First MDCT evidence of ruptured aberrant left subclavian artery aneurysm in right aortic arch, Kommerell's diverticulum and extrapleural hematoma treated by emergency thoracic endovascular aortic repair

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Abstract

Spontaneous ruptured aneurysm involving an aberrant subclavian artery with a right-sided aortic arch and Kommerell's diverticulum is a rare life-threatening condition that can be treated successfully if promptly identified. Multidetector Computed Tomography angiography is the first line imaging modality of thoracic vascular anomalies diagnosis. We report the case of a 74-year-old man suffering from this emergency condition with medi-astinal hematoma mostly extending to the left-side extrapleural cavity. The patient underwent successful emergency thoracic endovascular aortic repair and an Amplatzer vascular plug was placed into the first segment of the ALSA. Post-procedural imaging showed complete exclusion of the aneurysm. Emergency endovascular repair can be effective in such cases.

Introduction

Right aortic arch (RAA) is a rare anatomical variant occurring in 0.05 to 0.1% of radiology series [1,2]and in 0.04%-0.1% of autopsy series [3]. About 50% of people with RAA also have an aberrant left subclavian artery (ALSA) originating from a Kommerell's diverticulum (KD) [4,5]; it is usually asymptomatic and incidentally detected. To the best of our knowledge, we describe the first thoracic endovascular aortic repair (TEVAR) in a patient with spontaneous ruptured ALSA aneurysm in RAA and KD, complicated by a large extrapleural hematoma (EPH).

Case Report

A 74-year-old man presented at emergency department with 6 hours of abrupt onset of acute chest pain, anemia, hypotension (70/40 mmHg), and shortness of breath; the left hemithorax was dullness to percussion. Pulse was 160 beats/min, respiratory rate 36/min; at blood tests Ht was 30.4% and Hb 6.6 g/dL. Cardiac enzymes and ECG findings were normal. The patient's condition rapidly deteriorated up to require intubation. After resuscitation procedures, an emergency multidetector computed tomography angiography (MDCTA) revealed a RAA (Figure 1) and a saccular ruptured aneurysm (7 cm maximum diameter) coming immediately off the origin of a retroesophageal ALSA arising from a KD (Figure 2). In the upper mediastinum there was an anomalous course of the left brachiocephalic vein passing posteriorly to the ascending aorta (AAo) and forming the superior vena cava with the right brachiocephalic vein (Figure 3). This complex anatomy was associated to a hematoma around the descending aorta (DAo) separating the parietal pleura from the endothoracic fascia, mostly extending to the left-side extrapleural space, forming a large (10 x 13 x 32 cm) EPH with typical "extrapleural fat" and "fat ghost ribs" signs, related to the inward displacement of the extrapleural fat stripe medially to the ribs by the EPH (Figure 4) [6,7]. Through the right femoral artery two endoprosthesis were deployed in a tel-

escope modality with the proximal landing zone (LZ) just distal to the right subclavian artery and distal LZ in the proximal DAo (Ishimaru's Zone 4 or nobody's land), completely covering the aneurysm and the origin of the ALSA [8]. The subsequent angiography revealed no leak and initial sac thrombosis, confirmed by Transesophageal Echocardiography. To exclude retrograde patency of the aneurysm, an Amplatzer vascular plug was placed into the first segment of the ALSA (Figure 5). Twenty hours later a successful left carotid-axillary surgical bypass (CAB) was performed [9,10]. After 15 days the patient was discharged without complications except for a brief transient tetraplegia completely recovered and is alive and healthy at 2-years follow-up.

Discussion

In the aortic arch (AA) anatomical variants the first diagnostic step is to evaluate the laterality of the arch, defined as the side of the trachea on which the arch crosses, and the main bronchi. Another rule is that the first arch branch vessel that contains a common carotid artery is contralateral to the AA, or that the retroesophageal or ALSA is always contralateral to the arch. RAA results from the persistence of the right fourth arch and right dorsal aorta and involution of the left fourth arch and dorsal aorta [1-3,11]. In an autopsy study, 50% of cases of RAA were associated with an ALSA, which can be retroesophageal (in 80%), located between the trachea and the esophagus (in 15%), or anterior to the trachea (in 5%) [3]. Two anatomical RAA variants clinically differing in presentation, associations, and prognosis are described [11,12]:

- Type 1 with "mirror-image branching", in which a left innominate artery arises as the first branch of the AA, and then divides into a left common carotid and left subclavian artery; in approximately 75% of these patients, cyanotic congenital anomalies (including tetralogy of Fallot, pulmonary stenosis with ventricular septal defects, tricuspid atresia, and truncus arteriosus) are present;
- Type 2 with ALSA, more common, which arises as a fourth branch of the RAA or from an aortic diverticulum (KD); congenital heart anomalies are present in only 5% to 10%.

