

Thrombosis of Kommerell's diverticulum with subclavian steal phenomenon in a patient with non-small cell lung carcinoma under chemotherapy

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ABSTRACT

Kommerell's diverticulum (KD) is defined as a bulbous dilatation of the origin of an aberrant subclavian artery due to a remnant of the left fourth aortic arch. We report the case of an asymptomatic woman in whom progressive thrombosis of the KD extending to the prevertebral tract of an aberrant right subclavian artery was detected at multidetector computed tomography imaging for lung cancer staging performed before and after the beginning of chemotherapy. Reversed blood flow in the ipsilateral vertebral artery due to subclavian steal phenomenon was also observed by color Doppler ultrasound examination.

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1. Introduction

Kommerell's diverticulum (KD) is a rare condition consisting of both an aneurysm of the thoracic aorta and an aneurysmal orifice of an aberrant subclavian artery. It corresponds to a conical dilatation at the origin of the aberrant vessel and is therefore also known as “lusoria diverticulum”, “remnant diverticulum” or “lusoria root” [1–3].

Originally, Burckhard F. Kommerell reported an aortic diverticulum in a patient having a left aortic arch with an aberrant right subclavian artery in 1936 [4]. Basically, there are three types of aortic arch diverticulum:

- the diverticulum at the left aortic arch with aberrant right subclavian artery (more frequently: 0.5%–2.0% of the population);
- the diverticulum at the right aortic arch with anomalous origin of the left subclavian artery (0.05%–0.1%);
- the aortic diverticulum at the aortic-ductal junction [1–3].

Our purpose is to show the case of an asymptomatic patient with complete thrombosis of the prevertebral portion of an aberrant right subclavian artery, progressive KD thrombosis, and reverse ipsilateral vertebral artery flow that was detected by multidetector computed tomography (MDCT) imaging performed for lung cancer staging before and after the beginning of chemotherapy, and subsequently confirmed by a color Doppler ultrasound examination.

2. Case presentation

A 70-year-old Caucasian woman with biopsy proven non-small cell lung carcinoma (NSCLC) underwent a contrast-enhanced whole body MDCT examination for cancer staging. MDCT findings showed a left aortic arch with a KD, an autonomous origin of the left vertebral artery from the aortic arch, and origin of both common carotid arteries from a short common trunk. An aberrant (lusory) right subclavian artery stemmed from the KD and was completely thrombosed at its origin as well (Fig. 1). The patient received chemotherapy, and a follow-up MDCT examination performed two months later showed complete thrombosis of the KD extended to the prevertebral portion of the right subclavian artery. The right vertebral artery was patent as well as the post-vertebral portion of the ipsilateral subclavian artery, suggesting retrograde vertebral flow due to subclavian steal phenomenon (Fig. 2).

