

CASE REPORT

Endobronchial ultrasound for detection of tracheomalacia from chronic compression by vascular ringPYNG LEE,¹ SU-YING LOW,¹ HAN-LIM LIEW,¹ DIANA TAN² AND PHILIP ENG¹¹Department of Respiratory and Critical Care Medicine, Singapore General Hospital and ²Department of Diagnostic Radiology, Tan Tock Seng Hospital, Singapore**Endobronchial ultrasound for detection of tracheomalacia from chronic compression by vascular ring**LEE P, LOW S-Y, LIEW H-L, TAN D, ENG P. *Respirology* 2007; 12: 299–301

Abstract: A 67-year-old female chronic smoker was evaluated for an asymptomatic right paratracheal mass and the diagnosis of double-arch aorta was made. She returned 2 years later with dyspnoea on exertion, productive cough and wheeze on lying supine. Flow volume curve showed variable intrathoracic airway obstruction, and bronchoscopy revealed extrinsic compression of the trachea by double-arch aorta with destruction of the cartilaginous layer visualized on endobronchial ultrasonography. Endobronchial ultrasonography may be a useful adjunctive tool for the identification of adults at risk of postoperative tracheomalacia where tracheopexy or airway stenting can be performed concurrently or sequentially if surgery is contemplated.

Key words: double-arch aorta, endobronchial ultrasonography, tracheomalacia, vascular ring.

INTRODUCTION

Vascular rings are known to cause respiratory symptoms, however, double-arch aorta (DAA) presenting in adulthood is exceedingly rare due to its propensity to cause life-threatening symptoms that lead to early diagnosis and surgery within the first year of life.¹ Although significant relief of symptoms follows surgical correction, radiological narrowing of the trachea and oesophagus as well as impaired ventilatory function are long-term sequelae.^{2,3} This study reports an adult with DAA and discusses the mechanisms responsible for its late manifestation as well as the potential application of EBUS to identify patients at risk of tracheomalacia following surgical correction.

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CASE REPORT

A 67-year-old Chinese female was previously evaluated for an asymptomatic right paratracheal mass on CXR. She smoked 25 pack-years, and had a subtotal thyroidectomy for multinodular goitre. CT showed a DAA encircling the oesophagus with tracheal compression (Fig. 1). Both arches were balanced and functional, which joined to form a right-sided descending aorta.

The patient returned 2 years later with dyspnoea and chest pain on exertion, productive cough and wheeze on lying supine for 2 months. She did not complain of dysphagia or heartburn and had continued to smoke. Clinical examination showed a kyphotic patient with a thyroidectomy scar but no stridor, wheeze, crackles, cardiac murmurs, raised jugular venous pressure, ankle oedema or differential blood pressure recordings over the extremities. Two-dimensional echocardiography was normal except for atrial septal aneurysm, and dipyridamole perfusion study was negative for myocardial ischaemia.

Flow volume curve suggested variable intrathoracic airway obstruction. Flexible bronchoscopy showed a pulsatile indentation of the right lateral tracheal wall by the vascular ring, which caused 40% luminal obstruction and measured 2.5 cm in length. EBUS of the tracheal wall performed with a 20-MHz, radial ultrasonic probe (model UM-3R; Olympus, Tokyo,

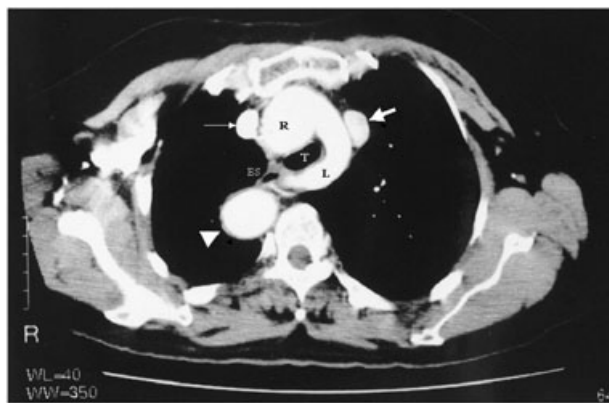


Figure 1 CT of thorax showing the double-arch aorta. Vessels are labelled clockwise from left to right: right-sided descending aorta (arrowhead), oesophagus (ES), right brachiocephalic vein (thin straight arrow), right aortic arch (R), left brachiocephalic vein (thick straight arrow), left aortic arch (L) and trachea (T).

