

# Demonstration of vascular abnormalities compressing esophagus by MDCT: Special focus on dysphagia lusoria

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## Abstract

**Purpose:** Dysphagia lusoria (DL) is described in the literature as difficulty in swallowing caused by vascular abnormalities. The most common cause is an aberrant right subclavian artery (SCA) which passes behind the esophagus and is also called arteria lusoria (AL). Our aim was to demonstrate the use of multidetector computed tomography (MDCT) in the diagnosis of AL, as there is no comprehensive study investigating the role of MDCT in such cases.

**Material and methods:** A total of 38 consecutive patients, comprising of 23 females (61%) and 15 males (39%), who had extrinsic compression were included in the study. These patients are selected from the cases who were admitted due to their gastrointestinal symptoms, such as dysphagia, epigastric pain, chronic nausea, vomiting, etc. The mean age of patients was  $40 \pm 25$  years (range 15–65). Following barium esophagogram and then endoscopy performed, MDCT angiography was carried out on the same or the following few days. MDCT sections were examined to determine the following: presence of vascular abnormality; the diameter and angle of that vascular structure; and the compressed area of esophagus. Radiological findings and dysphagia scores were also compared.

**Results:** In each of 15 cases, there was a compression due to vascular abnormality which were all located between the esophagus and the spine. There was an esophageal compression in each of 12 cases, due to right aberrant SCA, in one case due to right superior aortic arch and in two cases due to both right aortic arch and left SCA with Kommerell's diverticulum. The mean diameter and the angle of AL were 16.4 mm and  $48.8^\circ$ , respectively, and the mean area of pressured esophagus was  $194.7 \text{ mm}^2$ . Dysphagia scores of the cases was 1 in thirteen cases and 2 in two cases. However, dysphagia scores were not correlated with these parameters.

**Conclusions:** MDCT angiography is a useful diagnostic tool for evaluation of patients with dysphagia, especially caused by a vascular abnormality.

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**Keywords:** Dysphagia lusoria; Esophagus; MDCT

## 1. Introduction

Dysphagia lusoria (DL) is described in the literature as difficulty in swallowing. It is a birth defect encom-

passing any vascular ring anomaly (arteria lusoria: AL) that causes esophageal dysphagia [1]. The term “vascular ring” describes malformations of the aortic arch that render the esophagus or trachea partially or completely entrapped. The most common congenital abnormality of the aorta is an isolated aberrant right subclavian artery (SCA) [2,3]. A right aortic arch with an aberrant left SCA is less common, but may also result in esophageal compression [4].

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Vascular abnormalities do not usually lead to the symptoms, however, sometimes a dysphagia due to mass effect on esophagus, which is also called as DL, may develop. Abnormalities of the aortic arch and thoracic aorta are not uncommon and can result in esophageal compression and dysphagia. They can press on trachea and result in dyspnea, especially in children [5,6].

Several studies have used chest X-ray, barium esophagogram, and endoscopy in the evaluation of dysphagia. However, in the accurate and exact diagnosis of the abnormal vascular structures, computerized tomography (CT) and magnetic resonance angiography (MRA) or digital subtraction angiography (DSA) are needed. Recent advances in computed tomography techniques as multidetector scanners make it possible to visualize the vascular structure in detail. Multidetector computerized tomography (MDCT) is a reliable and noninvasive tool for diagnosing vascular abnormalities [7]. For this reason, MDCT can also detect the real incidence of arteria lusoria in vivo. We evaluated the presence of arteria lusoria in patients with dysphagia who are examined with multi-detector computerized tomography angiography (MDCTA).

## 2. Materials and methods

### 2.1. Patient selection and premedication

A total of 38 consecutive patients who have gastrointestinal symptoms such as dysphagia, epigastric pain, chronic nausea, vomiting but do not have any lumen pathology such as esophagitis, mass and the other intrinsic pathologies with barium esophagography and endoscopy, were included in this study. Twenty-three of patients were female (61%) and 15 of patients were male (39%). The mean age of patients was  $40 \pm 25$  years (with range 15–65).

In these cases, their dysphagia scores were found and categorized in increasing severity as follows: grade 0: normal swallowing; grade 1: unable to swallow solids; grade 2: unable to swallow semisolids; grade 3: unable to swallow liquids; grade 4: unable to swallow own saliva [8].

After initial diagnostic examination, MDCTA was carried out on the same day or the following few days. To obtain an optimal esophageal distention at a scout image in MDCTA, 5 g of bubble-making granules (sodium bicarbonate and 2,3-dihydroxybutanedioic acid; Baritogen Fushimi, Marugame, Japan) mixed with water were given orally to fasting subjects. Then MDCTA was performed after obtaining optimal esophageal distention.

