

Not All That Wheezes Is Asthma

A 62-year-old female complained of exertional dyspnoea associated with occasional wheeze over the past 10 years. She could not identify any trigger factors such as exposure to cold air, exercise or respiratory tract infections. She was treated empirically for bronchial asthma by her family physician with inhaled Salbutamol and inhaled Fluticasone without much relief in symptoms. She also recently complained of mild dysphagia on swallowing large boluses of food. There was no associated chest pain, weight loss or dyspepsia. The patient worked as a seamstress and was a non-smoker.

Physical exam was remarkable for expiratory wheeze, which was monophonic. Her voice was normal and there was no associated cervical adenopathy or goitre on neck examination. A flow-volume loop showed variable intrathoracic upper airway obstruction (Fig. 1A). Spirometry was normal. The chest radiograph is as shown in Figure 1B.

What is the diagnosis causing her symptoms?

- A. Intrathoracic goitre
- B. Paratracheal lymphadenopathy
- C. Aortic aneurysm
- D. Vascular ring
- E. Teratoma

The computed tomography scan of the chest (Fig. 2) confirmed a right-sided aorta associated with a diverticulum of Kommerell, an aberrant left subclavian artery and the left carotid artery forming a vascular ring. Bronchoscopy showed a mild stricture of the mid-trachea due to extrinsic compression. Therefore this patient has congenital vascular ring anomaly causing narrowing and displacement of the trachea and esophagus resulting in dyspnoea and dysphagia lusoria.

Discussion

Congenital anomalies of the great vessels are not uncommon, with a reported prevalence of between 0.5% and 1%.¹ They usually arise from incomplete or anomalous regression of the brachial arch system. These can occasionally lead to compression of the trachea and esophagus, especially when a vascular ring is formed as illustrated in this patient.

In our patient, a vascular ring surrounding the trachea and esophagus is formed by a right-sided aortic arch, the common carotid artery and a diverticulum of Kommerell together with an aberrant left subclavian artery (Fig. 2). A diverticulum of Kommerell is a pouch-like protrusion from the aortic arch and is formed by the persistence of the left fourth brachial arch during embryonic development.²

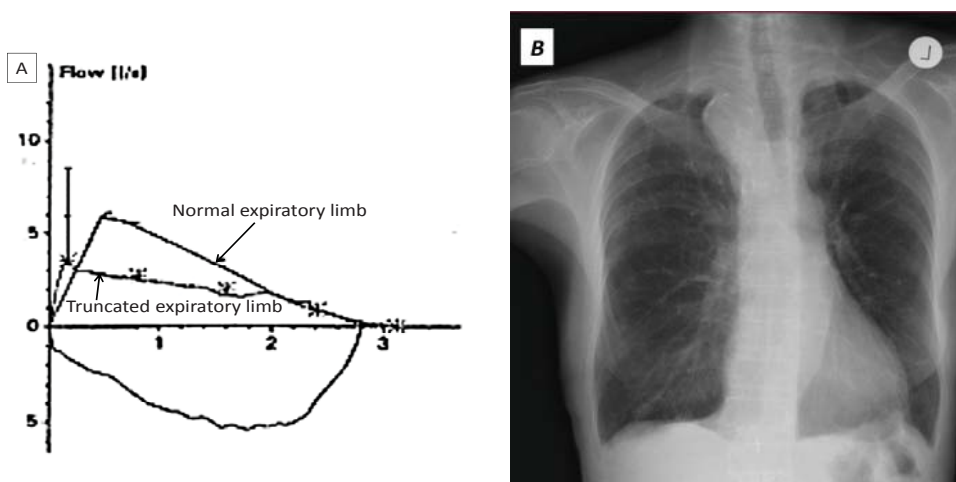


Fig. 1. At (A), chart of flow-volume loop showing truncation of the expiratory limb that is consistent with variable intrathoracic upper airway obstruction. At (B), right-sided aortic arch evident on the chest radiograph.

