ANTERIOR WALL MYOCARDIAL INFARCTION IN A PATIENT WITH RIGHT AORTIC ARCH WITH RETROESOPHAGEAL DIVERTICULUM OF KOMMERELL - CASE REPORT

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Abstract:
Right Aortic Arch with Retroesophageal Diverticulum of Kommerell is a rare anomaly. We report a case of Anterior wall myocardial infarction in a patient with such an anomaly. Myocardial infarction was managed with PTCA to Mid LAD. Right Aortic Arch with Retroesophageal Diverticulum of Kommerell was managed conservatively. A brief review of literature is discussed.

Keyword:
Anterior wall myocardial infarction, Right Aortic Arch, Retroesophageal Diverticulum of Kommerell, PTCA Stenting.

CASE REPORT
39 years old male was admitted for chest pain since 3 days. Chest pain was typical angina pain. Retrosternal, oppressive, radiating to both shoulders, associated with sweating, vomiting and giddiness. There was no history of breathlessness, palpitations, syncopal attacks, stridor, cough, wheeze, dysphagia. There was no history of similar illness in the past. There was no history of Type 2 Diabetes mellitus, systemic hypertension, COPD. He was a smoker and alcoholic. There was no Family history of Myocardial infarction.
Physical examination revealed normal stature male, not anaemic, not jaundiced, no cyanosis, no clubbing, no bilateral pedal edema, no lymphadenopathy. Pulse 76/min, regular, normal volume, no specific character, felt in all peripheral vessels, no radio-femoral delay. BP 110/80 mm Hg in right upperlimb in supine position. Cardiovascular examination showed normal JVP, Apical impulse felt in left 5th intercostal space just medial to mid clavicular line, S1S2 were normal, no added sounds, no murmur. Abdominal, Respiratory, CNS examination were normal. X-Ray showed Right Aortic Arch. ECG taken showed ST elevation from V1-V4. ECHO showed Akinesia of Anterior wall & Anterior septum at basal, mid ventricular & apical levels with LVEF-40%. He was diagnosed as suffering from Acute Anterior wall myocardial infarction. He was not thrombolysed due to late presentation. He was started on anticoagulants, antiplatelets, anti-ischemic medications. Coronary angiogram done three days later showed Single vessel disease Mid LAD showing 95% long segment lesion filled with Thrombus. Aortic root angiogram done showed Right sided aortic arch with Left Subclavian artery arising aberrantly from Kommerell's diverticulum arising from Descending Aorta. Following were the branches of the arch. 1st: Left common carotid artery. 2nd: Right common carotid artery. 3rd: Right Subclavian artery. Left Subclavian artery arising aberrantly from Kommerell's diverticulum arising from Descending Aorta.
PTCA stenting was done to Mid LAD lesion. Patient improved symptomatically. Right aortic arch with retroesophageal diverticulum of kommerell was managed conservatively.

**DISCUSSION:**

**Right Aortic Arch**

Types

- At least five different types
- Only two of importance

Mirror Image Type — Type I
Aberrant left subclavian — Type II

General

Recognized by leftward displacement of barium-filled esophagus

Of air-filled trachea

Aortic knob is absent from left side

Aorta descends on right

Para-aortic stripe returns to left side of spine just above diaphragm

Mirror-image type almost always has associated CHD

Usually Tetralogy of Fallot

Aberrant Left Subclavian type rarely has associated CHD

Most common variety of right arch

Type 1—Mirror Image Type

Secondary to interruption of left arch just distal to ductus arteriosis

Associated with congenital heart disease 98% of time

X-ray Findings

No posterior impression on trachea or barium-filled esophagus

Heart is usually abnormal in size or shape

Aorta descends on right

Type II—Aberrant Left Subclavian

Secondary to interruption of left aortic arch between LCC and LSC arteries

Associated with cardiac defects 5-10% of the time

Tetralogy of Fallot most often (71%)

ASD or VSD next most often (21%)

Coarctation of aorta rarely (7%)

Anomalous left subclavian artery (retroesophageal and retrotracheal)

Aorta descends on right

X-ray Findings

Posterior impression on trachea and barium-filled esophagus

Heart is usually normal in size and shape

Aorta descends on right

If there is a mirror-image right aortic arch, then

90% will have Tetralogy of Fallot

6% with Truncus Arteriosis

5% with Tricuspid Atresia

If the person has the following lesions, then the association with a mirror-image arch is

Truncus arteriosis 33%

Tetralogy of Fallot 25%

Transposition 10%
Tricuspid atresia 5%

VSD 2%

Aortic arch anomalies Classifications Edwards

Classification:

Anatomical Classification
1. Abnormalities of branching,
2. Abnormalities of arch position including right aortic arch and cervical aortic arch, supernumerary arches including double aortic arch and persistent fifth aortic arch,
3. Interrupted aortic arch,
4. Anomalous origin of a pulmonary artery branch from the ascending aorta or from the contralateral pulmonary artery branch.

