Aberrant Left Subclavian Artery Associated with an Aneurysmal Diverticulum of Kommerell: Multidetector CT Angiography Findings

Anevrizmal Kommerell Divertikülünün Eşlik Ettiği Aberran Sol Subklavyen Arter: Multidedektör BT Anjiyografi Bulguları

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ABSTRACT Aberrant left subclavian artery originating from a right sided aortic arch is an uncommon congenital aortic arch anomaly. Kommerell's diverticulum is defined as dilatation of the proximal portion of the aberrant subclavian artery near the origin from the aorta, and it represents the embryonic remnant of the dorsal aorta. Recognition of this anomaly has clinical importance because it may be associated with symptoms of tracheaoesophageal compression, aneurysm formation or rupture of the aneurysms. Multidetector computed tomographic (MDCT) angiography is able to display the detailed anatomy of the vascular structures and their spatial relationships with adjacent organs and this ability is the most important advantage of MDCT angiography over other imaging modalities in the assessment of the aortic arch abnormalities. In this report, we present the MDCT angiography findings of a rare case of a right aortic arch with aberrant left subclavian artery associated with an aneurysmal diverticulum of Kommerell causing dysphagia.

Key Words: Subclavian artery; aorta, thoracic; congenital abnormalities; tomography, X-Ray computed; angiography

ÖZET Sağ aortik arkdan köken alan aberran sol subklavyen arter nadir rastlanılan bir konjenital aortik ark anomalisidir. Kommerell divertikülü, aberran subklavyen arterin aortadan çıkışına yakın proksimal kesiminin dilatasyonu olarak tanımlanır ve dorsal aortanın embriyonik artığını temsil eder. Bu anomalinin farkında olmak, trakeaözofajial kompresyon semptomları, anevrizma oluşumu ve ya anevrizma rüptürü ile ilişkili olabilmesi nedeniyle klinik öneme sahiptir. Multidedektör bilgisayarlı tomografik (MDBT) anjiyografi, vasküler yapıların ayrıntılı anatomisini ve komşu organlarla ilişkilerini gösterebilme yeteneğindedir ve bu yetenek aortik ark anomalilerinin değerlendirilmesinde MDBT anjiyografinin diğer görüntüleme modalitelerine olan en önemli avantajıdır. Biz bu raporda disfajiye neden olan anevrizmatik Kommerell divertikülünün eşlik ettiği aberran sol subklavyen arterli sağ aortik ark olgusunun MDBT anjiyografi bulgularını sunuyoruz.

Anahtar Kelimeler: Subklavian arter; torasik aorta; konjenital anomaliler; bilgisayarlı tomografi; anjiyografi

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Congenital anomalies of the aortic arch and its main branches are rare and may be associated with other congenital cardiovascular diseases. Aberrant right subclavian artery originating from the normal left sided aortic arch is the most common aortic arch anomaly, with a reported incidence of 0.5-2%. The right aortic arch is relatively uncommon anomaly,

occuring in approximately 0.05% of the population.[1] Right aortic arch has been classified into three types according to the branching of the arch vessels. Of these, right aortic arch with aberrant left subclavian artery (ALSA) is the most common type.[1] An aortic diverticulum also known as Kommerell's diverticulum may be present at the origin of ALSA, representing the remnant of the embronic left dorsal aortic root. It is considered present when the diameter of the subclavian artery near its origin from the aortic arch is at least twice the size of its diameter more distally.[2] Recognition of this anomaly has clinical importance because it may be associated with symptoms of tracheaoesophageal compression, aneurysm formation or rupture of the aneurysms.[3-5] By tradition, aortic anomalies have usually diagnosed by conventional angiography. Recently, multidetector computed tomographic (MDCT) angiography has become a principal diagnostic method for the evaluation of aortic abnormalities and challenged the role of conventional angiography.[6,7] In this report we present the MDCT angiography findings of a rare case of a right aortic arch with ALSA associated with an aneurysmal diverticulum of Kommerell presenting as dysphagia.

CASE REPORT

A 52-year-old woman presented with intermittent dysphagia for solids, which had been progressive during the last two years. Her physical exam was unremarkable. A chest radiography showed superior mediastinal widening and aortic knob located to the right of the trachea. The oesophagogram could not be carried out because the patient did not tolerated the barium. An endoscopy showed a pulsating mass compressing the oesophagus. MDCT angiography was performed with a 16-row MDCT (Lightspeed Ultra, General Electrical Medical Systems, Milwaukee, Wisc.; USA). All images were acquired during a single breathhold, extending from base of the neck to the diaphragma. Imaging parameters were as follows: 120 kV, 16x1.25 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 bolus tracking technique, images were acquired 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 image 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 shows that the first branch arising from the right aortic arch is left carotid artery, which is followed by the right carotid artery, right subclavian artery and ALSA. It revealed an aneurysmal diverticulum of Kommerell at the proximal part of the ALSA compressing the posterior wall of the oesophagus and trachea (Figure 1, 2). The oesophagus appeared as dilated above the compression level. There was also a mild dilatation of the descending aorta. Based on these MDCT angiography findings, the patient was diagnosed as having right sided aortic arch with ALSA associated with an aneurysmatic diverticulum of Kommerell and she was operated due to progression of her dysphagia.