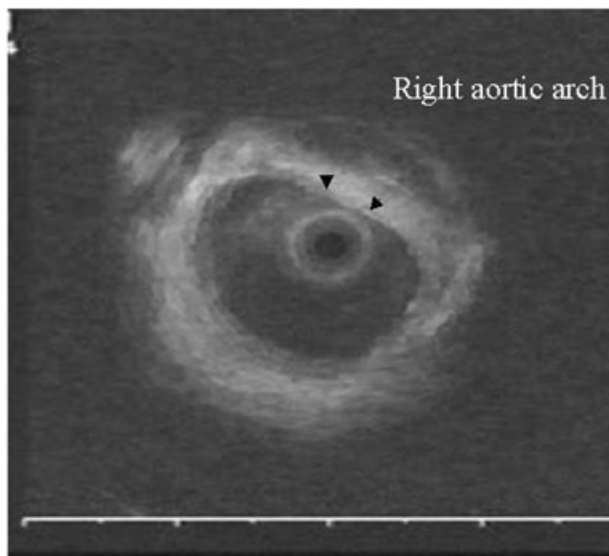


Figure 2 Endobronchial ultrasound demonstrating destruction of the hypoechoic cartilaginous layer of the tracheal wall (arrows) at the site of compression by right aortic arch.

Japan) inserted via the working channel of the bronchoscope revealed cartilaginous layer destruction at the site of maximal compression (Fig. 2). The patient's symptoms improved with smoking cessation and it was decided to defer vascular correction after considering her surgical risks.

DISCUSSION

Vascular rings are congenital anomalies of the aortic arch, which arise as a result of abnormal development of the branchial arch system. DAA is the most

common variant where both arches encircle and compress the trachea and oesophagus leading to symptoms, early diagnosis and surgery. However, late presentation in adults has been reported, and in these cases, symptoms arise as a consequence of static and dynamic changes of the aorta, thoracic cage and trachea.¹

Normal ageing causes 0.1-cm dilatation of the aorta per decade and the onset of kyphosis leads to greater compression of the trachea and oesophagus that can be exacerbated by the supine position.⁴ Symptoms can also occur intermittently with exertion especially if tracheomalacia has occurred due to long-standing compression by the vascular ring.⁵ These mechanisms coupled with persistent airflow inflammation due to chronic smoking, exacerbate airflow limitation and account for the symptoms experienced by this patient.

In recent years, magnetic resonance imaging has become the modality of choice for preoperative evaluation of vascular rings,⁶ and the 16-slice CT for the dynamic assessment of location and extent of tracheal compression. They, however, cannot detect tracheomalacia with certainty. Indeed, postoperative tracheomalacia is a challenging management problem and is the primary reason for dependence on mechanical ventilation, persistent respiratory symptoms, and impaired spirometry, which necessitates tracheal resection or intraluminal stent placement.¹⁻³

EBUS performed during bronchoscopy can demonstrate the layers of the tracheal wall clearly, and may be useful for the identification of patients at risk of postoperative tracheomalacia by determining the contiguity of the cartilaginous layer.⁷ In the current patient EBUS shows cartilage destruction of the affected tracheal segment that measures 2.5 cm in length. As it is very likely that the attendant tracheomalacia will not improve with airway remodelling following vascular decompression surgery⁸ and because a significant length of the trachea is involved, tracheopexy or stent placement should be considered together with the correction of DAA.

EBUS can be a useful adjunct to bronchoscopy for the evaluation of an adult with vascular ring who presents with variable intrathoracic airflow obstruction. The presence of cartilaginous destruction of the tracheal wall from chronic compression by the vascular anomaly should prompt careful postoperative observation of respiratory symptoms that may suggest tracheomalacia, or serve as an indication for concurrent tracheopexy or stenting especially when significant length of the trachea is affected.

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