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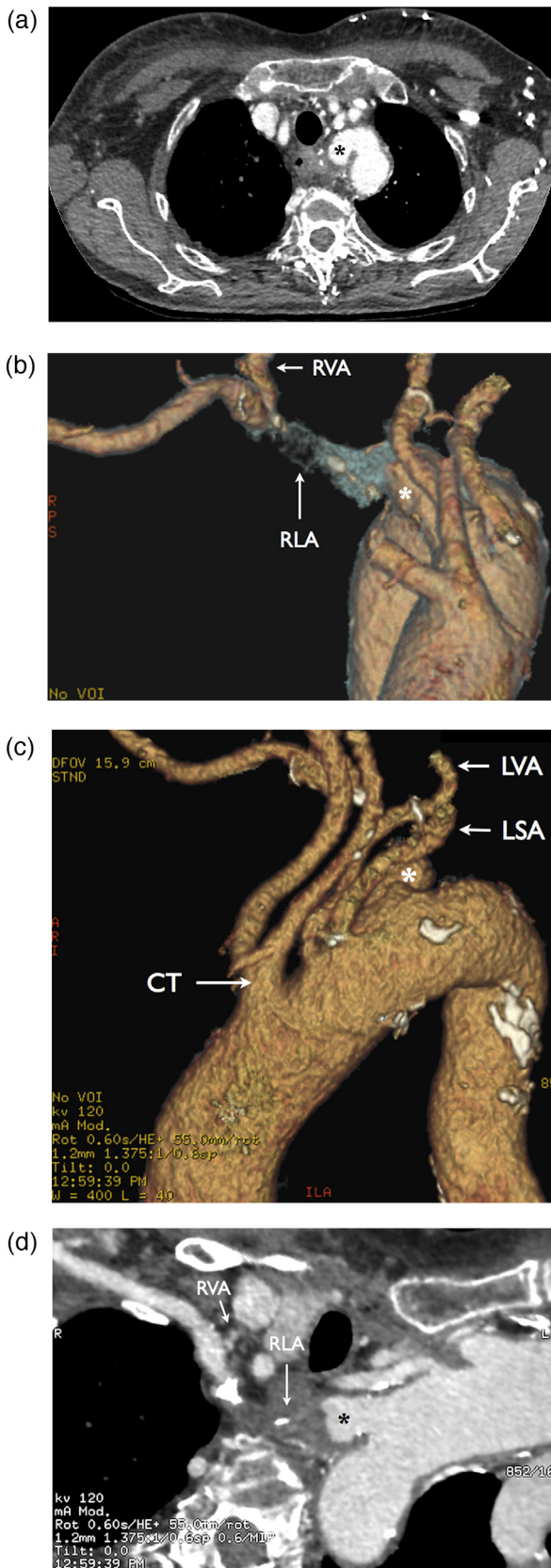


Fig. 1. Baseline MDCT examination. (a) Axial source image, (b, c) Volume Rendering and (d) Curved Planar views show patent KD (*) associated with thrombosed prevertebral right lusory artery (RLA), autonomous origin of the left vertebral artery (LVA) from the aortic arch, and origin of both common carotid arteries from a short common trunk (CT). The proximal portion of the right vertebral artery (RVA) is patent, as well as the RLA distal to it. LSA = left subclavian artery.

