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Case Report
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A Case of Double Aortic Arch

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Abstract

Vascular rings are an uncommon, but important, set of conditions to explore in infants suffering from recurrent respiratory or esophageal symptoms, especially those who present within six months of age. Vascular rings may be isolated or be associated with other congenital heart conditions. Most vascular rings are surgically correctable, and can result in improved quality of life for the infant. Comprehensive pre-operative imaging is a necessity for good surgical outcome and majority of patients report near-complete or complete cessation of symptoms post-operatively. Delays in surgical resection can result in patients presenting later in life with tracheomalacia or worsening respiratory or esophageal symptoms. We report a child who presented with persistent respiratory tract infections since birth, and was found to have a tight vascular ring causing tracheal stenosis on computed tomographic imaging during investigation by Otorhinolaryngology for laryngomalacia. Following full pre-operative assessment and imaging, the child underwent a left sided thoracotomy to divide the vascular ring. Post-operatively, there was a complete resolution of stridor, with no short-term complications. Though vascular rings are a rarity, it is important to have a good understanding about the condition, investigations required and treatment modalities.

Introduction

Vascular rings are a rare but important diagnosis to consider in infants presenting with chronic intermittent respiratory symptoms, or with esophageal symptoms. Patients oftentimes present with a clinical picture consistent with croup owing to the presence of stridor, or with a pneumonia-like picture, due to dysphagia and subsequent aspiration. If the symptoms persist, patients may thereafter be referred for Otorhinolaryngology diseases, such as tracheomalacia or laryngomalacia. However, patients who do not respond to treatment should also be investigated for possible vascular ring anomalies, many of which are amenable to surgical intervention. The double aortic arch, a type of complete vascular ring, is a treatable condition, which, once surgically repaired, can result in improved symptomology and quality of life for the patient.

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Case Presentation

An 8 month old child presented with recurrent croup-like symptoms since birth, including central cyanosis, stridor, cough, and irritability, requiring admission to the hospital in both the general ward and the Intensive Care Unit (ICU). She also had recurrent episodes of aspiration due to dysphagia, which contributed to her recurrent lower respiratory tract infections, as well as her early presentation. Treatment depended on the presentation at the time: Ventilator support and antibiotics for severe episodes of aspiration pneumonia, and adjunctive use of steroids and adrenaline nebulization. Both adjunctive treatments failed to relieve symptoms. She was consequently diagnosed with laryngomalacia, but was found to have a vascular ring causing a fixed tracheal stenosis due to a double aortic arch on Computed Tomographic (CT) imaging.

On physical examination, the child had marked laryngeal stridor with occasional alar flaring and coughing, without costal recessions or cyanosis. Growth trend showed a weight for height above the 97th percentile. Chest X-ray was grossly normal, except for poor visualization of the trachea. CT angiography confirmed the presence of a double aortic arch arising from the ascending aorta, encircling the trachea, and joining the descending aorta; the right arch was slightly larger than the left arch and the vascular ring caused significant tracheal stenosis at the level of T1 (Figure 1); a typical four vessel sign could be appreciated on CTA (Figure 2). Pre-operative echocardiogram showed a dominant right arch with some narrowing of the left arch, and a small patent ductus arteriosus (1.9 mm). An esophageal narrowing due to extrinsic compression at the level of T4/ T5 could be noted on barium oesophagogram. Electrocardiogram (ECG) showed a sinus rhythm at a rate of 150, a normal axis, and signs of left ventricular hypertrophy.

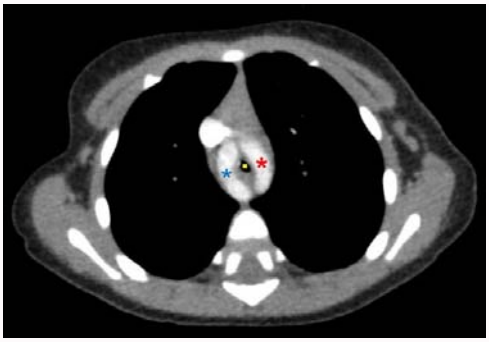

Figure 1: Double aortic arch (right arch – blue asterisk, left arch – red asterisk) encircling trachea with subsequent stenosis (yellow square).

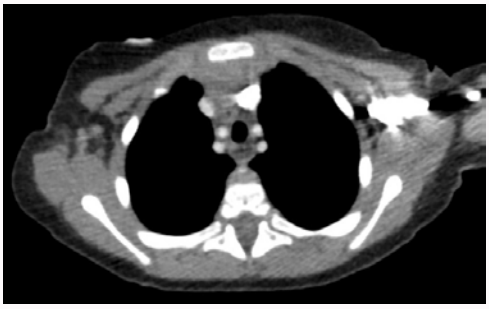

Figure 2: Four vessel sign, showing left and right common carotid arteries anteriorly and left and right subclavian arteries posteriorly.

A left posterolateral thoracotomy approach was used to enter the chest at the third intercostal space. A double aortic arch encircling both the trachea and esophagus were clearly visualized, with the left subclavian artery and left carotid artery coming off the left arch.