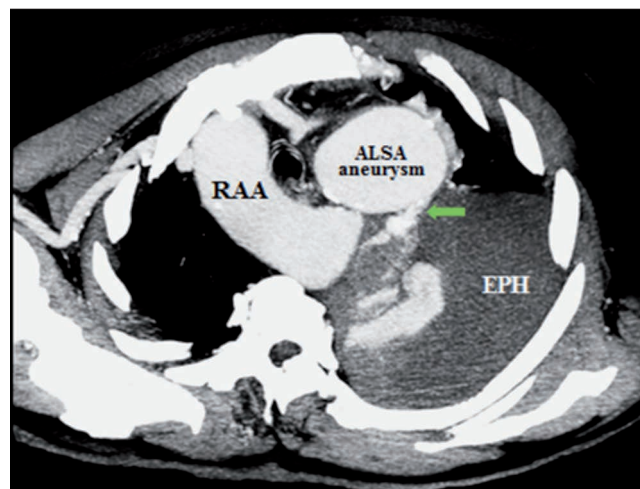In 10% of cases with a RAA and ALSA, there is a right ductus,

Figure 1. Maximum-intensity projection oblique axial reformat image shows a right aortic arch (RAA) and a ruptured (arrow) aneurysm of a retroesophageal aberrant left subclavian artery (ALSA). Discontinuity of ALSA aneurysm wall (green arrow) with extraluminal extravasation of contrast material and large haematoma in the posterior mediastinum, pleural and extrapleural spaces (EPH).
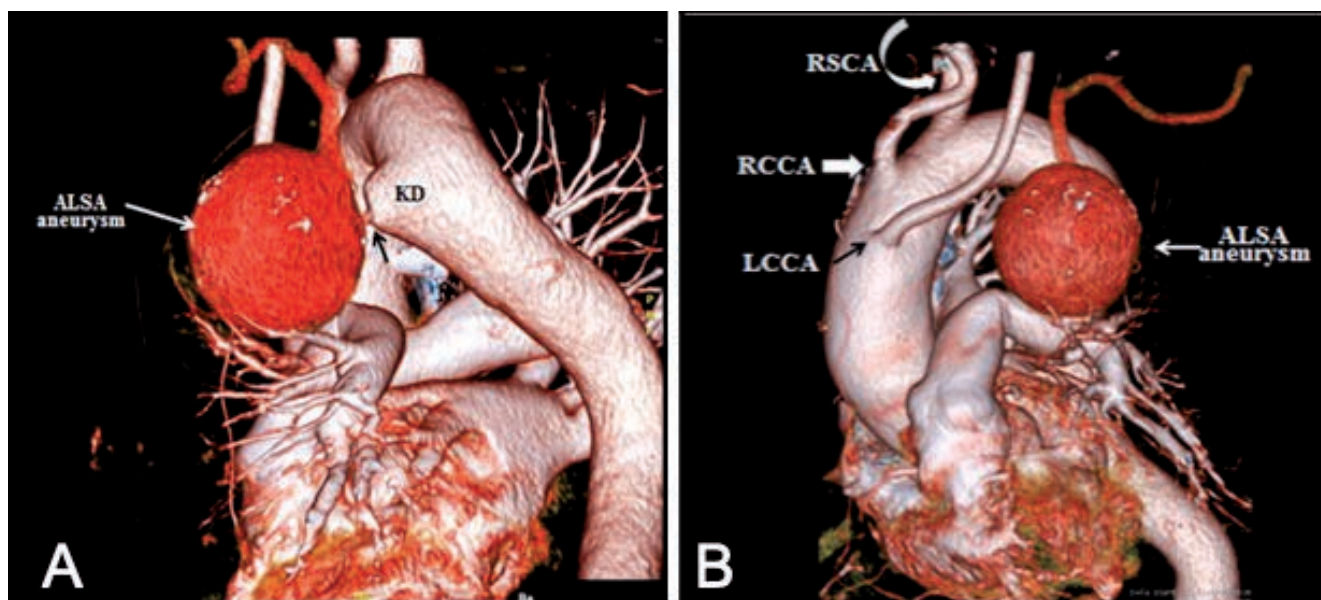