Clinical Classification
1. Vascular rings; nonring vascular compression of the trachea, bronchi, or esophagus;
2. Noncompressive arch malformations;
3. Ductal-dependent arch anomalies including interrupted aortic arches and 4. Isolated subclavian, carotid, or innominate arteries.

Left and Right arch Definition
Left and right aortic arch refer to which bronchus is crossed by the arch, not to which side of the midline the aortic root ascends. Practically, the sidedness of the aortic arch is usually determined indirectly with echocardiography or angiography by the branching pattern of the brachiocephalic vessels. As a rule, the first aortic vessel contains the carotid artery opposite the side of the arch.

Right Aortic Arch
Right aortic arches have in common (by definition) a single aortic arch that crosses over the right mainstem bronchus, passing to the right of the trachea. There are four major types of right arch:
(a) with mirror image branching,
(b) with retroesophageal left subclavian artery,
(c) with retro-esophageal diverticulum, and
(d) with left descending aorta.

There are also several infrequently occurring variations. The incidence of right aortic arch among patients with tetralogy of Fallot has been reported to be anywhere from 13% to 34% (35). The incidence in truncus arteriosus is generally higher than in tetralogy.

Right Aortic Arch with Mirror Image Branching:
Mirror-image right arch has the first branch as a left innominate artery, which, in turn, divides into left carotid and left subclavian arteries; the second as the right carotid; and the third a right subclavian. This is the left-right mirror of a normal left aortic arch. However, frequently that is the end of the symmetry, since the ductus arteriosus (or ligamentum arteriosum) is usually the left-sided one, arising from the base of the innominate artery rather than from the aortic arch. Therefore, typical mirror-image right aortic arch with left ductus or ligamentum does not form a vascular ring. This arch anomaly is almost always associated with congenital intracardiac disease. The most common association is with tetralogy of Fallot,
but truncus arteriosus communis, other conotruncal anomalies including transposition of the great arteries, double-outlet right ventricle, right ventricular aorta with pulmonary atresia, and anatomically corrected malposition were also seen. Embryology: The presumptive pattern of mirror-image right arch development includes dissolution of the left dorsal aorta distal to the origin of the seventh intersegmental artery (distal subclavian artery precursor) so that the left fourth arch becomes the proximal subclavian artery rather than remaining as an aortic arch. Typically, the right sixth (ductal) arch involutes while the left sixth persists.

Diagnosis and Management: Since this type of right arch usually produces no retro-esophageal compression or vascular ring, there are, with rare exception, no symptoms produced by the arch itself. Therefore, the diagnosis is usually made during imaging of the associated congenital intracardiac disease. No treatment of right aortic arch per se is required.

**Right Aortic Arch with Retroesophageal Diverticulum of Kommerell:**

Right arch with diverticulum of Kommerell is the second most common vascular ring after double aortic arch. It is far more common than its mirror image described above. It has as its first branch the left carotid artery, second the right carotid artery, third the right subclavian artery, and finally, a retroesophageal vessel from which the left subclavian artery arises and the left ductus arteriosus or ligamentum arteriosum connects. This combination of vessels produces a vascular ring. Many people with this arch anomaly are asymptomatic and therefore go unrecognized except for incidental discovery. If the ductus arteriosus is patent, it is easy to understand how one might visualize the complete ring formed by the aortic arch on the right, the pulmonary artery anteriorly. However, if the ductus is closed, it is not intuitive that one could distinguish this from the more benign right arch with retroesophageal subclavian artery (nonring) discussed below. The difference is the diverticulum of Kommerell, which is a much larger vessel than the subclavian artery itself. Typically, the origin of the diverticulum is equal in diameter to the descending aorta and tapers to the subclavian artery caliber at the site of juncture with the left ligamentum.

Embryology: Disappearance of the left fourth embryonic arch with persistence of the left sixth (ductal) arch between the truncoaortic sac (pulmonary artery precursor) and left dorsal aorta accounts for the findings in this arch anomaly. Note that the left dorsal aorta is not merely the proximal left subclavian artery but is also the continuation of the left sixth aortic arch leading to the fused dorsal aortae.

