

A Case of Double Aortic Arch

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Herein we report an 18 months old male infant diagnosed as double aortic arch by angiography. He was presented with persistent stridor and recurrent lower respiratory infections which had developed in neonatal period.

Case Report

An 18 months old male infant was referred to the hospital because of cough, dyspnea and stridor. He was born at term weighing 3.2 kg after an uncomplicated pregnancy and delivery. He was breast-fed until 15 months. By the age of 6 months, his mother started feeding by solid foods. However, she stressed that he could not have eaten solid foods easily. He was referred to another hospital when he was one month old because of stridor where he was diagnosed as laryngomalacia. Since then, he had been hospitalized five times because of lower respiratory infection.

On admission his weight, length and head circumferences were 25 to 50 percentile. Stridor and respiratory distress were noted. Chest X-ray and echocardiogram were normal whereas barium esophagography showed a narrowing on middle esophagus (Figure 1). Aortography revealed double aortic arch (Figure 2). By the age of 20 months surgical division of the vascular ring was performed. He is now 3 years old and completely asymptomatic.

Discussion

Congenital abnormalities of the aortic arch have been known for years, and in addition to the anatomical categorization, can be subdivided according to clinical features. A vascular ring refers to a group of anomalies of the aortic arch in which the trachea and esophagus are surrounded completely by vascular structures that cause respiratory symptoms or feeding problems.

Vascular ring represents less than 1% of all congenital cardiovascular anomalies, but this may be an underestimate because some conditions are asymptomatic. Double aortic arch, the most common type, constitutes 40% of vascular ring anomalies in which both right and left aortic arches are present. The suggested pathogenesis of this anomaly is a failure of the regression of both the right and left fourth branchial arches resulting in right and left aortic arches, respectively (1).



Figure 1 . Barium esophagogram of the patient showing a narrowing on middle esophagus.



Figure 2. Aortography of the patient with double aortic arch.

Several variations on this basic theme occur: both arches widely patent, hypoplasia of one arch and atresia of one arch (usually left). In addition, a ductus arteriosus or ligamentum may also be present. Typically, the right aortic arch is the more superiorly located. Double aortic arch is rarely associated with congenital heart disease, but when it is present, tetralogy of Fallot is most common.

The clinical manifestations of double aortic arches, as with the other vascular rings, are related to the tightness of the ring. With both arches widely patent, the rings are typically tight and patients present with stridor in the first weeks of life. Whereas with double arch and atretic left arch, the rings are usually looser, with presentations at 3 to 6 months of age or later (2). Rarely double aortic arches present in adulthood with swallowing or respiratory symptoms (3). Our patient had been symptomatic since early infancy suggesting that both arches were patent. However, he was diagnosed by the age of 18 months with a delay in diagnosis.

Chest X-ray and echocardiogram are rarely diagnostic. The findings may be more obvious with barium esophagography. Angiography has long been standard for diagnosis but can be confusing because of overlapping structures. In addition, confirmation by angiography or magnetic resonance imaging may be required for differential diagnosis (2, 4). Occasionally differential diagnosis from other vascular abnormality of aortic arch can be established during surgery.

Surgical division of the vascular ring is indicated in any patient who is symptomatic. In conclusion, persistent or recurrent stridor associated with feeding difficulties should prompt an investigation for a vascular ring. Once imaging studies have clearly delineated the causal pathologic vascular structures, surgical division is often effective (5, 6).

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