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A rare cause of dysphagia – Aberrant Right Subclavian Artery (ARSA)

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Abstract. – Aberrant Right Subclavian Artery (ARSA) is a rare vascular anomaly. It is though the most frequent anomaly involving the aortic arch, being present in about 1% of the general population.

We present a case of a 71 year old male with symptoms of chronic cough and dysphagia. He was referred to our clinic for further evaluation of a mediastinal mass. A second Computed Tomography of the thorax was performed, this time with the use of i.v. contrast. The latter examination confirmed the diagnosis (ARSA), sparing the patient of invasive and potentially life threatening diagnostic procedures (invasive angiography, mediastinoscopy).

This case highlights the significant pitfalls regarding the mediastinal evaluation, especially when i.v. contrast is not used. The majority of the mediastinal structures are vessels. Vessels abnormalities should be included in the differential diagnosis of a mediastinal mass.

Key Words:

Aberrant Right Subclavian Artery (ARSA), Computed Tomography (CT), Edwards hypothetical double aortic arch.

Case Presentation

A 71 year old man presented to our Clinic for further evaluation of a mediastinal mass. The patient complained for dysphagia and chronic cough. This symptom urged his primary care physician to obtain a chest X-ray and subsequently a Computed Tomography (CT) of the thorax. Initially CT was performed without the use of i.v. contrast.

The patient was a former smoker for 20 years with a total of 20 pack/year. He stopped smoking at the age of 50. Auscultation of the lungs was within normal limits. Spirometry was normal.

He had arterial hypertension and was treated with hydrochlorothiazide and losartan.

At admission blood pressure was 135/70 mmHg.

Physical examination did not provide any significant abnormalities.

Total blood count and biochemical profile were within normal limits.

Computed Tomography (CT)

At admission the presence of a "soft tissue mass" was confirmed on Computed Tomography scans (Figures 1 to 5).

This "mass":

- had distinct margins;
- there were no signs of trachea infiltration (the inner contour of the trachea was smooth);
- had a "bizarre" elongated shape, as it is shown in sequentia l sections;
- most importantly there was a direct anatomic relationship with the descending aorta.

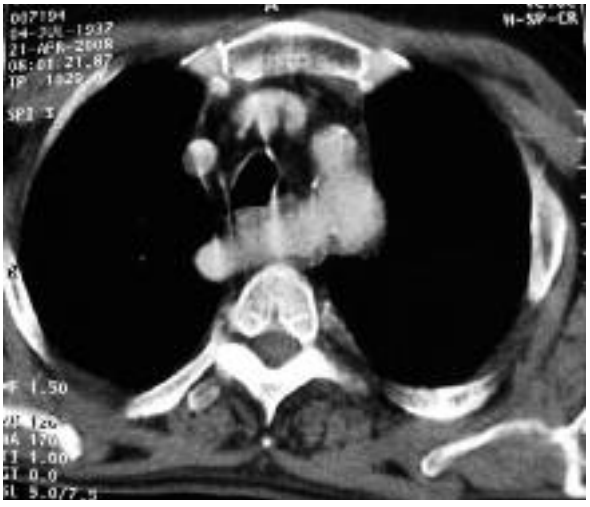

Figure 1. Aberrant Right Subclavian Artery (ARSA) is originating from the medial side of the aortic arch. Without the use of i.v. contrast the vascular origin cannot be confirmed.

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Figure 2. Aberrant Right Subclavian Artery (ARSA) is originating from the medial side of the aortic arch. Without the use of i.v. contrast the vascular origin cannot be confirmed.

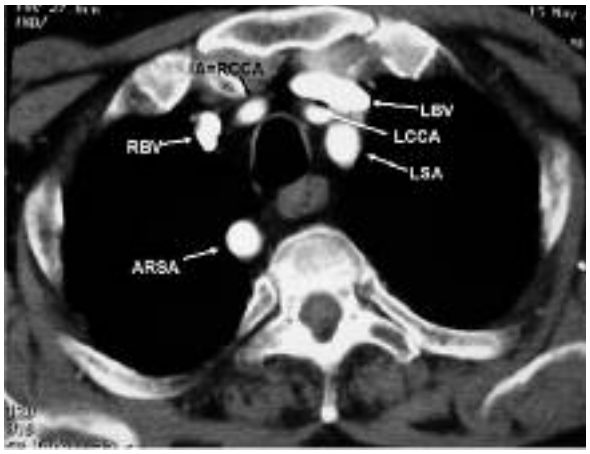

Figure 4. ARSA = Aberrant Right Subclavian Artery; LSA = Left Subclavian Artery; LCCA = Left Common Carotid Artery; RCCA = Right Common Carotid Artery IA = Innominate Artery; LBV = Left Brachiocephalic Vein; RBV = Right Brachiocephalic Vein.