The procedures used were in accordance with the recommendations announced in the Helsinki declaration. Informed consent was obtained from all patients.

### 2.2. MDCT protocol, image analysis

Multidetector computed tomography was performed on a 16-detector-row CT scanner (Aquillon; Toshiba Medical Sys-

tems, Tokyo, Japan) during one breath hold time (24–30 s). Scans were obtained with  $16 \times 0.5$ -mm collimation, 1.0-mm slice thickness and 1.0-mm reconstruction interval. Ninety milliliters iodinated contrast medium (Omnipaque, Amersham Health, Cork, Ireland) was injected intravenously into the antecubital vein, at a rate of 4.5 ml/s. The sections were taken from the lower cervical region to the top of the liver.

Images were then transferred to a processing workstation for further analysis with specialized software (Vitrea 2, Vital Images, Inc., Minneapolis, MN). In addition to the traditional axial images, all the other available techniques (multiplanar reconstructions, curved multiplanar reformation, sliding thin-slab maximum intensity projection (MIP) and three dimensional volume rendering (3D VR)) images were used for the assessment of the thoracic vascular structures.

All MDCTA studies were retrospectively reviewed by two radiologists who were aware of endoscopic and barium esophagogram findings. MDCT sections were examined to determine the following: presence of vascular abnormality; the diameter and angle of that vascular structure; the compressed area of esophagus which is measured in the sections (especially with coronal MIP images) that esophagus and arteria lusoria superposed; esophagus wall thickness and presence of extra esophageal lesions (mass, vessels, lymph nodes, trachea, mediastinal fat, lung, vertebral colon and upper abdominal organs).

Radiological findings in cases with arteria lusoria are compared to the dysphagia scores of the same cases.

### 2.3. Statistical analysis

Data were analyzed using the statistical software SPSS for windows version 10.0. Spearman's rank order correlation ( $\rho$ ) was used to determine the correlation between dysphagia scores and other parameters including the diameter of AL and compressed area of esophagus. A  $p$ -value less than 0.05 was considered to indicate statistical significance. Data were expressed as mean  $\pm$  S.D.

## 3. Results

With MDCT, there was an esophageal compression due to vascular abnormality in 15 (40%) cases and mediastinal masses in 23 (60%) cases. Of the cases with vascular abnormality only nine (60%) had positive results with barium esophagogram or endoscopy. Of the cases with mediastinal masses 14 had multiple lymphadenopathy (8 due to lymphoma and 6 due to tuberculosis), 3 bronchogenic cyst, 2 neurogenic tumors, 2 bronchial carcinomas, 1 esophageal leiomyoma and 1 posterior mediastinal hydatid cyst.

Of the 15 cases with AL 9 were women and 6 were men. The detected AL in all cases was between the esophagus and spine. There was an esophageal compression in 12 cases due to right aberrant SCA (Fig. 1), in one case due to right superior aortic arch (Fig. 2) and in two cases due to both right