DISCUSSION

ALSA originating from a right sided aortic arch is a rare aortic arch anomaly. In this anomaly, the first branch arising from the aortic arch is left carotid artery, which is followed by the right carotid, right subclavian arteries, and ALSA in that order. According to Edward's hypothetical double aortic arch model; the right aortic arch with ALSA results from interruption of the dorsal segment of the left arch between the left common carotid and left subclavian arteries with regression of the right ductus arteriosus.[1]

An ALSA is generally asymptomatic and has been diagnosed incidentally, but rarely adults with this anomaly have dysphagia because of extrinsic compression of the oesophagus due to its retrooesophageal course.[7,8] Kommerell's diverticulum can compress trachea or oesophagus either directly, by aneurysmal dilatation, or age related atherosclerotic changes.[3,5] Rarely, with failure in

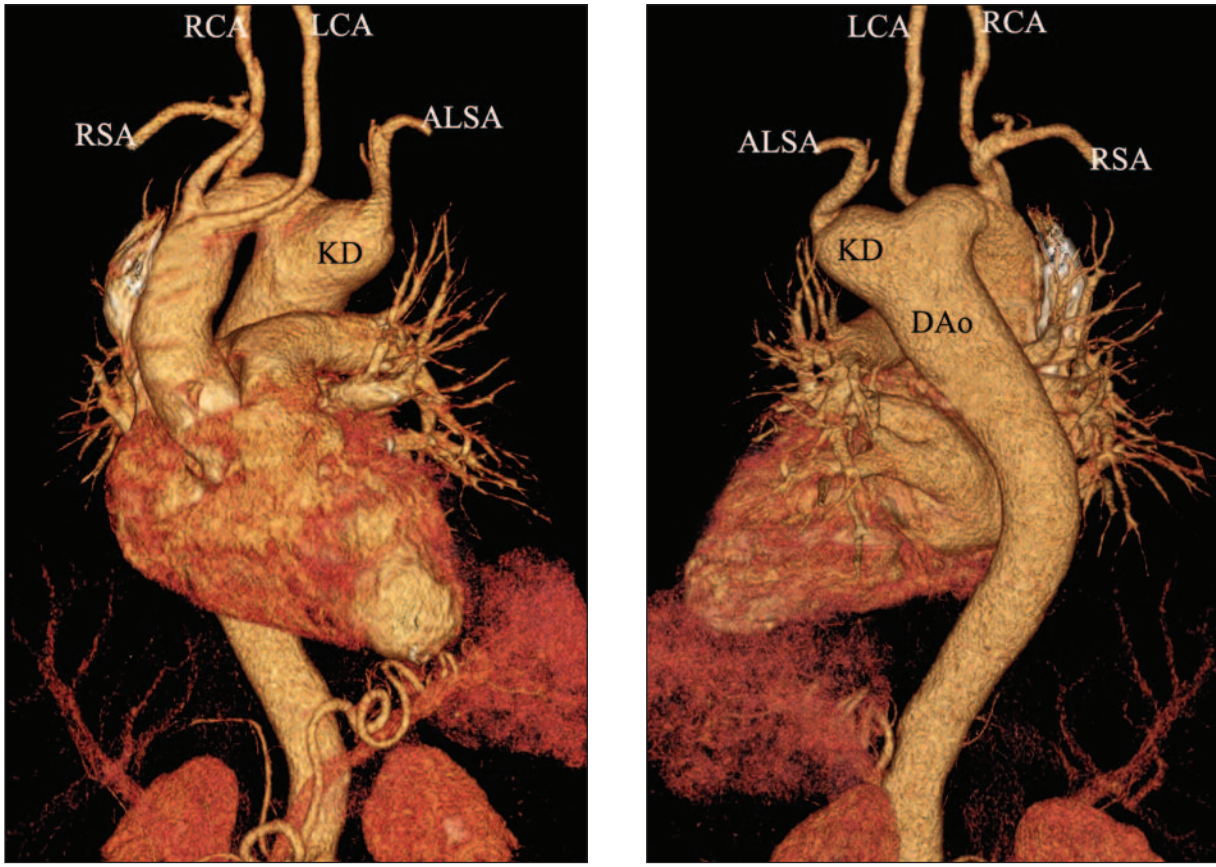
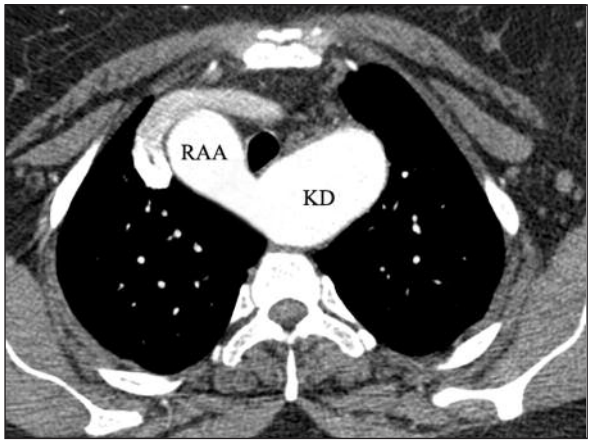


