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A Study of Anomalies of Great Vessels of Heart

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Abstract:

Introduction: Abnormalities of great vessels of heart, especially arch of aorta, are of embryological interest and have already been described in many text books of anatomy. The embryological development of aortic arch is very complex that finally results in the formation of three branches: 1) brachiocephalic trunk 2) left common carotid artery and 3) left subclavian artery. In the present study we have found a case of abnormal aortic arch giving rise to four branches and also ligamentum arteriosum. We also found three more cases of ligamentum arteriosum.

During the foetal life, Ductus arteriosus provides communication between pulmonary and systemic circulations. This normal anatomical structure closes soon after birth and undergoes fibrosis which is termed as ligamentum arteriosum that persists up to 3 months of post natal life.

Aims and Objectives: The clinicians and surgeons should be aware of wide range of congenital anomalies of great vessels of heart during the management of such cases. Therefore the present study focuses on reporting of such cases to reduce the risk of iotrogenic injuries.

Materials and Methods: We had studied 80 embalmed cadavers for a period of 3 years for the presence of anomalous great vessels of heart.

Results: We have found four hearts with anomalous great vessels. One of the hearts had abnormal aortic arch that had given rise to four branches. It also had persistent ligamentum arteriosum. In the other three cases, we found only persistent ligamentum arteriosum.

Conclusion: Knowledge of variations in great vessels of heart is important for the practitioners to reduce the disastrous vascular complications.

Keywords: Great vessels; Ligamentum arteriosum; anomalies.

Introduction

The development ofaortic arch is very complex and finally results in the formation of left aortic arch with three branches. They are 1) brachiocephalic trunk 2) left common carotid artery and 3) left subclavian artery. 1–3% of congenital heart diseases account for anomalies of aortic arch. When the arch crosses the right main bronchus, it is called as right aortic arch and further descends to the right side of the spine. This variation does not cause oesophageal compression. But with the presence of both Kommerell's diverticulum and ligamentum arteriosum, oesophagus may get compressed resulting in dysphagia. Patients usually remain asymptomatic till later decades of life as the arteries become tortuous and aneurysmal.[1]

A recent study suggests that only 0.36% of patients with dysphagia had aberrant subclavian arteries.[2] An aberrant right subclavian artery is found in 0.5% of population, whereas aberrant left subclavian artery is found in only 0.06–0.01% of population.[3] The right-sidedaortic arch is a rare congenital malformation occurring during embryologic development. Majority of these cases

present with aberrant left subclavian artery (LSA) which can cause compression of thoracic structures. While this aberrant vessel causes mostly benign symptoms, patients may first present with rupture of a thoracic aneurysm. This thoracic aneurysm is commonly known as Kommerell's diverticulum. Kommerell's diverticulum is a developmental anomaly named after Dr. Kommerell who diagnosed it first. It is a remnant of fourth dorsal aortic arch. The diverticulum may be present on both right and left aortic arches, from which aberrant subclavian artery arise to the contralateral side.[4]

Embryological aspects

The various congenital anomalies of the aortic arch is based on the hypothetical theory of double aortic arch, which was described by Edwards et al.[5] In this hypothetical theory, the ascending aorta divides into Right and Left aortic arches which encircle trachea and esophagus and then unite to form descending aorta. A common carotid artery and subclavian artery arise from each aortic arch. A ductus arteriosus connects the aortic arch and pulmonary artery on each side. Usually, the regression occurs between the origin of right subclavian artery and descending aorta in the Right aortic arch (RAA). Right ductus also undergoes regression. Thus, the normal aortic arch is formed. If the regression occurs between the origin of left subclavian artery (LSCA) and the descending aorta in the left aortic arch, it then becomes an RAA with mirror-image branching. If the interruption takes place between the left carotid artery and the LSCA in the left aortic arch, it will form an RAA with aberrant LSCA.[6] There can be a dilated segment at the proximal part of the aberrant LSCA. It is known as the diverticulum of Kommerell. Thus depending on inappropriate persistence orregression of different segments, different types of aortic arch anomalies occur.[6]

Materials and Methods

The present studywas conducted on 80 embalmed cadavers, out of which 19 were female and 61 were male, for the period of 3 years from 2015 to 2018 in KAHER's Jawaharlal Medical College, Belagavi, Karnataka. All the 80 cadaveric hearts were observed during the routine dissection hours of MBBS students and presence ofany abnormalities or congenital anomalies of great vessels of heart were noticed.

We found an anomalous heart in a 60 year old male cadaver with the ascending aorta originating from the left ventricle posterior to the commencement of pulmonary artery (Fig. 1). It travels cranially and to the left and became continuous with the arch of aorta. The arch passes posterior to the trachea and oesophagus becoming continuous with the descending thoracic aorta which was on the right side of vertebral column. This anomaly is because of persistence of right fourth aortic arch and regression of left fourth aortic arch.