Diagnosis and Management: The presentation in symptomatic patients is usually that of a vascular ring. With that history, the appearance of a right aortic arch on a plain chest roentgenogram should raise the question of this anomaly and prompt further, more specific evaluation. Barium esophagram reveals a large posterior indentation on the esophagus. Echocardiography will show the left carotid artery arising alone as the first arch vessel, but definitive diagnosis requires that the diverticulum be followed out to the point at which the caliber changes to that of the smaller subclavian artery. Angiography demonstrates the characteristic branching pattern, but more important, shows the abrupt change in caliber from diverticulum to subclavian artery. Magnetic resonance imaging is ideal for making this diagnosis, being noninvasive and having
airway structures. Axial or transverse imaging demonstrates the aortic arch to the right of the trachea and the diverticulum posterior to it. Most people with this arch anomaly are asymptomatic. Treatment is surgical division of the ductus or ligamentum in those patients who are symptomatic. This is usually performed through a left thoracotomy, although a median sternotomy approach is preferred by some. Still others advocate not only division of the ligamentum but resection of the diverticulum and transfer of the left subclavian artery to the left carotid artery because of the recurrence of symptoms in some patients owing to compression from an aneurysmal diverticulum and traction by the subclavian artery. In those patients undergoing surgery for another lesion such as an intracardiac abnormality or repair of tracheoesophageal fistula, even an asymptomatic ligamentum should be divided. With esophageal atresia, a symptomatic ring may be avoided when repairing the esophagus outside the ring.

**Right Aortic Arch with Retroesophageal Left Subclavian Artery:**

Right aortic arch with retroesophageal (also called anomalous or aberrant) left subclavian artery consists of an arch passing to the right of the trachea with the following sequence of brachiocephalic arteries: Left carotid, right carotid, right subclavian, and retroesophageal left subclavian. This differs from the previous arch in that the proximal left subclavian artery is not significantly larger in caliber than its more distal portion (i.e., no aortic diverticulum). Therefore, there is no left-sided ductus arteriosus or ligamentum arteriosum and thus no vascular ring. Many of these patients have associated conotruncal anomalies. As has been noted previously, aberrant subclavian artery has a higher incidence of 22q11 deletion than does mirror-image right aortic arch.

**Embryology:**

This is similar to the right arch with retroesophageal diverticulum with involution of the left fourth aortic arch but with loss of the left sixth (ductal) arch. Diagnosis and Management: The diagnosis may be suspected from barium esophagography with a relatively small posterior indentation on the esophagus passing upward to the left. Since there is no vascular ring, the trachea is unaffected except for the slight leftward deviation seen with virtually all right aortic arches. Echocardiography may be expected to identify the first branch of the aorta as a left carotid artery (since it does not bifurcate proximally as an innominate artery does and is similar in caliber to the second branch—right carotid artery), but appreciation of the size of the retroesophageal vessel itself may be difficult. Both magnetic resonance imaging and angiography can demonstrate the size of the left subclavian artery, which distinguishes this lesion from right aortic arch with retroesophageal diverticulum. Since there is no vascular ring, there is usually no need for treatment other than that of associated anomalies.

**Right Aortic Arch with Left Descending Aorta and Left Ductus Arteriosus or Ligamentum Arteriosum:**

Right arch, left descending aorta, also known as right aortic arch with retroesophageal segment or circumflex right aortic arch, is similar in presentation to right arch with retro-esophageal diverticulum (above), but it is less common. Unlike cases with retroesophageal diverticulum in which the aorta, after passing over the right mainstem bronchus, descends for some distance on the right, then gradually crosses to the left before reaching the level of the diaphragm, right arch left descending (aorta) has the aortic arch itself cross the midline to the left at the
level of the T4 or T5 vertebral body, at which point it gives rise to the left ductus arteriosus or ligamentum arteriosum. The first arch vessel may be the left innominate, followed by right carotid and right subclavian, or the left carotid artery alone, followed by the right carotid, right subclavian, and finally the left subclavian. However, it is the aortic arch that is retroesophageal, not the subclavian artery or an aortic diverticulum.

Embryology: Two forms exist with dissolution of either the left dorsal aorta distal to the take-off of the left subclavian artery or the left fourth arch. The distal portion of the definitive arch is composed of the retroesophageal right-sided dorsal aorta. The persistent left sixth arch connects to the left-sided dorsal aorta, completing a vascularring.

Diagnosis and Management: The findings on chest roentgenogram and barium esophagography may be similar to those in right arch with retroesophageal diverticulum. Differences include a downward to the left instead of upward to the left orientation of the esophageal indentation. Furthermore, in some cases the descending aorta itself can be seen on the left of the spine instead of the right, but this is not a consistent finding. When associated with a hypoplastic arch, this anomaly can be mistaken for interrupted aortic arch. With a projection image such as angiography, it may not be clear whether the aorta passes anterior to the trachea as it courses from right ascending to left descending, as is seen with a normal left aortic arch, or posterior to the trachea, i.e., a right aortic arch with left descending. Magnetic resonance imaging can avoid some of the pitfalls seen with projection images and can delineate the entire aorta, not only in its normal rightward ascending and leftward descending segments, but definitively in its relationship to the trachea. Division of the vascular ring is indicated when patients are symptomatic, although these are typically loose rings. However, an adult with dysphagia may require more than simple division of the ligamentum. Actual division of the aortic arch with mobilization of the retroesophageal portion and reanastomosis of ascending and descending aorta using a tube graft may be necessary to relieve the esophageal compression.