Answer: D

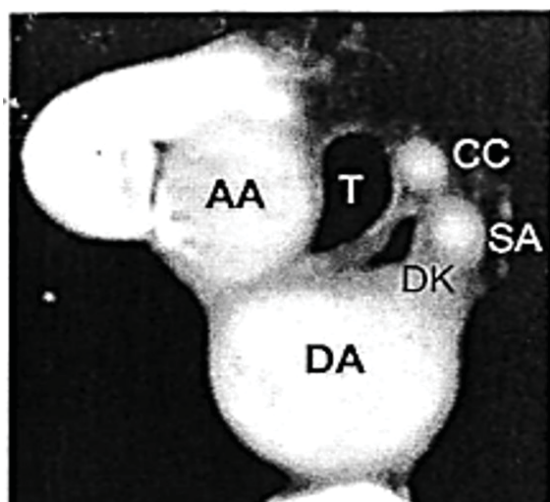


Fig. 2. Computed tomography showing an almost complete vascular ring surrounding the trachea and esophagus. The presence of a right-sided aortic arch, with aberrant left subclavian artery (SA), arising from the diverticulum of Kommerell, the adjoining esophagus being compressed by the aberrant SA resulting in dysphagia lusoria.

AA: Ascending aorta; DA: Descending aorta; SA: Subclavian artery (left); CC: Common carotid artery; DK: Diverticulum of Kommerell; T: Trachea

It usually gives rise to an aberrant left subclavian artery. Other vascular ring configurations have been described, including a right aortic arch with a persistent ligamentous arteriosum, a double aortic arch, an aberrant right subclavian artery and a pulmonary sling (an anomalous left pulmonary artery arising from the right pulmonary trunk which courses posteriorly between the esophagus and trachea).³

These anomalies usually present in childhood, with dysphagia, dyspnoea or wheeze.^{1,3,4} An adult presentation, especially presenting with both dyspnoea and dysphagia, is rare. Misdiagnosis as asthma⁵ and late development of esophageal compression from the vascular ring have been reported.⁶ Our patient had a few unusual clinical features for asthma, in that she had dyspnoea on exertion and dysphagia in the latter part of her presentation. The absence of paroxysmal episodes of wheeze with readily identified trigger factors as well as the lack of response to inhaled steroids and bronchodilators were also unusual. This case also highlights the importance of a chest radiograph and spirometry with a flow-volume loop, especially in patients with atypical features.

The management of a vascular ring varies according to the specific configuration but almost always involves surgical correction and division of the ring.^{1,4} It has been suggested that a diverticulum of Kommerell should always be excised, as there is a risk of rupture. Our patient had declined surgical correction as she deemed her symptoms to be mild and tolerable.

In conclusion, this case highlights the age-old adage that “not all that wheezes is asthma”. It is important to consider vascular ring anomalies, albeit rare, as a cause of dyspnoea and wheeze, especially in patients with aortic arch abnormalities on chest radiograph.

REFERENCES

1. Cordovilla ZG, Cabo SJ, Sanz GE, Moreno GF, Alvarez DF. Vascular rings of aortic origin: the surgical experience in 43 cases. *Rev Esp Cardiol* 1994;47:468-75.
2. Steiner RM, Wechler RJ, Granger RG. The thoracic aorta. In: Granger RG, Albion DJ, editors. *Diagnostic Radiology*. 2nd ed. New York: Churchill Livingstone, 1995.
3. Dillman JR, Attali AK, Agarwal PP, Dorfman AL, Hernandez RJ, Strouse PJ. Common and uncommon vascular rings and slings: a multi-modality review. *Pediatr Radiol* 2011;41:1440-54;quiz 1489-90.
4. Kir M, Saylam GS, Karadas U, Yilmaz N, Çakmakçi H, Uzuner N, et al. Vascular rings: presentation, imaging strategies, treatment, and outcome. *Pediatr Cardiol* 2012;33:607-17.
5. Parker JM, Cary-Freitas B, Berg BW. Symptomatic vascular rings in adulthood: an uncommon mimic of asthma. *J Asthma* 2000;37:275-80.
6. Nicolosi AC, Cambria RA. Late development of esophageal compression from a vascular ring. *Ann Thorac Surg* 1995;60:1413-5.

Serena LW Chong,¹ Anne AL Hsu,² MBBS (S'pore), FRCP (Edin.),

FAMS

¹Department of Medicine, The University of New South Wales, Sydney, Australia

²Department of Respiratory & Critical Care Medicine, Singapore General Hospital, Singapore

Address for Correspondence: Dr Anne AL Hsu, Department of Respiratory and Critical Care Medicine, Singapore General Hospital, 20 College Road, Academia Level 3, Singapore 169856.

Email: gm3hal@sgh.com.sg