FIGURE 1: Coronal anterior (A) and posterior (B) three-dimensional volume rendering multidetector computed tomography images show that the first branch arising from the right aortic arch is left carotid artery (LCA), which is followed by the right carotid artery (RCA), right subclavian artery (RSA), and aberrant left subclavian artery (ALSA). An aneurysmal diverticulum of Kommerell (KD) is present at the proximal part of the ALSA. There is also a dilatation of the proximal descending aorta (DAo).

regression of the left ductus, a loose vascular ring may be formed by the right arch with ALSA, left pulmonary artery, and left ductus arteriosus.[1] Respiratory compromise due to tracheal compression may be present in especially pediatric patients.[6,8] An elderly patient may present with dysphagia when ALSA becomes tortuous and ectatic. Aneurysmal dilatation of the proximal portion of ALSA is uncommon. Such an aneurysm may be associated with Kommerell's diverticulum and is believed to the result of atherosclerotic disease. Symptoms are related to pressure of the aneurysm on adjacent structures and include dysphagia, dyspnea, cough, hoarseness, superior vena cava syndrome and Horner's syndrome.[5] Fatal rupture of these aneurysms has been reported. Early diagnosis and elective surgical management of these aneurysms seems necessary because of high mortality rate associated with rupture.[9] Generally, an aberrant left subclavian artery is an isolated anomaly, but may be associated with other cardiovascular anomalies such as tetralogy of Fallot, atrial septal defect, ventricular septal defect or coarctation of aorta.[1]

MDCT angiography is a noninvasive and reliable technique for diagnosis of thoracic vascular anomalies. With MDCT, images can be reconstructed in various planes and used as a road map for endovascular and surgical treatment planning. Although axial images are diagnostic for the evaluation of aortic arch position, aberrant vessels and vascular compression of the trachea and oesopha-



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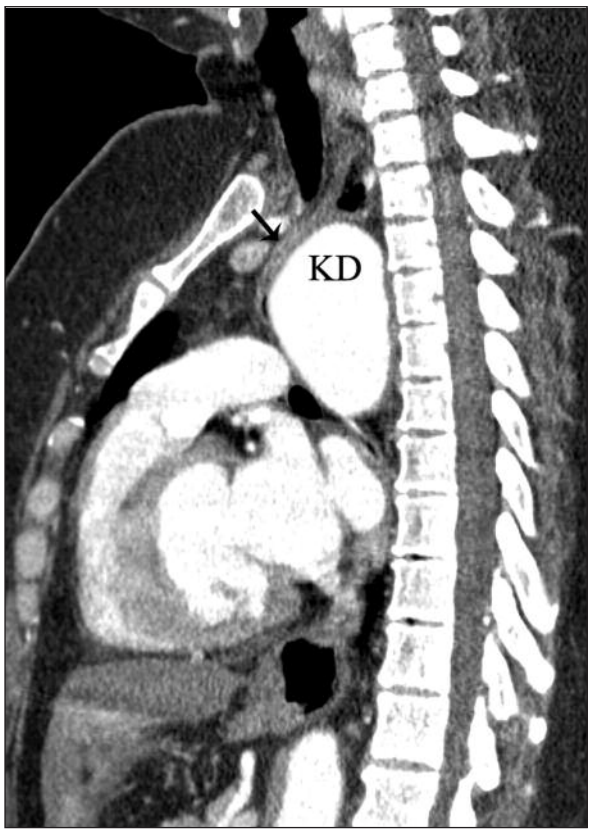

B

FIGURE 2: Axial (A) and sagittal multiplanar reformatted (B) multidetector computed tomography images show an aneurysmal diverticulum of Kommerell (KD) causing tracheal and oesophageal compression (arrow). (RAA=Right aortic arch)).

gus, multiplanar and three-dimensional volume rendered images can enhance the diagnostic value of MDCT. With MDCT, supraaortic branches can be well delinaeted with excellent image quality.[6,7] The disadvantages of this method especially in pediatric patients, are to need iodinized contrast material and radiation exposure. Magnetic resonance (MR) angiography is an alternative non-invasive tool in the evaluation of thoracic vascular anomalies without the need for iodinated contrast material or radiation exposure. But it is time-consuming and, may require prolonged sedation in pediatric patients. In addition, MR images are prone to cardiac or respiratory artifacts.[10]

In conclusion, recognition of the ALSA has clinical importance because it may be associated with clinical symptoms of tracheaoesophageal compression, aneurysm formation, rupture of the aneurysm and be accompanied by other vascular and cardiac anomalies. In patients with unexplained dysphagia, an aberrant subclavian artery should be considered as a possible cause. MDCT angiography is able to display the detailed anatomy of the vascular structures and their spatial relationships with adjacent organs and this ability is the most important advantage of MDCT angiography over other imaging modalities in the assessment of the aortic arch abnormalities.

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