The left arch measured 5.0 mm and the right arch measured 6.0 mm. A left-sided patent ductus arteriosus was present, measuring 4.0 mm. The left arch was divided at the junction between the descending arch and the left subclavian arteries. The patent ductus arteriosus was divided and ligated. Post-operatively, the patient had complete relief of symptoms, with an uneventful stay in hospital there after (Figure 3).

Discussion

Congenital aortic arch anomalies are a rare subset of malformations that emerge due to dysfunctional embryogenesis of the branchial arches. Vascular rings compose 1% of all congenital cardiac abnormalities and the double aortic arch, specifically, is the most common type encountered [1,2].

Rathke's model (Figure 4A) describes the development of the branchial arches, which sequentially undergo formation and regression, from the second to the seventh week of gestation, and ultimately form the normal aortic arch and its branches. Edward's theoretical model (Figure 4C) further postulates the presence of a double aortic arch during embryogenesis, formed by the fourth branchial arches, with a ductus arteriosus on each side, formed by the sixth branchial arch, with the descending aorta found at the midline [3-5]. Regression of the right ductus arteriosus and the right dorsal arch form the normal aortic arch anatomy; however, persistence of this form results in the double aortic arch. In fact, most aortic arch anomalies can be explained using Edward's model, wherein persistence or regression of certain segments of the double aortic arch can ultimately lead to different types of malformations.

Double aortic arches can be classified as complete or incomplete, depending on the patency of the arches. Complete, or functional, double aortic arches are the less common type, with both arches being patent and functional. The right aortic arch is dominant in approximately 73% of patients [6]. Incomplete arches are double arches with atresia of one of the arches, usually the left, which may be present as a fibrous continuity [4,6]. A right arch atresia is possible, but extremely rare.

Patients generally present within the first six months of life with stridor, recurrent respiratory tract infections, or dysphagia [7]. Oftentimes, dysphagia results in aspiration, which leads to patient presentation to the hospital, as was the case with our patient. It is rarely associated with other congenital heart diseases. The severity of symptoms depends on the tightness of the vascular ring [6,8]. Rare case reports of patients presenting in adulthood with symptoms due to a vascular ring have been reported [8].

Imaging is crucial in the pre-operative setting for patients with vascular rings, both for its use in diagnosis, and in operative planning. Chest X-ray may appear normal or show abnormalities such as narrowing of the trachea (Figure 5A), atypical mediastinal contours, or widening of the mediastinum. Barium oesophagram is an effective non-invasive modality that can be used to confirm the presence of a vascular ring by demonstrating a persistent, pulsatile, extrinsic compression of the esophagus [2] (Figure 5B).

CT angiography is central in identification and measurement of the aortic arches and their branches, which further assists in planning the surgical approach for treatment [3,9]. Magnetic Resonance Imaging (MRI) is an efficacious alternative to CT, which allows for detailed evaluation of the aortic arch and its branches without exposure to ionizing radiation. However, due to the limitations of MRI, such as the prolonged scanning time and need for general

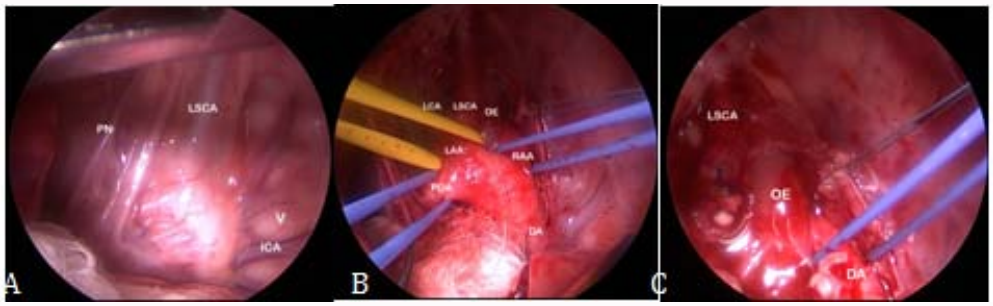

Figure 3: Initial view upon entering the chest cavity (A), showing the Phrenic Nerve (PN), Left Subclavian Artery (LSCA), Vertebral body (V) and Intercostal Artery (ICA).

