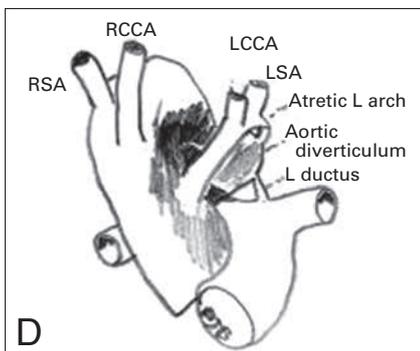
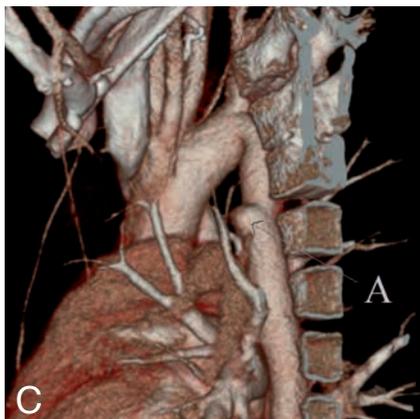
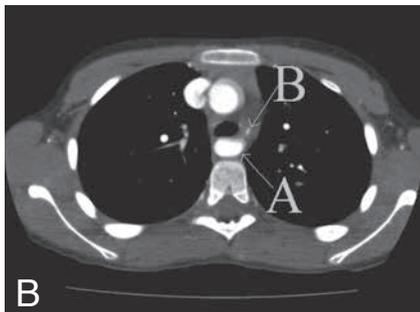


IMAGES IN CLINICAL RADIOLOGY



Unusual case of “airway hyperreactivity” in a child with an incomplete double aortic arch with atresia of the left aortic arch

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A 10-year-old girl had an obstructive breathing pattern with recurrent episodes of wheezing and stridor since infancy, identified and treated as hyperreactivity of the airways with little improvement. Clinical examination revealed a protosystolic ejection murmur (graded 2/6). Subsequent echocardiography revealed a dominant right aortic arch and an atretic left arch.

Cardiac computed tomography was performed and confirmed the dominant right aortic arch (Fig. A) with mirror-image branching. Additionally there was a diverticulum of Kommerell (Fig. A, B, and D arrows A) and a small persisting ductus arteriosus (Fig. B, arrow B). There was a discrepancy between the level of the ductus arteriosus and the tracheal narrowing, suggesting the presence of a fibrous cord.

Only one third of the patients with a vascular tracheobronchial compression will demonstrate significant symptoms and thus the majority remains asymptomatic although a number of them are currently diagnosed with atypical asthma.

According to the classification of Shuford and Sybers there is a spectrum of congenital anomalies from the single right aortic arch to the double aortic arch. Firstly, differentiation between a single right aortic arch with mirror-image branching from a double aortic arch with an atretic left arch (Fig. D) is important because there is an important correlation of a single right aortic arch with congenital heart diseases. Secondly, because the fibrous cord in patients with an atretic left aortic arch has to be searched and transected by the surgeon to relieve the tracheobronchial compression.

Schlesinger et al described some objective characteristics in favour of the diagnosis of an incomplete double aortic arch with atresia of the distal left arch. First, there is a symmetric appearance of the subclavian and common carotid artery originating from the right and incomplete left aortic arch. Next, the incomplete left aortic arch is positioned more posteriorly in comparison with the innominate artery in the right aortic arch, however this is to some extent subjective. Finally there is a diverticulum on the descending aorta when there is an incomplete left aortic arch.

Multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI) are the modalities of choice and the advantage of MDCT over MRI is its superior spatial resolution and the shorter acquisition time, in contrast MRI has the advantage of a higher intrinsic contrast resolution and its superiority in cardiac anatomy and physiologic evaluation. Both techniques do not allow to directly visualizing obliterated vascular segments and do not evaluate dynamic compression of the airway.

In conclusion we state that it is important to consider anatomical variants causing a vascular ring in children having atypical respiratory complaints and recurrent respiratory infections despite adequate treatment.

References

1. Marmon L.M., Bye M.R., Haas J.M., et. al.: Vascular rings and slings: long term follow-up of pulmonary function. *J Paediatr Surg*, 1984, 19: 683-690.
2. Kussman B.D., Geva T., McGowan F.X.: Cardiovascular causes of airway compression. *Paediatr Anaesth*, 2004, 14: 60-74.
3. Shuford W.H., Sybers R.G.: The aortic arch and its malformations. Charles C. Thomas Publisher, Springfield, Illinois, 1974.
4. Schlesinger A.E., Krishnamurthy R., Sena L.M., et al.: Incomplete double aortic arch with atresia of the distal left arch: Distinctive image appearance. *Am J Roentgenol*, 2004, 184: 1634-1639.

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