Aneurysm of the aortic arch was also present.

The above CT findings were highly suggestive of a vascular aberrancy and specifically of Aberrant Right Subclavian Artery (ARSA). In order to confirm the diagnosis a second CT of the thorax was performed, this time with the use of i.v. contrast. The latter test confirmed the vascular origin of the lesion and established the final diagnosis. This way the patient avoided invasive diagnostic procedures (invasive angiography, mediastinoscopy) and further psychological stress.

The patient refused to undergo barium esophagogram. At 1 month follow up-visit he reported improvement following the appropriate dietary changes that were instructed.

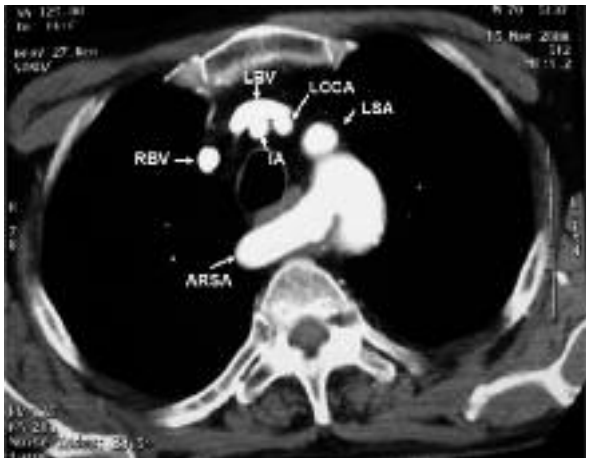

Figure 3. ARSA = Aberrant Right Subclavian Artery; LSA = Left Subclavian Artery; LCCA = Left Common Carotid Artery; RCCA = Right Common Carotid Artery IA = Innominate Artery; LBV = Left Brachiocephalic Vein; RBV = Right Brachiocephalic Vein.

Discussion

To be able to identify aberrancies of the aorta and great vessels the fundamentals of embryology are needed. The double arch system, described by Edwards, although hypothetical is of great practical help[1]. According to this, there is an aortic arch and a potential *ductus arteriosus* on each side. The descending aorta is in the midline posteriorly. Interruption of this arch at different locations can explain the various aortic arch anomalies. These can be divided into three main groups: left aortic arch anomalies, right aortic arch anomalies, and double aortic arch anomalies. Normally, the hypothetical right arch is interrupted distally to the right subclavian artery (Figure 5). The right common carotid and subclavian arteries fuse to become the Innominate Artery (IA). The proximal portion of the embryologic right arch becomes incorporated into the left arch. The result is the normal left-sided aortic arch.