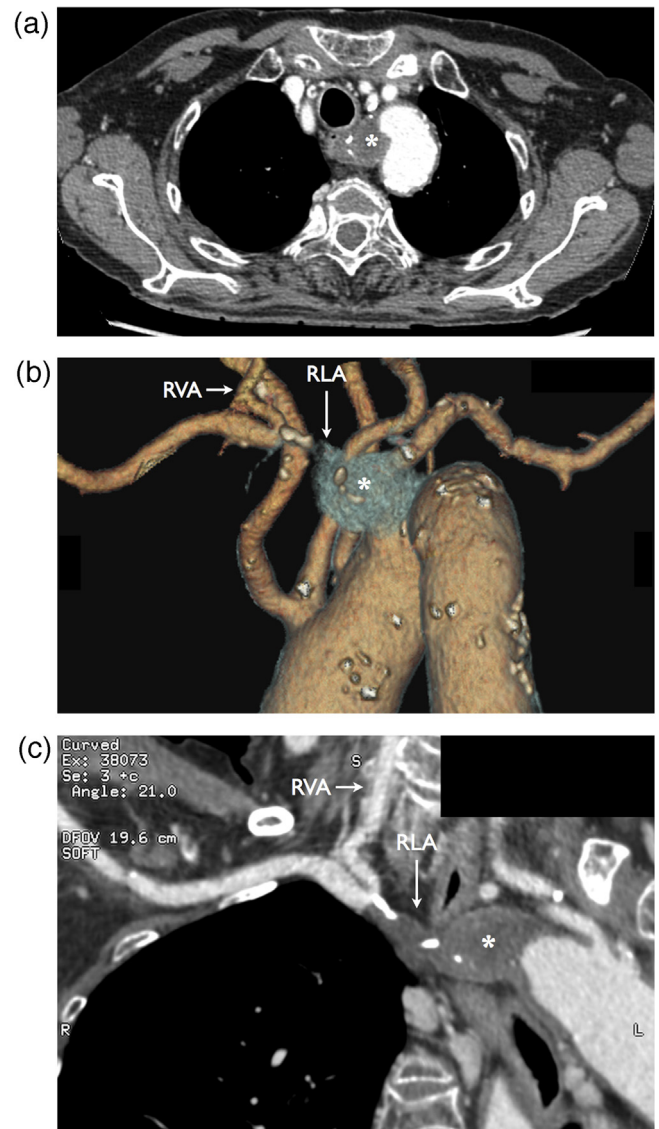


Fig. 2. Two-month follow-up MDCT examination. (a) Axial source image, (b) Volume Rendering and (c) Curved Planar views show complete thrombosis of the KD extending from the prevertebral RLA. As at baseline, the proximal portion of the RVA and the RLA distal to it are patent.

A Doppler ultrasound examination was performed some days later for hemodynamic assessment, confirming reverse flow in the right vertebral artery compared with the left side (Fig. 3). The patient had never complained of any symptoms related to subclavian steal syndrome or tracheo-esophageal compression, and laboratory tests did not reveal any pre-existing hematologic disorders potentially favoring a pro-thrombotic condition.

3. Discussion

The embryological development of the aorta begins during the second week of gestation and is completed by the seventh week. Between the fourth and fifth weeks of embryonic life, blood leaves the heart by a single vessel (i.e. the truncus arteriosus), which divides into two branches named the ventral and dorsal aortae, respectively. The ventral aortae are connected with the dorsal aortae by six branchial vessels, called aortic arches. These latter are numbered from cephalad to caudal and normally develop into the thoracic aorta and its branches.

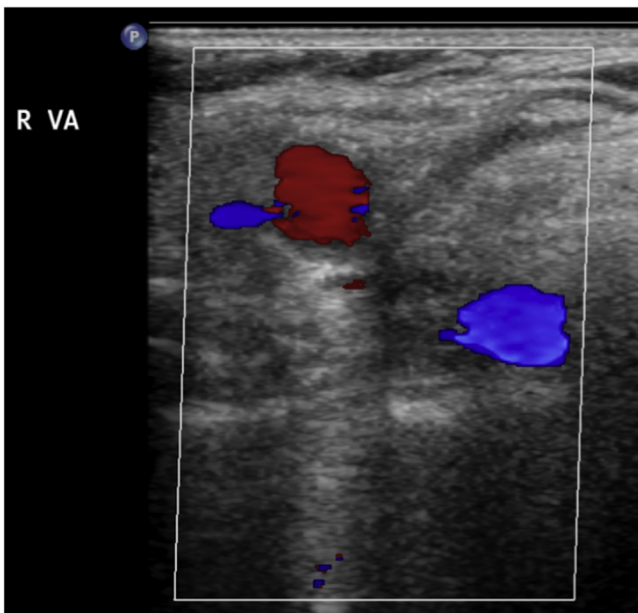


Fig. 3. Doppler ultrasound examination obtained with a transoccipital approach shows reversed flow in the RVA compared with the LVA.

