“TYPE-I VASCULAR RINGS”

EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ARTICLE INFO

Article history:
Received: 12 October 2016;
Received in revised form: 13 November 2016;
Accepted: 23 November 2016;

Keywords
Tracheal stenosis,
Subglottic stenosis,
Rima Glottides,
Subglottoplasty,
Nasal congestion,
Dyspnea,
Airway stenosis.

Abstract

The congenital abnormalities of the aortic arch derived vascular and ligamentous structures are vascular rings. They encircle trachea and esophagus at variant degrees which results in respiratory or feeding difficulties in children. An abnormality of pulmonary arterial system results in airway compression is known as a sling. CT gives excellent spatial and temporal resolution cona a wide field of view, multi planer reconstruction capabilities and simultaneous evaluation of the airway. A brief discussion of the embryology of the branch vessels and aorta is followed by discussions and illustrations of some uncommon and common vascular rings with critical informations required by surgeons.

Introduction

Vascular rings are (<1%) congenital abnormalities of aortic arch-derived vascular or ligamentous structures that enclose the trachea and esophagus to varying degrees [1-2]. A sling is anomaly of pulmonary arterial system results in airway compression. The clinical presentation is variable since vascular rings and slings may be challenging to diagnose [3-6]. They can be asymptomatic with respiratory symptoms like respiratory distress, stridor, seal-bark cough, apnea, cyanosis or recurrent infection typically in first year of life. Feeding difficulties like dysphagia slow feeding and hypertension of head while eating may present later in life still liquid diets are tolerated earlier and weight may be found [7-9]. Some abnormalities present later in life with feeding difficulties during pregnancy and when there is ectasia os vessels. Cough, stridor, wheezing, tachypnea, noisy breathing and subcostal retractions may be apparent while examinations. Vascular rings may be associated with congenital anomalies, particularly conotruncal abnormalities tetrology, transposition, truncus and syndromes including deletion [3-8]. There is no gender, ethnic and geographic predictions. Symptomatic vascular rings are resection in the first year of life to avoid complications like hypoxic spells, sudden death, aneurysm, dissection and erosion of the aorta into the trachea or esophagus [8]. Imaging plays an important role in the evaluation and management of vascular rings. In this review the current role and techniques of ct in the elevation of vascular rings discuss. Some common and uncommon vascular rings with its critical information required for surgeons are discussed and illustrated in the review of embryology of the aorta [9-10]. The innominate artery compression syndrome (IACS) and pulmonary artery are the other two syndromes of tracheoesophageal compression. IACS acquires at abnormal distal and posterior origin of the innominate artery and the trachea gets compressed anteriorly when it courses from left to right arm of the mediastinum [10]. When the left pulmonary artery originates from right pulmonary artery and encircles the distal trachea, the pulmonary artery sling occurs. Also the right main stem branches as it courses between trachea and esophagus to the left lung [11].

Incidence

In the vascular ring the anomalous left pulmonary artery or pulmonary artery sling makes about 10% of the cases [10]. The pulmonary artery sling arises from an anomaly of the sixth brachial arch and produces a complete ring. This anomaly is linked with intra cardiac defects in 10 to 15% of the cases.

Ontogenesis for the structures forms the Type-I Vascular ring

These embryonic structures form during the enlargement of the arterial system in intrauterine life. An aortic arch is a division from the arterial aortic sac to the dorsal aorta [11]. It travels in the centre of each pharyngeal arch, surrounded in mesenchym. Initially, there are five pairs of arches, but these undergo structural changes and variance to form the definite vascular patterns for the head and neck, aorta, and pulmonary circulation [12-14]. During the fourth and fifth weeks of embryological growth, when the pharyngeal arches form, the aortic sac gives rise to the aortic arches [12]. The aortic sac is the endothelial ruled opening just distal to the truncus arteriosus; it is the primordial vascular channel from which the aortic arches rise. Each pharyngeal arch has its own cranial nerve and its artery [13]. Hence, we can determine that the growths of the aortic and pharyngeal arches are very closely associated. The aortic arches dismiss in the right and left dorsal aortae. The dorsal aortae remain paired in the division of the arches, though below this region they fuse to form a single vessel (the dismiss in the right and left dorsal aortae. The dorsal aortae remain paired in the division of the arches, though below this region they fuse to form a single vessel (the descending/thoracic/abdominal aorta) [15].
The pharyngeal arches and their vessels seem in a cephalo-caudal order, so they are not all existing at the same time. As a new arch form the aortic sac donates a branch to it. In the early stage, there are pairs of aortic arches, which are numbered I, II, III, IV, and VI. This system becomes changed in further development [16].