Figure 2. 3D volume rendering oblique sagittal reformat image. In (A) it is clearly appreciable how the aneurysm comes immediately off the aberrant left subclavian artery (ALSA) origin (arrow) from a Kommerell's diverticulum (KD); in (B) the right aortic arch branching pattern is the left common carotid (LCCA), right common carotid (RCCA) and right subclavian (RSCA) arteries followed by an ALSA aneurysm, which originates as the last branch from the proximal descending aorta and has a retroesophageal course to reach the left subclavian region.
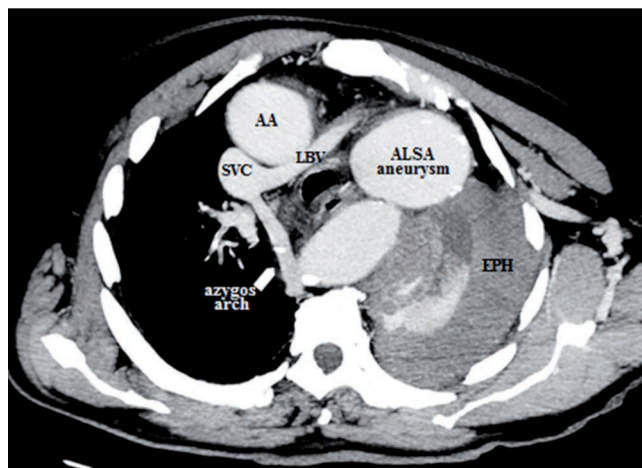
Figure 3. Maximum-intensity projection reformat axial image shows the anomalous course of the left brachiocefalic vein (LBV), posterior to the ascending aorta (AA) to form the superior vena cava (SVC). Hyperacute hypodense hematoma is associated to hyperdense extraluminal extravasation of contrast material.
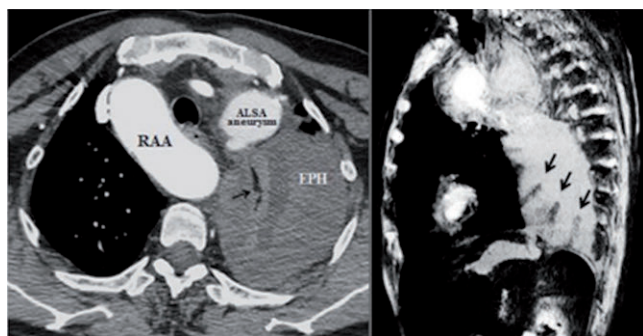

Figure 4. A) Maximum-intensity projection axial reformat image shows inward displacement of the extrapleural fat stripe (arrow) by extrapleural hematoma (EPH). B) Oblique sagittal minimum-intensity projection reformat image shows "fat ghost ribs" sign (arrows) related to EPH; it is caused by a greater presence of displaced fat in the intercostal spaces compared to that present inside of the ribs.

Figure 5. After thoracic endovascular repair (TEVAR) 3D volume rendering oblique sagittal reformat image shows complete aneurysmal exclusion and plug (arrow) in ALSA.

without ring or associated intracardiac defect, but in 90% of the cases there is a left ductus, that is the second most common cause of a symptomatic vascular ring. In the adult population a RAA is often asymptomatic unless aneurysmal disease develops [13].

A meticulous analysis of vascular structure, obtained with a rigorous multiphasic MDCTA, is useful to define multiple combined vascular anomalies, their relationships with the trachea and esophagus, and it is essential for facilitate endovascular repair or surgical planning [14]. In our case the branches originate from the RAA in the following order: left and right common carotid arteries, right subclavian artery (RSCA) and ALSA which originates as the last branch from the proximal DAo (Type 2) and has a retroesophageal course to reach the left subclavian region [1-3,12]. The ALSA originates in the proximal DAo directly or more commonly from a bulbous diverticulum (KD), that represents the remnant of the left dorsal aortic root in the RAA and may to cause tracheal compression or dysphagia [15-17].

In adults, clinical implications of a RAA with ALSA and KD include esophageal compression, stridor, wheezing, dysphagia and, in the presence of atherosclerotic changes, complications such as dissection or aneurysmal dilatation up to vessels rupture [18]. Aberrant subclavian arteries are more prone to dilate than non-aberrant vessels and in the rupture of an aneurysmal aberrant subclavian artery a 100% mortality rate has been described [19]. One of the elective therapeutic options is its exclusion by TEVAR, preceded by a carotid-subclavian (or axillary) debranching bypass. In case of ruptured ALSA/diverticulum, the first-line treatment consists in TEVAR, followed by surgical bypass as soon as possible. The aim is to reduce the risk of spinal ischemia in cases of vertebral artery origin from the ALSA and to reduce the risk of upper left limb ischemia in cases of vertebral artery origin directly from the arch.

Conclusions

A correct contrast enhanced multiphasic MDCT is essential for the study of vascular anomalies allowing to obtain an adequate surgical planning of their complications even with less invasive and less time-consuming procedures such as TEVAR.

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