Right Aortic Arch with Retroesophageal Innominate Artery:

Right aortic arch with retroesophageal (or aberrant) innominate artery is another rare abnormality of the aortic arch system. Contrary to the general rule that the first arch vessel contains a carotid artery contralateral to the aortic arch, in these cases the sequence of brachiocephalic vessels is right carotid, right subclavian, retroesophageal left innominate artery. The ductus arteriosus or ligamentum arteriosum completes a vascular ring as it connects the left pulmonary artery with the base of the so-called innominate artery. Embryology: The apparent site of arch dissolution is in the left branch of the truncus aorticoaortic sac. Thus the arch is formed by the right branch leading to the right fourth arch, which, in turn, connects to the right dorsal aorta. The left dorsal aorta supplies the left seventh intersegmental artery (distal left subclavian) and the left third arch (common carotid artery). It is not clear whether any remnant of the left fourth arch persists, although it may form the proximal left carotid artery. Diagnosis and Management: Tracheal compression seems to be the rule, although the degree of symptomatology varies considerably. The important anatomic clues to the diagnosis by any imaging modality are the presence of a single carotid artery arising from the proximal aorta. The other anomalies with that finding are also rare:
Interrupted aortic arch with interruption between the two carotid arteries and isolated left carotid or innominate artery. The differentiating feature is the presence of a normal-sized (right) aortic arch (missing in arch interruption) and the distal origin of the carotid artery from that arch (not present with isolated carotid or innominate). If symptomatic from the vascular ring, division of the ductus or ligamentum is in order. Conceivably in the adult, detachment of the innominate artery from the distal arch and reimplantation in the ascending aorta might be necessary based on similar arch anomalies mentioned above in which the retroesophageal vessel continued to cause dysphagia even after relief of the ring by division of the ligamentum.

**Right Aortic Arch with Isolation of Contralateral Arch Vesse:**

Isolation of brachiocephalic vessels is relatively uncommon. The term isolation means that the particular vessel arises exclusively from the pulmonary artery via the ductus arteriosus (or ligamentum) but without connection to the aorta. Three different forms have been noted: Isolation of the left subclavian artery, isolation of the left carotid, and isolation of the left innominate artery. Isolated left subclavian is by far the most common of the three.

**Embryology:**

All cases of isolated arch vessels derive from two ipsilateral breaks in the aortic arch system. In isolated subclavian artery, the distal left dorsal aorta involutes after cephalad migration of the left seventh intersegmental (subclavian) artery to the level where left sixth (ductal) arch normally joins the proximal dorsal aorta. This together with involution of the left fourth arch leaves the subclavian isolated from the aortic arch but connected to the pulmonary artery via the ductus.

**Diagnosis and Management:**

Cases of isolated brachiocephalic arteries may have diminished pulses or lower blood pressure in the affected artery. When the subclavian and vertebral arteries are involved, the possibility of subclavian steal syndrome exists in which blood flows down the vertebral artery into the subclavian, particularly when the arm is exercised. If the ductus remains patent, pulmonary artery steal can occur with flow down the vertebral artery through the ductus into the low-resistance pulmonary artery (47). The diagnosis should be suspected in any patient with right aortic arch and diminished pulse amplitude or blood pressure in the left arm. Contrast injection in the aortic arch shows delayed filling of the subclavian artery via the vertebral and various collateral arteries. Doppler echocardiography may be able to demonstrate the reversal of flow in the vertebral artery, which would corroborate this diagnosis, but phase-encoded velocity mapping on MRI can also be definitive. Surgical management in children consists of repair of the accompanying heart disease and ligation of the ductus, if patent, to prevent pulmonary steal. Patients with central nervous system symptoms or claudication of the left arm may require surgical reimplantation of the subclavian artery into the carotid or aorta.

Dr. Hsiao-Huang Chang et al reported a case of right-sided aortic arch with Kommerell’s diverticulum of the aberrant left subclavian artery presenting with dysphagia, syncope, and left subclavian steal syndrome. Fewer than 50 cases have been reported in the literature. The repair was accomplished by division of a left ligamentum arteriosum, obliteration of the Kommerell’s aneurysm, and an aorto-subclavian bypass.
Hoarseness and left ptosis recovered spontaneously 3 months after surgery, and the patient remained symptom-free at the 1-year follow-up. He believes that a left thoracotomy for direct repair of Kommerell’s diverticulum is a simple and safe method without the increased morbidity found in other procedures.

References:
1. Moss and Adams, Heart Disease in Infants, Children, and Adolescents 7th edition, 2008; Aortic Arch Anomalies, Pg730-60