The right sided aortic arch gave four branches: from right towards the left they were 1) right subclavian artery 2) right common carotid artery 3) left common carotid artery and 4) aberrant left subclavian artery. The aberrant left subclavian artery was arising from a conical dilatation of the proximal portion of its origin from the aorta. This conical dilatation is called as Kommerell's diverticulum which accounts for 1.25% in the present study.

Three more adult male cadavers had only persistent ligamentum arteriosum with no other associated anomalies which accounts for 3.75% (Fig. 2a, b, c).

Fig. 1: Anomalous Heart

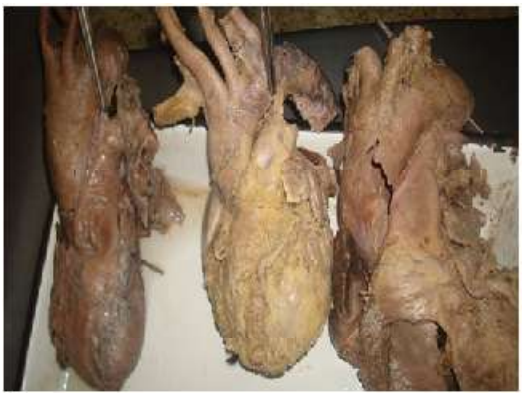

Fig 2a, b, c: Hearts Showing Ligmentum Arteriosum.

Results

Among 80 dissected cadavers, we got four anomalous hearts. All these were found in adult male cadavers in the age group of 55–70 years. One of hearts (Fig. 1) had right sided arch of aorta giving rise to four branches including aberrant left subclavian artery. The rest of the three hearts had persistent ligamentum arteriosum with no other anomalies (Fig. 2a, b, c).

Discussion

In 1763, Fioratti and Aglietti first documented a case of right sided aortic arch.[7] This is later classified in 1948 by Edward, in 1963 by Palayew and in 1964 by Steward et al.[8]

According to a study done by Cina CS, et al. in 2009, only 0.1% of adult population was found with Right sided aortic arch which is a very rare congenital anomaly.[9] Half of these cases were associated with aberrant left subclavian artery (0.05–0.1%) which has been found in our case as well. Only about 50 cases of Right sided aortic arch with aberrant left subclavian artery have been found in the previous studies so far.[10]

Right aortic arch (RAA) is divided into two types: mirror-image branching and aberrant left subclavian artery (LSCA).[11] RAA with aberrant LSCA is rarely associated with other congenital heart diseases, which accounts for only about 10%, where as in RAA with mirror-image branching, this risk of association with other congenital heart disease is more than 90%.[12] The most common association is Tetralogy of Fallot. Hastreiter et al. in his study, found that the RAA with mirror-image branching was associated with double-outlet right ventricle in 20%, with truncus arteriosus in 13–15% cases and with Tetralogy of Fallot in 13–34% of cases.[7]

The etiology of right sided arch of aorta is still unknown. A deletion of chromosome 22q11 is said to be associated with 24% incidence of isolated anomalies of laterality of aortic arch.[13]

Right sided aortic arch with aberrant left subclavian artery is rarely associated with other congenital heart diseases and accounts for only 10%.[14]

Jung et al. in 1978 reported five cases of right sided aortic arches with aberrant left subclavian artery and persistent leftligamentum arteriosum.[15]

Kommerell's diverticulum is also called as "remnant diverticulum", "lusoria diverticulum"or "lusoria root". This diverticulum was originally described by Burckhard Friedrich Kommerell in 1936 and is a remnant of left fourth aortic arch.[16]

Two types of aortic arches have been described. In the first type, a vascular ring is formed around the trachea and oesophagus.[17] Bothare enclosedin the ring formed by the right sided aortic arch, and left ligamentum arteriosum. Similar formation has also been found in our study as well. In the other type, both the ductus arteriosus and aorta lie to the right of the trachea without a vascular ring.[14]

Vascular rings are formed by double aortic arches with small, atretic or equal right and left components. The descending thoracic aorta may be left or right sided, and there may be a left, right, or bilateral patent ductus arteriosus, or a ligamentum arteriosum.[18,19]

In the previous studies, it has been reported that right sided aortic arch with aberrant left subclavian artery must always have a ductus arteriosus.[20] Similar findings have been noted in our studies as well. With the presence of ductus arteriosus, a vascular ring is formed which may or may not cause compression of trachea and oesophagus.

Conclusion

The branching pattern of great vesselsthat are arising from the arch of aorta are of major interest to the cardiologists, cardiothoracic surgeons and clinicians who perform imaging studies and catheter-based techniques to reduce the risk of iotrogenic injuries. It helps them to appreciate these anomalies pre-operatively and can reduce the risk of disastrous vascular complications.

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