Anomalous Origin of Bilateral Vertebral Arteries Associated with an Aberrant Right Subclavian Artery and a Common Carotid Trunk: Case Report

Aberran Sağ Subklavyan Arter ve Ortak Karotid Trunkus ile Birliktelik Gösteren Bilateral Anormal Çıkışlı Vertebral Arter

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ABSTRACT Variations in the origin and the course of the vertebral arteries are uncommon and regarded as a congenital anomaly occurring in the early stages of embryonic development. These variations of the vertebral arteries occur mostly on one side and usually on the left. The most frequent variant is a left vertebral artery arising directly from the aortic arch between the left common carotid and the left subclavian arteries. However, an anomalous origin of the the right vertebral artery is much rarer and detected as an incidental finding at surgery or autopsy. Anomalous origins of both vertebral arteries are exceedingly rare. There are only a few case reports in the radiology literature that discuss the conventional angiographic findings of this rare anomaly. Knowledge of the variations in the aortic arch branching pattern is important prior to endovascular interventional procedures or vascular surgery in the neck region. In this report, we present multidetector computed tomographic angiography findings of an extremely rare combination of anomalies of the aortic arch consisting of an aberrant right subclavian artery; a common origin of both common carotid arteries with a right vertebral artery originating from the right common carotid artery and a left vertebral artery originating from the aortic arch between the common carotid trunk and left subclavian artery.

Key Words: Vertebral artery; subclavian artery; congenital abnormalities; tomography, X-ray computed; angiography

ÖZET Vertebral arterlerin çıkış ve seyrindeki varyasyonlar yaygın değildir ve embriyonik gelişimin erken evrelerinde oluşan bir konjenital anomali olarak kabul edilirler. Vertebral arterlerin bu varyasyonları büyük çoğunlukla tek taraflı ve genellikle soldadır. En sık görülen varyant, sol ana karotid arter ile sol subklavyan arter arasında aortik arkdan direkt olarak köken alan sol vertebral arterdir. Ancak, anormal çıkışlı sağ vertebral arter daha nadirdir ve ameliyat veya otopsi sırasında rastlantısal bir bulgu olarak saptanır. Her iki vertebral arterin anormal çıkışlı olması son derece nadirdir. Radyoloji literatüründe bu nadir anomalinin konvansiyonel anjiyografik bulgularını tartışan yalnızca birkaç olgu sunumu vardır. Aortik arkın dallanma paternindeki varyasyonları bilmek, endovasküler girişimsel prosedürler ve boyun bölgesinin vasküler cerrahisi öncesinde önemlidir. Biz bu raporda, aberran sağ subklavyan arter, her iki ana karotid arterin ortak kökeni, sağ ana karotid arterden çıkan sağ vertebral arter ve aortik arkdan direkt olarak çıkan sol vertebral arterden oluşan aortik ark anomalilerinin son derece nadir bir kombinasyonunun çok kesitli bilgisayarlı tomografik anjiyografi bulgularını sunuyoruz.

Anahtar Kelimeler: Vertebral arter; subklavyan arter; konjenital anomaliler; bilgisayarlı tomografi; anjiyografi

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Usually, the vertebral artery (VA) is the first branch of the ipsilateral subclavian artery, originating from the posterosuperior surface of the ascending part of the subclavian artery and it passes backward to en-

ter the transverse foramina of sixth cervical vertebra. Variations in the origin and the course of the vertebral arteries are uncommon and regarded as a congenital anomaly occurring in the early stages of embryonic development.[1] Embryologically, vertebral arteries are formed by the fusion of the longitudinal anastomoses that link the cervical intersegmental arteries, which branch off the primitive dorsal aorta. The intersegmental arteries eventually regress, except for the seventh artery, which forms the proximal portion of the subclavian artery, including the point of origin of the vertebral artery. Failure of involution in one of the first six intersegmental arteries causes a variety of abnormal origins of the vertebral artery. If the first or second cervical intersegmental arteries persists, the result is an anomalous origin of the vertebral artery from the internal or external carotid artery. If the third through sixth cervical intersegmental arteries persists, the result is an anomalous origin of the VA from the aortic arch or the common carotid artery.[1,2] Variations of the vertebral artery frequently occur only on one side and usually on the left. The most frequent variant is a left vertebral arteries arising directly from the aortic arch between the left common carotid and left subclavian artery, with a reported prevalance of 2.4-5.8% in several large autopsy series.[3] However, an anomalous origin of the the right vertebral artery is rarer and detected as a coincidental finding during surgery or autopsy. The right vertebral artery originating from the right carotid artery is a rare variant, with a reported incidence of 0.18%.[4] Anomalous origins of both vertebral arteries are very rare. There are only a few case reports in the radiology literature that discuss the conventional angiographic findings of this rare anomaly.[5-7]

