

Stridor serving as a harbinger of the mirror-image right aortic arch carrying three anatomic dispositions contributing to a noosing vascular ring

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Introduction

Right aortic arch (RAA) is a rare congenital aortic arch anomaly, accounting for 1 out of 1,000 individuals in radiographic and autopsy studies (1). RAA was classified by Edwards into three types: RAA with mirror-image branching (MIRAA) (type III A), RAA with aberrant left subclavian artery (type III B) (with or without Kommerell diverticulum), and RAA with aberrant left brachiocephalic artery and isolated left subclavian artery (type III C) (2). MIRAA, which is the second most common type of RAA, results from evolutionary regression changes of the left dorsal aorta distal to the seventh intersegmental artery, regression of the left sixth arch, and persistence of the right sixth arch (2). MIRAA was frequently associated with concomitant congenital heart disease in pediatric patients, especially those with tetralogy of Fallot, pulmonary atresia, and truncus arteriosus (1, 3). We reported on a 4-month-old boy who had a complete vascular ring (VR), composed of an "isolated" MIRAA (III A RAA), a right-sided retroesophageal aortic diverticulum (AD), and a ligamentum arteriosum arising from the AD to the left pulmonary artery (LPA). The anatomic dispositions that are liable to form a complete VR will be discussed.

Case Report

A 4-month-old boy, who had suffered from stridor since his birth and presented with tachypnea, cyanosis, and loss of consciousness related to postprandial choking at 2 months old, was referred to our hospital for the evaluation of noisy stridor associated with aspiration pneumonia. Prenatal examinations were unremarkable. He was born at 38⁺4 weeks by normal vaginal delivery of a 26-year-old, gravida-3, and para-3 female. His birth weight was 3,630 gm. Apgar scores were 8 and 9 points, respectively. On admission, weight and height were 6.3 kg and

56 cm, respectively. Heart rate was 155/min, respiratory rate 56/min, and blood pressure 87/37 mm Hg. Stridor with subcostal retraction was discernible. Serial examinations, including a radiogram, esophagogram, echocardiography, bronchoscopy, angiography, and computerized tomography showed a complete VR (Fig. 1a-1g, Video 1). After informed consent was obtained from his parents, cardiac surgery through a left thoracotomy was performed. During surgery, a large ligamentum arteriosum was found to originate from a large right-sided retroesophageal descending AD, crossing the midline, and connecting with the LPA (Fig. 1h). Resection of this ligamentum arteriosum and aortoplasty after removal of AD were performed. He was gradually waived of stridor in a 24-month follow-up. Chromosome study for 22q11.2 deletion was negative.

Discussion

Three anatomic dispositions are in a proclivity to form a complete VR in our patient, including RAA; ligamentum arteriosum originating from a right-sided retroesophageal descending AD, crossing midline, and leading to the left side; and connection of the ligamentum arteriosum to the LPA. It is noteworthy that if the patent ductus arteriosus (PDA) or ligamentum arteriosum arises from a right-sided retroesophageal descending AD posteriorly and connects with the LPA anteriorly, there will be a taut traction, between the descending AD and LPA, to noose the trachea and esophagus as a slipknot. Rather, if PDA or ligamentum arteriosum originates from the left subclavian artery and connects with the LPA in MIRAA, there will be no VR (1, 3). In the present case, the right-sided retroesophageal descending AD has an egregious anatomy compressing the esophagus from the right-posterior aspect, and RAA is another one compressing the trachea and esophagus right-laterally. Meanwhile, the curvature angle of a MIRAA, especially in tetralogy of Fallot with pulmonary atresia, between the ascending aorta (which is prone to shift rightward) and right-sided descending aorta (which is prone to cross midline) is lesser than that of a left aortic arch, whose disposition may foster a shorter distance between the ascending aorta and midline descending aorta and result in an extrinsic compression of the trachea and right main bronchus (4). We highlight that the anatomic position of the ligamentum arteriosum may tip the scale to a worsening condition by hauling the LPA forcefully backward, producing a tense traction between LPA and descending AD, tightening this VR as a slipknot, and noosing the trachea and esophagus relentlessly.

There is a trend shifting from a combination of echocardiography, esophagogram, and angiography to a set of magnetic resonance imaging, computerized tomography (CT), and CT angiography in diagnosing VR (1, 5, 6). Bronchoscopy was recently applied as a diagnostic tool in patients with VR (5), providing dynamic visualization of tracheobronchial compression (4, 5).