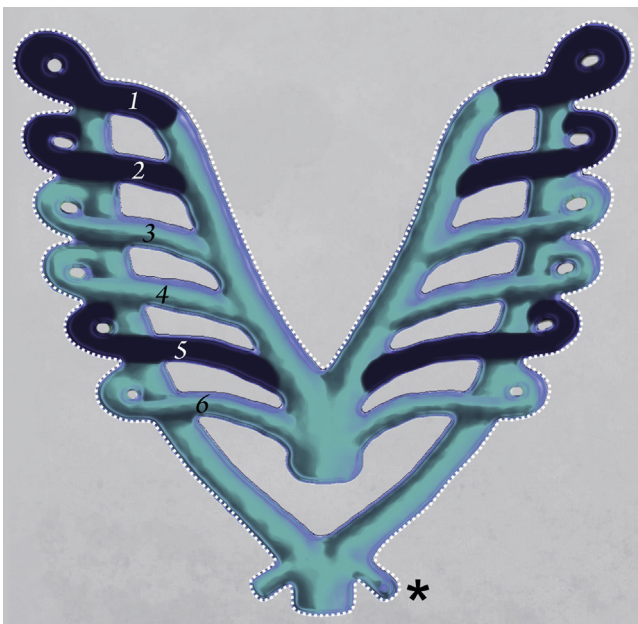


Fig. 4. Embryological development of the aorta and the supra-aortic vessels. (*) intersegmental artery.

In particular, the first and the second pair contribute to the formation of the maxillary and external carotid arteries and of the stapelial arteries, respectively. The proximal segments of the third pair form the common carotid arteries. Together with segments from the dorsal aortae, the distal portions contribute to the development of the internal carotid arteries. The left arch of the fourth pair forms the segment of normal left aortic arch between the left common carotid and subclavian arteries. The right fourth arch forms the proximal right subclavian artery, whereas the distal right subclavian artery is derived from a portion of the right seventh intersegmental artery. The fifth arches are both reabsorbed, and the sixth arches form the pulmonary artery and the ductus arteriosus (Fig. 4).

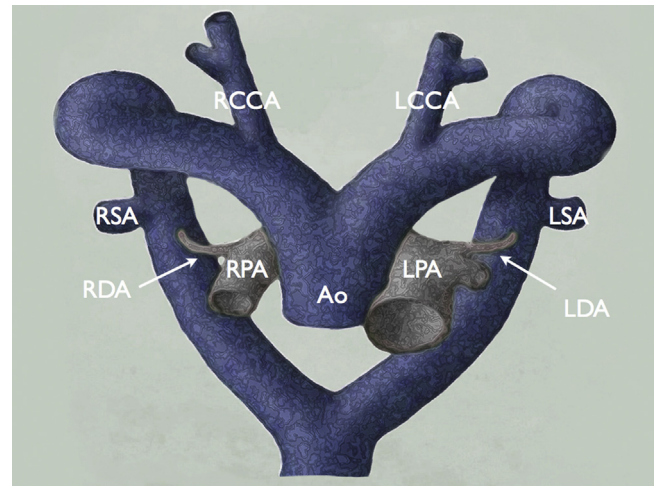


Fig. 5. Illustration of Edwards' hypothesis. Ao = aorta, RCCA = right common carotid artery, LCCA = left common carotid artery, RSA = right subclavian artery, RPA = right pulmonary artery, LPA = left pulmonary artery, RDA = right ductus arteriosus, LDA = left ductus arteriosus.

If the left fourth arch disappears and the right one persists, a right aortic arch develops, whereas if both arches persist, they form a double arch or a vascular ring encircling the trachea and oesophagus [5,6].

Edwards' hypothesis is useful to understand the various aortic arch anomalies [3,5–7]. According to this theory, the double aortic arch with the ductus arteriosus is located on each side (thus surrounding the trachea and the oesophagus) and the descending aorta lies in the midline. The common carotid and subclavian arteries on each side stem from the ipsilateral aortic arch. The normal aortic arch is formed by interruption of the dorsal segment of the right aortic arch distally from the right subclavian artery to the descending aorta and regression of the right ductus arteriosus (Fig. 5).

Based on the different sites of interruption of the double aortic arch, aortic arch anomalies can be classified into the following three types:

- type I: the left arch is interrupted between the descending aorta and left subclavian artery. This arch branching pattern is the mirror image of the conventional branching pattern of the normal left aortic arch;
- type II: the interruption occurs between the left common carotid and subclavian artery;
- type III: the interruption takes place proximal and distal to the left subclavian artery, which is therefore isolated and may be connected either to the pulmonary artery by the ductus arteriosus or to the left vertebral artery [3,5–8].