In this report, we describe multidetector computed tomographic (MDCT) angiography findings of an extremely rare combination of anomalies of the aortic arch consisting of an aberrant right subclavian artery; a common origin of both common carotid arteries with the right vertebral artery originating from the right common carotid artery and the left vertebral artery originating from the aortic arch between the common carotid trunk and left subclavian artery.

CASE REPORT

A 40-year-old woman with a past medical history of hypertension presented with central chest pain. She had been followed up by using echocardiography due to type 3 aortic dissection for 10 years. On physical examination, her blood pressure was 110/65 mmHg and her pulse was 76 beats/min. The electrocardiography showed no abnormalities. A chest radiograph showed mediastinal widening. With echocardiography, an aneurysm of the descending aorta with an intimal flap was observed. The patient was referred to our department for MDCT angiography of thorax as a further investigation. MDCT angiography was performed using a 16-row MDCT (Lightspeed Ultra, General Electrical Medical Systems, Milwaukee, Wisc.; USA). Imaging parameters were as follows: 120 kV, 16 x 1.25 mm collimation, speed 27.5 mm/rot, rotation time 0.5 sec, pitch value of 1.375:1. After determining the contrast agent transit time using the smart prep bolus technique, we acquired image data during an intravenous injection of 100 ml iodinated contrast agent (Iodixanol, Visipaque 320 mgI/ml, GE Healthcare, Milwaukee, Wisc.; USA) at a rate of 4 ml/sec. For three-dimensional image reconstruction, all acquired data were processed on a separate workstation (Advanced Workstation 4.2, GE Healthcare, Milwaukee, Wisc.;USA) with multiplanar reformatting, maximum intensity projection and volume rendering. MDCT angiography showed a descending aortic aneurysm, 5.2-cm in diameter with a dissection flap. There were also dilatation of the ascending and arcus aorta. Mural calcifications of the descending aorta were present. MDCT angiography revealed a left sided aortic arch with the branches from right to left as follows: a common trunk for the right and left common carotid arteries with the right vertebral artery arising from the right common carotid artery, the left vertebral artery, left subclavian artery and an aberrant right subclavian artery (Figure 1). The aberrant right subclavian artery crossed the mediastinum from left to right passing behind the trachea and oesophagus and causing mild compression of the latter. There was calcified plaque and short segment occ-

