the left main coronary artery. One year later, she noted a voice change and bovine cough, suggesting vocal cord dysfunction. She was admitted to the hospital with a lower respiratory tract infection and worsening shortness of breath, which only partially resolved with antibiotics and then recurred. Because of progressive shortness of breath and sputum production, she had high-resolution CT that showed aneurysmal dilatation of the central pulmonary arteries. The right pulmonary artery had increased in size to 91 mm in diameter and was associated with left main bronchus compression (Figs. 1, 2).

Spirometry was not tolerated because of cough, and flow-volume loops could not be recorded. Peak flow was severely reduced to an average of 183 L/min from 3 attempts. Her treatment included bosentan and sildenafil. She was subsequently referred to specialist palliative care for symptom control and has continued on medical therapy.

Address correspondence to Dr. J. Simon R. Gibbs, Pulmonary Hypertension Service, Hammersmith Hospital, London, United Kingdom. E-mail: s.gibbs@imperial.ac.uk

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Figure 1. Contrast-enhanced computed tomography of the chest reconstructed in the coronal plane of patient A showing left main bronchus compression (arrow) by an enlarged main pulmonary artery.

Patient B is a 63-year-old male with Eisenmenger's complex associated with right-sided aortic arch, chronic obstructive pulmonary disease, and atrial fibrillation who had a 10-year history of increasing shortness of breath. Severe pulmonary hypertension with a bidirectional shunt across the ventricular septal defect was confirmed on echocardiography, with an estimated right ventricular systolic pressure of 116 mmHg. Over time, he progressed to NYHA class IV symptoms, and his current treatment includes warfarin, bosentan, and sildenafil.

For 2 years, he has had an increasing frequency of lower respiratory tract infections and has expectorated an eggcup full of gray-green sputum every day. CT 3 years ago showed bronchus intermedius and middle lobe bronchus compression with right middle lobe collapse. This progressed 2 years later with aneurysmal dilatation of the right pulmonary artery to 51 mm with intrinsic thrombus, occlusion of the right middle lobe bronchus with consequent collapse of the right middle lobe, and significant compression of the bronchus intermedius (Fig. 3). He has continued on medical therapy.

DISCUSSION

In contrast to adults, the cartilaginous, muscular, and elastic supports of the airway in infants are weak, making the tracheobronchial tree vulnerable to compression. The left main bronchus is located between the descending aorta and the pulmonary artery, making it susceptible to compression with dilation of these vessels.^{9,10} Acyanotic congenital heart disease including absent pulmonary valve syndrome and atrial septal defects causing pulmonary arterial dilatation,⁹⁻¹³ right-sided aortic arch, double aortic arch, and abnormal ligamentous arteriosus insertion,⁶ associated with pulmonary arterial dilatation and tracheobronchial compression, have been reported.

Pulmonary arterial dilatation is a common complication of severe pulmonary arterial hypertension and is not associated with increased risk of death. Our review of the literature found 2 cases of tracheobronchial compression from dilated pulmonary arteries in adults. Symptomatic tracheobronchial compression was reported in an 18-year-old female with newly diagnosed absent pulmonary valve syndrome,¹⁴ and asymptomatic left bronchial compression was reported in a 55-year-old male with chronic thromboembolic disease and congenital pulmonary stenosis with consequent pulmonary arterial dilatation.¹⁵ Left main coronary artery compression and left recurrent laryngeal nerve compression are otherwise well-reported complications of pulmonary arterial dilatation.²⁻⁵

Vascular tracheobronchial compression syndromes rarely present in adulthood. The most common acquired vascular tracheobronchial compression syndrome in adults is due to compression from aortic aneurysms in the setting of either atherosclerosis or Marfan's syndrome.⁸ In adults, CT is the investigation of choice and avoids invasive bronchoscopy. Flow-volume curves may show flattening of the expiratory portion of the curve, suggesting variable intrathoracic obstruction, and the chest x-ray may show a widened upper mediastinum.

Treatment of tracheobronchial compression in infants and children has been successful with surgical decompression^{6,13,16} or endobronchial stenting.⁹ In adults, patients are likely to be unsuitable for surgical intervention. In the 2 cases previously reported in the literature, one patient died soon after diagnosis,¹⁴ and the other managed with pulmonary vasodilators.¹⁵ Endobronchial stenting has not been performed.

In conclusion, we present 2 patients with massive pulmonary arterial dilatation causing tracheobronchial compression. Diagnosis was made on CT, with the clinical presentation of recurrent lower respiratory tract infections in both patients. Large airway compression should be considered in patients with pulmonary arterial

Figure 2. Contrast-enhanced computed tomography image of patient A showing left main bronchus compression (arrow) by the main pulmonary artery.

Figure 3. Contrast-enhanced computed tomography reconstructed in the coronal plane of patient B showing right bronchus compression (arrow).

hypertension associated with markedly enlarged pulmonary arteries as a potential cause of worsening breathlessness.

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