Among the aforementioned conditions, type II right aortic arch with an aberrant left subclavian artery is the most common one. In this condition, an aberrant left subclavian artery arises either as the last branch of the right-sided aortic arch or from an aortic diverticulum, such as the KD, which results from reabsorption of the left fourth aortic arch proximal to the origin of the left subclavian artery.

Most patients with KD are asymptomatic unless aneurysmal disease develops. KD dilatation may increase the risk of aneurysm rupture, and recent histopathological studies have revealed a high prevalence of cystic medial necrosis in the KD wall, potentially accounting for the increased risk of KD dissection and rupture [3,9]. Moreover, KD aneurysm may result in compression of the surrounding mediastinal structures (especially the trachea and the

esophagus), causing symptoms such as dysphagia, dyspnea, stridor, wheezing, cough, recurrent pneumonia, obstructive emphysema, or chest pain [1–3].

The most severe issues in the course of aneurysms are their higher propensity to acute rupture with increasing aneurysm size, and especially in the case of aneurysms of the supra-aortic vessels, the risk of hemodynamic impairment with distal hypoperfusion or embolization due to thrombosis of the aneurysm lumen. Clinical and imaging diagnoses focus on the reduction of morbidity and mortality associated with such risks, yet aneurysmal disease may often go undetected due to lack of symptoms unless imaging procedures spanning the upper chest are performed for diagnosis or follow-up of concurrent disease [1–4].

Our patient showed a KD associated with a type II left sided aortic arch variant, and to our knowledge, this is the first report to date of a thrombosed prevertebral right lusory artery associated with a progressively thrombosed KD, detected by both MDCT and color Doppler ultrasound in a patient with no history of ischemic symptoms or swallowing disorders. Unfortunately, the patient had not received any prior imaging tests that might have been useful to date the onset of her right prevertebral lusory artery thrombosis. Vlummens et al. reported a similar case of aberrant right subclavian artery originating from a thrombosed KD in an 87-year-old male patient with prostate cancer and history of swallowing difficulties and stagnation of saliva, in whom secondary subclavian steal syndrome was suspected based on the anatomical finding of opacification of the ipsilateral vertebral artery on MDCT images, but not confirmed functionally by a Doppler ultrasound examination as in our own case [10]. In analogy with this case, the hematologic workup revealed no pre-existing coagulation disorders, and the fact that progressive thrombosis of the KD occurred between the baseline and post-chemotherapy MDCT examinations may raise the suspicion that a pro-thrombotic state may have occurred either as a paraneoplastic syndrome due to the underlying NSCLC or as a side effect of chemotherapy.

Surgical indications for the KD have not been established systematically because of its rareness. Nonetheless, early surgical treatment of the KD is recommended because of the risk of aneurysm rupture, and usually entails resection of the diverticulum and reimplantation of the aberrant subclavian artery into the aorta. However, the type of surgical procedure depends on the patient's age and relationship between the KD and the surrounding tissues, and especially in elderly or other frail patients (in whom open surgery is associated with a higher mortality and the risk of neurological complications), hybrid techniques tend to be preferred, such as endovascular occlusion followed by open revascularization [2,10,11].

In the specific case of our patient, anticoagulation therapy was started and no vascular treatments have been performed so far, due to the lack of KD-related symptoms and to the underlying

lung cancer with brain metastasis (as shown by staging MDCT examinations). From a diagnostic standpoint, the excellent spatial and contrast resolution and the panoramacity afforded by modern MDCT technology allow an accurate evaluation of the aneurysm size and intraluminal thrombosis, patency and course of the thoracic aorta and subclavian vessels, presence of collateral pathways via the vertebral circulation, and relationship with the neighboring nonvascular structures, thus providing the key elements for diagnosis and potential treatment planning [3,12].

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Conflict of interest

None.

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