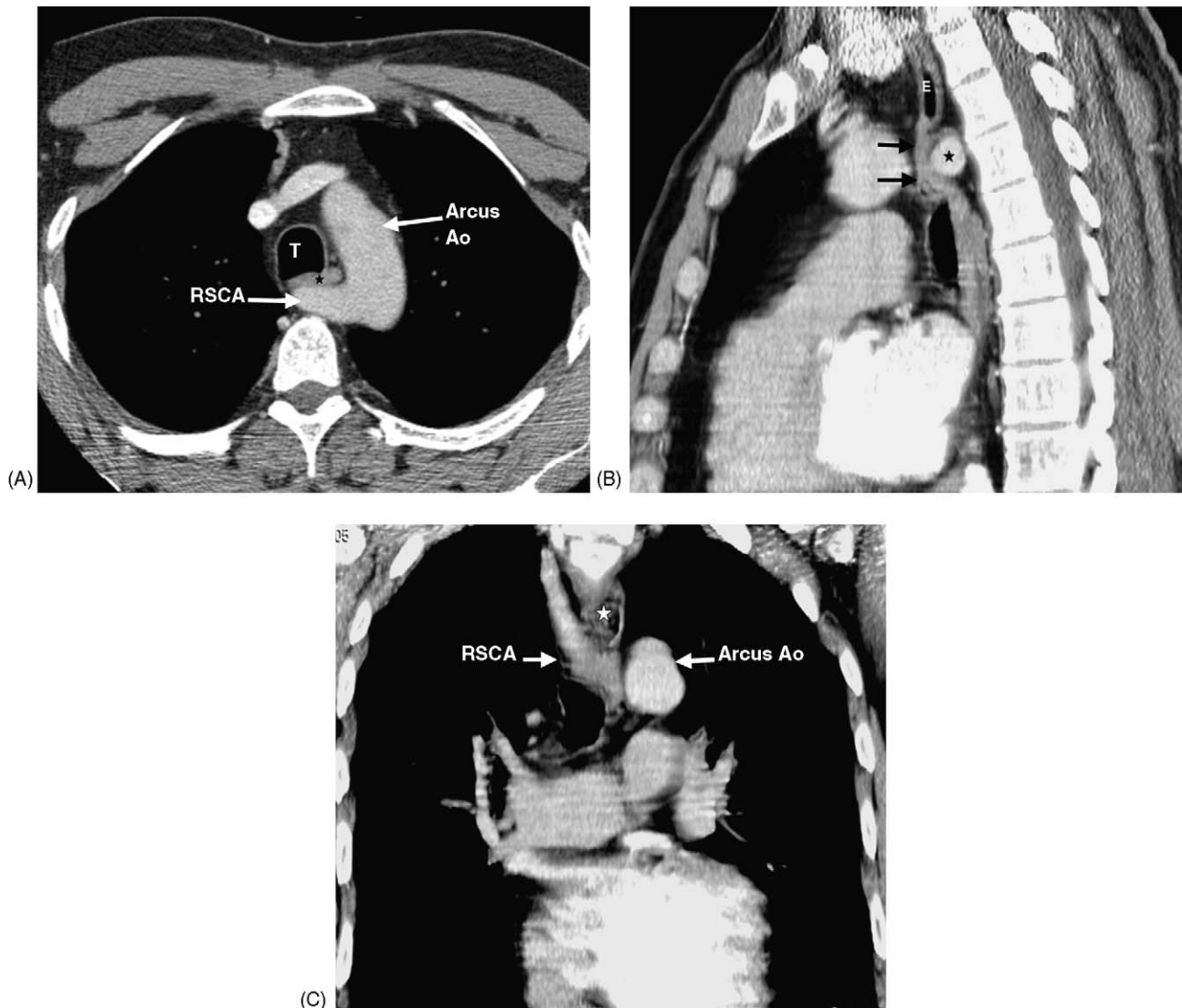


Fig. 1. A patient with aberrant right subclavian artery. (A) Axial MDCT image shows aberrant right subclavian artery (RSCA) and compressed esophagus (black star). T: trachea; Arcus Ao: aortic arch or arcus aortae. (B) Sagittal MDCT image shows compression of esophagus (black arrows) and its proximal dilatation (E). It also shows aberrant RSCA (black star) placed between spine and esophagus. (C) Coronal MDCT image shows RSCA crossing the spine from left to right side. White star: esophagus; Arcus Ao: aortic arch.

aortic arch and left aberrant SCA with Kommerell's diverticulum (Fig. 3). The compression was caused by right aberrant SCA placed between spine and esophagus in 12 cases, by distal of aortic arch in one case, by Kommerell's diverticulum extending between spine and esophagus in two cases.

The course of AL of all cases was between Th2 and Th4. The mean diameter of AL was  $16.4 \pm 4.3$  mm. The AL was arising from the aortic arch with an average angle  $48.8 \pm 10.7^\circ$ . The mean area of compressed esophagus was  $194.7 \pm 90.9$  mm<sup>2</sup>.

Of the cases with AL, all of them had dysphagia. Dysphagia scores of the cases were 1 in 13 cases and 2 in 2 cases. Dysphagia scores were not correlated with the measurements of AL ( $\rho=0.13$ ,  $p=0.64$  with diameter,  $\rho=0.16$ ,  $p=0.90$  with angle and  $\rho=0.31$ ,  $p=0.14$  with compressed area). Surgery was recommended for two patients who had

severe dysphagia (dysphagia score: 2), but those patients did not accept surgery. All cases were followed up with a medical treatment.