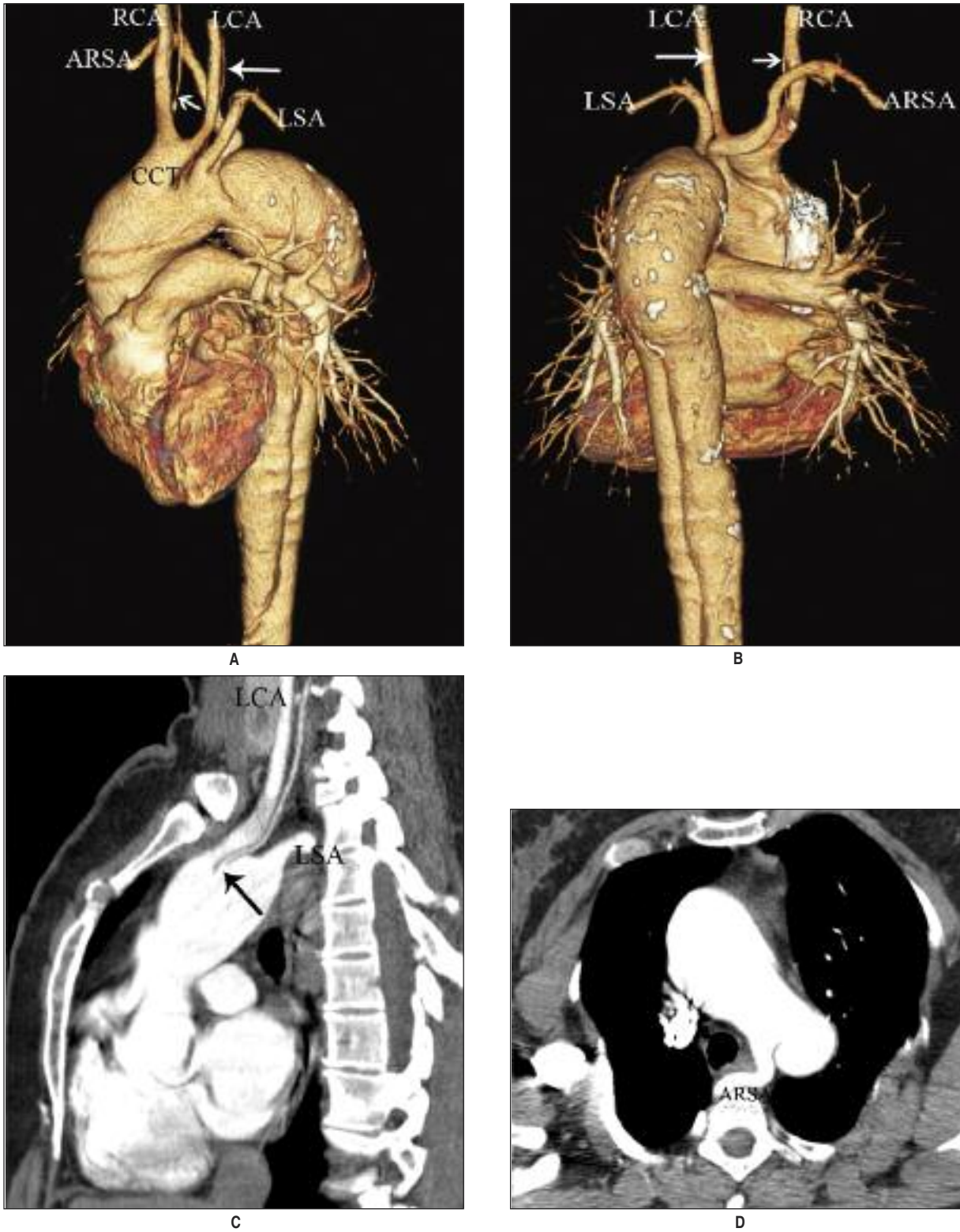

FIGURE 1: Anterior **(A)** and posterior **(B)** three dimensional volume rendering multidetector computed tomography images show a descending aortic aneurysm with a dissection flap. The supraaortic branches from right to left are as follows: a common trunk (CCT) for the right (RCA) and the left (LCA) common carotid arteries, with a right vertebral artery (short arrow) arising from the right common carotid artery, the left vertebral artery (long arrow), the left subclavian artery (LSA) and an aberrant right subclavian artery (ARSA). Sagittal multiplanar reformatted image **(C)** shows the left vertebral artery (long arrow) arising from the aortic arch. Axial image **(D)** shows the aberrant right subclavian artery crossing the mediastinum from left to right, passing behind the trachea and oesophagus and thus causing compression of the latter.

lusion in the proximal portion of the right vertebral artery. The MDCT findings of multiple supraaortic arterial anomalies were incidental and without significant clinical consequences, hence, no further evaluation was warranted. The patient is under follow-up in order to control progression of descending aortic aneurysm with dissection.