The aortic-pulmonary septum splits the truncus arteriosus into the ventral aorta and pulmonary trunk. They indicate the discharge channels of the heart [17]. After this, the aortic sac then forms right and left horns. The brachiocephalic artery is the right horn and the proximal part is the left horn of the aortic arch. On the day 27, the 1st and 2nd Arch start to regret by approximately. However, parts of each persist as the maxillary artery, as hyoid and stapediales arteries, respectively [18]. By day 29 both these arches completely dissolve. Around the time reversion of the 1st and 2nd arches, the 3rd is large and 4th and 6th arches are forming. Soon the 3rd, 4th, and 6th arches all seem large [19]. Because of separation of the truncus arteriosus, the 6th arches are now continuous with the pulmonary trunk, with the primitive pulmonary artery existing as a major branch. The pulmonary arteries initiate from the truncus arteriosus and also from the sixth pharyngeal arch. The truncus arteriosis is a structure that procedures during the development of the heart as a replacement to the conus Arteriosus [20-23].

The endocardial tubes have established a swelling in the part nearby to the heart, by the third week of embryological life [23]. The swelling is known as the bulbus cordis and the upper part of this swelling change into the truncus arteriosus. The structure is eventually mesodermal in origin. During development of the heart, the heart tissue feels the folding, and the truncus arteriosus is visible to what will finally be both the left and right ventricles. As a septum grows between the two ventricles of the heart, two bulges form on either side of the truncus Arteriosus [24]. These progressively increase until the trunk splits into the aorta and pulmonary arteries. The ductus arteriosis connects the pulmonary trunk and the arch of aorta, allowing blood to bypass the lungs during the embryological life [25].

Fig 1. Primitive Pharyngeal arch left (L) and right (R) external carotid (EC), internal carotid (IC) pharyngeal arches – 4th (IV) and 6th (VI) pharyngeal arches Dorsal Aortas (DA), 7th intersegmental arteries (VII).

At the early manifestation of the respiratory diverticulum, the Pair of bronchial buds appears at its end [26]. And then it seems that the precursors of the windpipe and lung buds are derived from different source of cells, and that the lung buds give rise the bronchi and distal respiratory tree [27]. The straight portion of the respiratory diverticulum is the primordial of the wind pipe. Then the bronchial buds ultimately becomes the primary bronchi, give rise to an additional buds on the right side and 2 on the left side [26-28].

Then these buds develops and becomes the secondary, or stem bronchi, their number signify the formation of the 3 lobes of the right side lung and 2 lobes of the left side lung. From that point each secondary bronchial buds undergoes a long series of branching during embryonic and fetal life [29]. Then the mesoderm surround the endoderm controlling the extent of branching within the respiratory tract. Abundant tissues recombination experiments have shown that the mesoderm surrounds the wind pipe and inhibits its branching, the mesoderm surrounds the bronchial buds promotes branching [30]. If the tracheal endoderm is joins with the bronchial mesoderm, anomalous budding is induced. On the other hand, tracheal mesoderm placed around the bronchial endoderm which inhibits the bronchial budding [31-35]. The mesoderm of the certain additional organ, such as the salivary glands, can promotes the budding of the bronchial endoderm. The pattern of branching characteristic of the mesoderm is induce [34]. The mesoderm is capable of promoting or supporting budding must maintain a high rate of production of epithelial cells. Generally, the pattern of epithelial organs is largely separation of the epithelium is a definite property of the epithelial cells, but the epithelial phenotype corresponding to the regions occupies by the mesoderm [35]. By week eight (28-30mm embryo), mesenchyme essential rudiments of the sixteen to twenty tracheal cartilages are seen and the next two weeks, the masses formation cartilage begins cranially and extending caudally. At the same time fibroblastic tissue of the tracheal wall arising from the mesenchyme between the cartilages a then posteriorly between the ends of the embryonic rings of smooth muscles arises [36]. Cilia appears at the tenth week of the development (51-53mm embryo). By the 12th week of development the mucosal glands are seen and developed in a crania caudal direction. By the end of the 20th week development, all major microscopic features of the trachea are visible. It is short and narrow while the larynx is relatively long [37].

Ontogenesis for the abnormal Type-1 Vascular ring
Abnormal development of Double aortic arch:
The double aortic arch is a rare abnormality caused by perseverance of the fetal double aortic arch system. The ascending aorta splits into two arches that passes to either side of the esophagus, trachea and reunify to form the descending aorta [36-40]. Consequently, it is a form of complete vascular ring, resulting in no cardiac disease, but rarely related with intracardiac defects. The descending aorta is regularly on the left side. Most commonly, one arch is leading, whereas the other may be of small ability or signified by a fibrous band [40].

Fig 2. Segments of the pharyngeal arch system that regress (shown in black) in the normal formation of the thoracic great arteries. (Left pulmonary artery (LPA); right pulmonary artery (RPA); subclavian artery (SCA); right common carotid arch(R CCA); left common carotid arch (L CCA)).
Abnormal development of Right aortic arch with retro esophageal component

The left fourth branchial arch involves and right leftovers, a right aortic arch is existent. Right aortic arch occurs less commonly than 1 in 100,000 times in the general people and may be in the absence of any other abnormalities [41]. Its presence is indicative of the existence of an associated abnormality. About 30% of patients with tetralogy off allot have an related right aortic arch. Perseverance of the right arch with involution of the left generates a state in which the beginnings of the left subclavian artery and ductus arteriosus can differ. Several of these formations can produce a vascular ring [42-45].