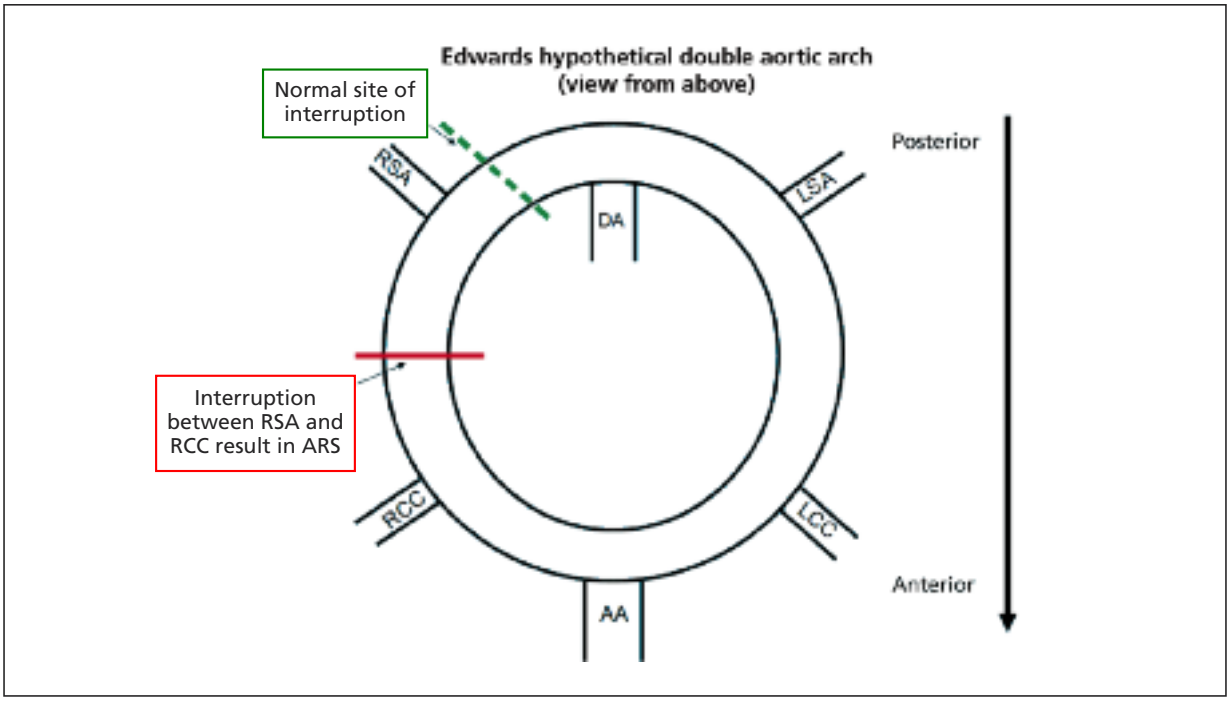

Figure 5. Interruption between RSA and RCC (indicated by the first left arrow) makes impossible the fusion of these arteries to form the innominate artery. So, while the aortic arch remains in the normal left position, the right subclavian artery originates as the fourth and last branch of the aortic arch (aberrant right subclavian artery). ARSA = Aberrant Right Subclavian Artery; LSA = Left Subclavian Artery; RSA = Right Subclavian Artery; LCC = Left Common Carotid Artery; RCC = Right Common Carotid Artery; DA = Descending thoracic aorta; AA = Ascending thoracic aorta.

Aberrant Right Subclavian Artery (ARSA) is the most common congenital aortic arch anomaly. It is present in approximately 1% of the general population[2-4]. According to Edward's model, this occurs when there is interruption of the embryologic right aortic arch between the right common carotid and the right subclavian arteries. The ARSA originates from the posterior portion of the otherwise normal left-sided arch. It does not originate from the Innominate Artery (IA) but is in fact a discrete fourth (and last) branch of the aortic arch after the origin of the Left Subclavian Artery (LSA). It crosses the mediastinum obliquely from left to right, lying posterior to the trachea (95%). In 80% of cases it lies posterior to both the trachea and esophagus, while in 15% of cases it lies between the trachea and esophagus. Rarely (5%) it crosses the mediastinum anterior to the trachea. Usually it is asymptomatic. When there is dilation of the aberrant vessel at its origin, this is known as Kommerell's diverticulum. In the last case the possibility of dysphagia is increased. The term "*dysphagia lusoria*" is used when there is a symptomatic, extrinsic compression of the esophagus, due to aberrancy in the aortic arch[5] (Table I). It was first described by Bayford in 1794[6].

Before the era of computed tomography, ARSA was a cause of confusion with other diseases such as mediastinal masses and could even lead to un-

Table I. Learning points.

- Aberrant Right Subclavian Artery (ARSA) is a relatively common vascular aberrancy (1%). It is the most common vascular anomaly involving the aortic arch
- Always think of a vascular anomaly when there is "extra tissue" in the mediastinum with anatomic correlation to vessels
- ARSA is usually asymptomatic but can cause dysphagia (*dysphagia lusoria*) due to extrinsic compression of the esophagus
- Use of i.v. contrast is of utmost importance for the radiological evaluation of the mediastinum

necessary surgery[7-8]. A detailed knowledge of the mediastinal structures is needed to avoid diagnostic pitfalls which could lead to unnecessary and potentially dangerous diagnostic procedures such as invasive angiography, mediastinoscopy or even surgery.

The use of i.v. contrast is of utmost importance in the evaluation of the mediastinum. In the case of ARSA it produces a virtually pathognomonic picture, as in our example.

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