DISCUSSION

With the increased use of imaging studies, anomalies of the aortic arch and supraaortic branches have been identified more frequently. Various anatomic variants of the supraaortic arteries have been reported in the literature. Among all the variants, the most frequent anomaly is the common origin of the left carotid and innominate arteries, also known as a bovine arch, which accounts for 73% of all aortic arch vessel anomalies and is prevalent in approximately 22% of the population. An aberrant right subclavian artery as the most distal branch of the aortic arch is uncommon, representing approximately 1% of all arch vessel anomalies.[8,9] It has been reported that the aberrant right subclavian artery typically travels in a retrooesophageal course in 80% of patients, between the trachea and oesophagus in 10-15% of patients, and anterior to both structures in 5% of patients.[8] Although most cases of aberrant right subclavian artery are asymptomatic, only 10% of adults have symptoms of dysphagia because of extrinsic compression on the posterior aspect of the oesophagus.[8]

A vascular ring caused by an aberrant right subclavian artery associated with a common origin of the both carotid arteries is rare and its incidence may be less than 0.05%.[8,10] The association of an aberrant right subclavian artery and a common carotid trunk can be explained by the embryologic double aortic arch reported by Edwards, who described various aortic arch abnormalities by selective regression of various parts of each arch. An aberrant right subclavian artery occurs as a result of the interruption of the embryonic right aortic arch proximal to the seventh cervical intersegmental artery and both sides of common carotid arteries arise from the aortic sac without separation from each other to form a common origin of the carotid arteries.[10] Most cases with common carotid trunk have no symptoms except for a few reported cases of respiratory distress during early infancy. Respiratory distress may be ascribed to superior mediastinal crowding and tracheal compression by the common carotid trunk forming a 'V' shape in front of the trachea and running along both sides. The tracheal compression and respiratory distress may improve with age due to lessening of the degree of such crowding. Thereby, patients with an aberrant right subclavian artery associated with common carotid trunk may be asymptomatic or have dysphagia or respiratory symptoms.[8-11]

Anomalous origin of the vertebral artery is only present in 15.7% of cases with aberrant right subclavian artery.[11-13] The clinical significance of anomalous vertebral artery origins is not clear, yet in most cases described in the literature, anomalous vertebral artery origins did not result in clinical symptoms. There are very few reports describing patients who complained of symptoms of vertebrobasillary insufficiency. Kinking of vertebral artery and increased incidence of arterial dissection with anomalous origin of the vertebral artery was also reported. Theoretically, altered hemodynamics cause turbulence, which may predispose the patients to aneurysms, and therefore increase the risk of a cerebrovascular accident. However, there is no conclusive evidence that these variants lead to a predisposition to cerebrovascular disorders.[4,5]

Although supraaortic vascular anomalies are considered as anatomic variants, they are potentially important to be recognized prior to vascular surgery in the head and neck region, as well as during the aortic arch surgery. This is important in order to avoid accidental vascular injury and optimize access for vascular anastomosis during surgery. Detailed knowledge of anomalous origin of supraaortic branches is also of importance for patients who are to undergo four-vessel cerebral angiography in an emergency unit to rule out, for example, the possibility of intracranial aneurysm after subarachnoid hemorrhage. If the vertebral artery origin is unknown, the patient might be wrongly diagnosed as vertebral artery hypoplasia or aplasia. This leaves the probability of missing the

pathology, such as dissection or aneurysm formation in the missed vertebral artery. Additionally,, knowledge of the variations in the aortic arch branching pattern becomes more important in the era of carotid or vertebral artery stents and new therapeutic options for intercranial interventions.

Even though conventional angiography remains the gold standard method for imaging the supraaortic branches, MDCT angiography is playing increasingly important role in the evaluation of the thoracic vascular pathologies including supraaortic branches. The advantages of MDCT angiography in comparison with conventional angiography include ability for unrestricted reconstruction of images, more rapid image acquisition, and lack of potential angiographic complications. It is also able to display the detailed anatomy of the vascular structures, and with MDCT, images can be reconstructed in various planes and used as a roadmap for interventional and surgical treatment planning, especially in patients with complex aortic arch anatomy. The supraaortic branches can be well delineated with excellent image quality by MDCT.[11,13] The disadvantages of this method are the need for an iodinized contrast material and radiation exposure.

In conclusion, the understanding of the variations in the aortic arch branching pattern is important prior to endovascular interventional procedures or vascular surgery in the neck region. MDCT angiography is able to exhibit the detailed anatomy of the aorta and supraaortic arteries with excellent image quality.

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