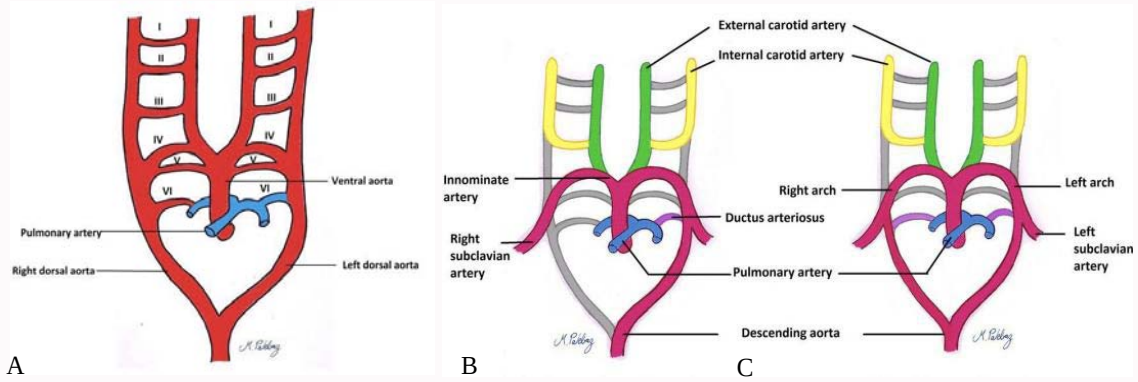

Figure 4: A) Rathke's diagram showing the 6 symmetrical paired arches and dorsal aortas, B) The normal embryological development showing the atretic areas (in grey) over the right aortic arch between the right subclavian and descending aorta, and the remaining patent ductus arteriosus on the left, C) Edward's hypothesized model showing the double aortic arch from which regression occurs at different areas, resulting in various types of aortic arch abnormalities.

Table 1: Benefits and Drawbacks of different diagnostic imaging modalities.

Modality	Benefits	Drawbacks
Ultrasound	Readily available , no ionizing radiation, ability to detect congenital cardiac defects	Operator dependent, limited ability to evaluate complex lesions
CT	Multiplanar, high spatial resolution, readily available	Exposure to ionizing radiation, contrast material required
CT angiography	Multiple planes , high spatial resolution, readily available, ability to grade tracheal stenosis, ability to delineate complex connections	Exposure to ionizing radiation, contrast material required, invasive, time consuming, requiring general anesthesia/sedation
MRI	multi planer and 3D images, good soft tissue contrast, no contrast material required	Long waiting times, limited availability, expensive time consuming, requiring general anesthesia/sedation

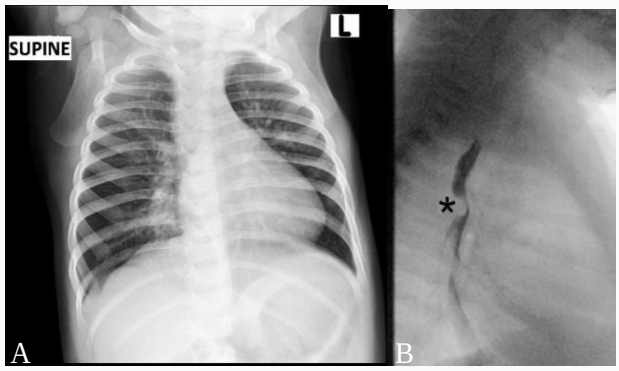

Figure 5: A) Chest X-ray showing poor visualization of trachea and abnormal mediastinal contours. B) Extrinsic compression on barium oesophagram (asterisk).

anesthesia in pediatric patients, CT angiography was favored in our patient [2,5]. Detailed advantages and disadvantages of the different imaging modalities have been outlined in Table 1. Echocardiogram is also important in assessing for other cardiac abnormalities, whose presence is usually uncommon (12%) [2,9]. Together, these forms of imaging are required for accurate planning of operative intervention.

Surgical correction of vascular rings is done most commonly *via* a left thoracotomy, unless pre-operative assessment of the anatomy suggested the need for a right-sided approach [2,7,9,10]. Median sternotomy is reserved for patients with concurrent intracardiac anomalies [2,7,10]. The aim of surgery is to divide the vascular ring, usually at the lesser arch or at an atretic area over one of the arches, and ligate both vascular (ductus arteriosus) and ligamentous structures (ligamentum arteriosum) that may be present, in order to free the trachea and esophagus from compression. If the arches are similar in size and patency, pulses and blood pressure is monitored over the carotids, upper limbs, and lower limbs as the vascular clamps are applied intraoperatively in order to see which arch will be spared, as its patency results in improved palpable pulses and pressures. Another consideration is resection of a Kommerell diverticulum. Commonly, Kommerell diverticulum are found in right aortic arches with aberrant left subclavian artery as a dilated portion of the left subclavian artery at the junction to the descending aorta, which is approximately the same diameter as the descending aorta. This diverticulum is resected and the subclavian artery is anastomosed to the carotid artery. The aorta is repaired with simple continuous suturing, sometimes reinforced with a patch. An important final step is dividing all fibrous bands that may be present adjacent to the esophagus and trachea, to ensure that, following surgery, no remnant incomplete rings are present. Post-operative complications include chylothorax, unilateral vocal cord paralysis, and transient hypertension [6,7]. Incidence of post-operative complications is approximately 15% [7]. Long-term recovery is excellent, with low rates of mortality recorded in multiple institutes.

Conclusion

Double aortic arch is an important diagnosis to consider in patients where there is a high index of suspicion for a vascular ring [11]. Chest X-ray and barium oesophagram are sufficient to diagnose the condition, however, CT angiography (or MRI) and echocardiography are fundamental for surgical planning. Surgical repair of a double aortic arch have excellent outcomes, with low rates of complication. Long-term recovery is good, with effective relief of symptoms.

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