Abnormal development of Mirror-image branching with retro esophageal ligamentum arteriosum:

Sometimes described as the most mutual, secretarial for up to 59% of all right sided arches. In most of the nonfiction it is less common than Type-II [46]. Occurs from disruption of the dorsal subdivision of the left arch between the left subclavian artery and the descending aorta, with reversion of the right ductus arteriosus in the hypothetical double aortic arch [44]. Usually associated with cyanotic congenital heart disease which include

1. Tetralogy of Fallot, 2.truncus arteriosus, 3.Tricuspid atresia, 4.Transposition of the great arteries.

Abnormal development of Retro esophageal left subclavian artery with ligamentum arteriosus:

A section of thoracic aorta (about 6 cm) around the aortic diverticulum was resected, and the proximal and distal ends of the aorta were anastomosed openly, so that the thoracic aorta would be at significant distance from the vertebral column and the brachial plexus [46]. Kommerell’s diverticulum is the main part of the vascular ring, it was included in the resected section of thoracic aorta. Right thoracotomy is the favored method for this lesion, because it gives outstanding contact of the ascending aorta through to the descending aorta [47].

Abnormal development of cervical aortic arch complex:

The cervical aortic arch mentions to an abnormally high position of the aortic arch in the low or mid neck region. This infrequent type of aortic arch abnormality is supposed to result from perseverance of the third aortic arch and reversion of the normal fourth arch [50]. Anomalies of brachiocephalic arterial branching and arch laterality are common in patients with a cervical aortic arch. There is no suggestion with congenital heart disease, and the irregularity occurs most often in suggestion with a right aortic arch. Most of the patients with this abnormality are asymptomatic, but signs of dysphagia and respiratory distress due to the density by the vascular ring
Vascular ring anomalies causing tracheoesophageal firmness cover 1-3% of all congenital cardiac abnormalities. The reported occurrence of respiratory symptoms and signs in patients with vascular rings is 70-95% and that of gastrointestinal signs 5-50% [54]. Patients with severe firmness incline to present symptoms earlier in life. More than half the patients showed indications during the newborn period and respiratory symptoms were more protruding. The most public respiratory and gastrointestinal symptoms were alike to those found in previous studies. Some patients may show signs much later in life and some remain asymptomatic through their entire life [55-57].

The occurrence of mutual pediatric symptoms connected with tracheoesophageal firmness should aware us to the prospect of a vascular ring. The diagnosis should be made instantly because patients with any type of vascular rings are at hazard of life-threatening difficulties (8.6% of our patients) such as respiratory arrest or apnea [58].

As shown in earlier studies, chest radiography and barium esophagography are main for not including other causes of mutual respiratory symptoms [59]. In our learning, chest radiography was done in four patients and barium esophagography in twelve patients. But these modalities were of lesser reputation to the diagnostic process associated to Echo, cardiac CT or MRI [60].

Echocardiography is a noninvasive, certainly obtainable, and significant diagnostic modality to correctly assess anatomy and eliminate other intracardiac abnormalities. It is imperative to recognize connected cardiac abnormalities because they are quite frequent and related with a poor prognosis [61]. However, Echo has its limits for exactly recognizing vascular structures in these abnormalities. The diagnostic procedure has been assisted by cardiac catheterization in the past [62]. Currently, CT angiography is a significant modality used in our center. It exactly defines the type of abnormality in most patients and directly shows the connection of the arch to the trachea and bronchi [63].

In our study, the middle interim between diagnosis and surgical procedure was only 6 days and the time between indications and surgery was about 2 months [64-68]. The intermission between symptoms and surgery was mainly reliant on the time between indications and diagnosis, because we quickly did surgical repair after authorizing the diagnosis [69]. We predictable that early reparation (surgery within 1 month of age) would lead to better mortality, yet no association was found between early repair and mortality. Three patients died after operating repair and two of them were asymptomatic at the time of optional operation for complex heart syndrome [70-72]. The main reasons of death were postoperative difficulty such as sepsis and respiratory disaster.

Overall prediction for a vascular ring was comparatively good in this study; however vascular rings shared with complex heart syndrome showed a deprived prognosis [73].

### Conclusion

Vascular rings contain some types of abnormalities, each with different indications and projection. The occurrence of complex heart syndrome was suggestively accompanying with mortality. Surgical reparation for a vascular ring has a comparatively good prognosis and the operative danger in the nonappearance of complex heart syndrome is low [74]. Consequently, early diagnosis and well-timed surgery in indicative patients are dominant. In difference, patients with slight symptoms can be accomplished medically with close follow-up [75].

### References