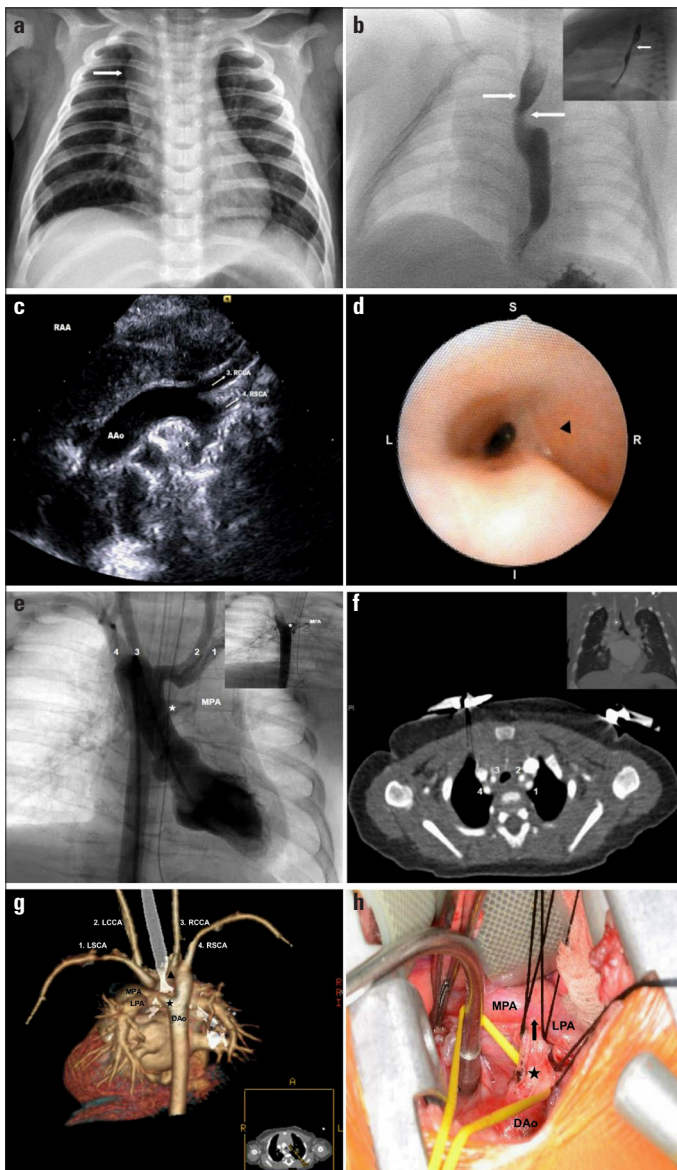


Figure 1. (a) Chest radiogram showed the silhouette of the right aortic arch (RAA) (arrow). (b) Barium esophagogram showed bilateral indentations of the esophagus (arrows) on the frontal projection. The larger left-sided indentation was caused by the aortic diverticulum (AD), while the smaller right-sided indentation was caused by the RAA. Lateral projection of the barium esophagogram, as a miniature panel at the right upper corner, showed posterior indentation of the esophagus (arrow). (c) Suprasternal long-axis view of the two-dimensional echocardiography, with a right-sided long-axis plane passing between the left nipple and right scapular tip, showed a large AD (star) taking off from a right descending aorta, right common carotid artery (RCCA) as the third branch, and right subclavian artery (RSCA) as the fourth branch of the RAA. (d) Bronchoscopy showed an extrinsic compression on the trachea (triangle) by the RAA from the right-anterolateral aspect. The abbreviations of S, I, R, and L indicate superior, inferior, right, and left, respectively. (e) Left ventriculogram showed a mirror-image right aortic arch, with a branching sequence of the left subclavian artery (numeral 1), the left common carotid artery (numeral 2), RCCA (numeral 3), and RSCA (numeral 4). The distal blind end of the AD (star), transformed as a ligamentum arteriosum, was found to be pointing to the main pulmonary artery (MPA) horizontally, rather than pointing to the left subclavian artery vertically, as is seen in incomplete double aortic arch with distal left aortic atresia. Descending aortography, as a miniature panel at the right upper corner, showed that the ductus arteriosus was non-patent, arising from the right retroesophageal descending AD (star). The left-sided indentation visualized in the frontal esophagogram is a repercussion of the extrinsic compression by the aforementioned descending AD. (f) Multidetector computerized tomography (CT) showed a four-vessel sign. However, the brachiocephalic arteries were branched in a sequence of the RAA, and are indicated by the numerals 1, 2, 3, and 4 as those seen in panel E. A miniature of the CT, inserted at the right upper corner, showed extrinsic compression on the lower third trachea by a RAA (triangle). (g) Three-dimensional volume-rendered CT angiography, with reconstruction of the trachea, showed extrinsic compression on the lower third portion of the trachea by a RAA from the right-anterior aspect (triangle). A large diverticulum (star) was visualized to be taking off posteriorly from a right-sided retroesophageal descending aorta. The distal portion of the AD was transformed into a large ligamentum arteriosum as a blind end, which was found pointing to the junction of the MPA and the left pulmonary artery (LPA). (h) At the left thoracotomy, a large ligamentum arteriosum (arrow) was found to originate from a large AD (star) of the right-sided retroesophageal descending aorta (DAo) and connecting to the junction of the MPA and LPA.

Due to a risk of rupture and dissection of the diverticulum in the follow-ups of adults incurring MIRA (7), surgical removal of the diverticulum is indicated in children (8).

Conclusion

Isolated MIRA is a rare aortic arch anomaly and can be masqueraded with the Janus face of stridor among infants in whom three anatomic dispositions are found to be in a proclivity to form a noosing VR. Chest radiogram, esophagogram, echocardiography, bronchoscopy, CT, and CT angiography can make the diagnosis. Surgical resection of the ligamentum arteriosum and AD, and aortoplasty are warranted to prevent rupture and dissection.

Informed consent: Written informed consent was obtained from the patient's family for publication of this case report and any accompanying images.

Video 1. Bronchoscopy showed extrinsic and pulsatile compression on the trachea (black triangle) by a mirror-image right aortic arch.

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