#### 4. Discussion

Esophageal dysphagia is mainly caused by esophageal cancer, esophageal stricture and webs, achalasia, diffuse esophageal spasm and esophagitis [9,10]. Rarely, it may also result from extrinsic causes including mediastinal mass (such as thyroid carcinoma, lymphoma and germ cell tumor), aortic aneurysm, vertebral spur and AL. Dysphagia lusoria is used to describe symptomatic extrinsic compression of the esophagus from any vascular abnormality of the aortic arch which is called as AL and was first described by Bayford in 1787

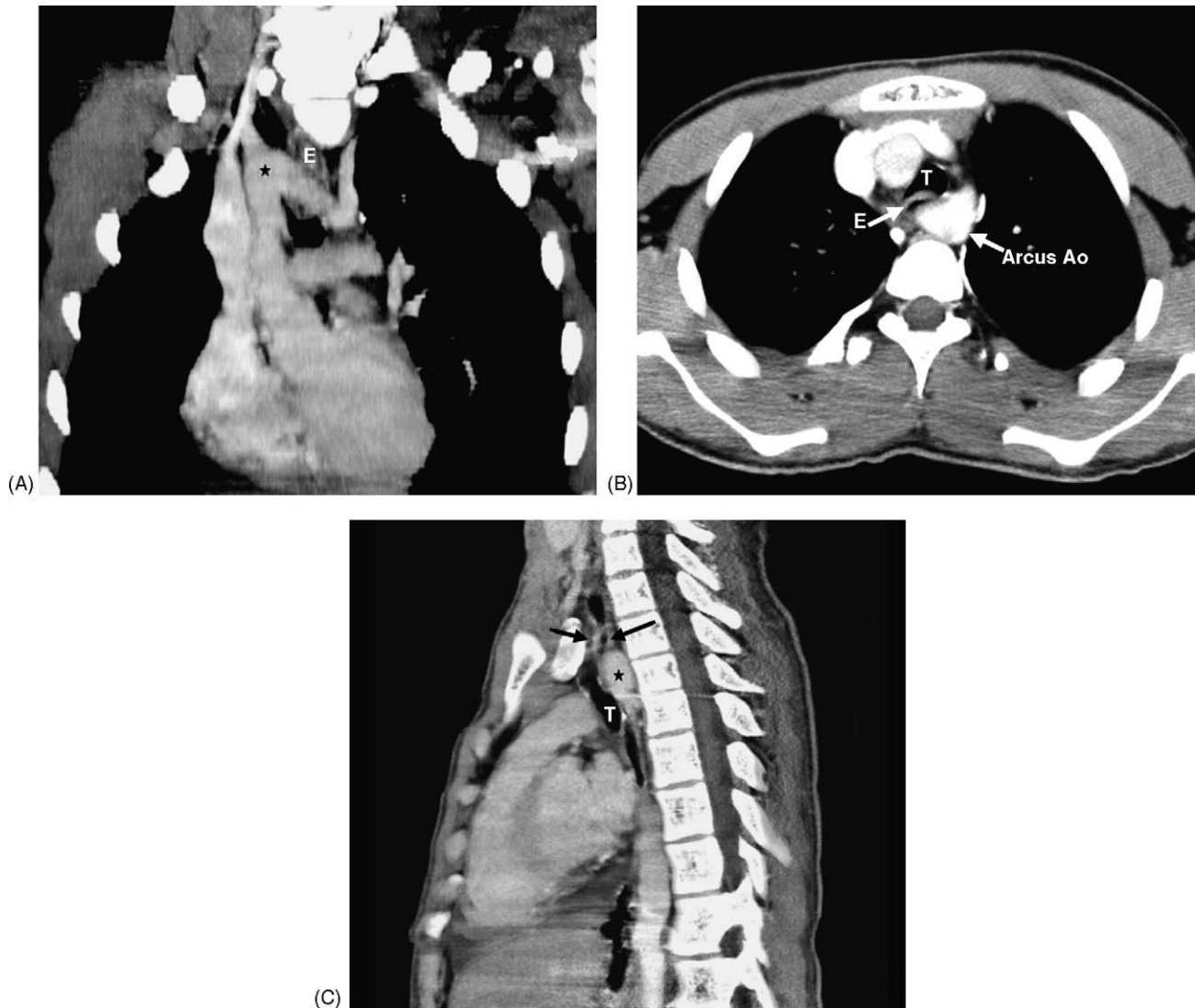


Fig. 2. A patient with superior and right aortic arch. (A) Coronal MDCT image shows right sided aortic arch (black star); E: esophagus. (B) Axial MDCT image shows distal of aortic arch passing behind of esophagus. T: trachea; E: esophagus; Arcus Ao: aortic arch. (C) Sagittal MDCT image shows compression due to aberrant aortic arch (black star); T: trachea. Black arrows: dilated proximal esophagus.

[11]. Additionally, there have been some articles suggesting that dysphagia lusoria may result after the development of atherosclerosis and dilatation due to aneurysm [12,13].

The most common embryologic abnormality of the aortic arch is an aberrant right SCA, which occurs in 0.5–1.8% of the population [14,15]. As hypothesized by Edwards, this abnormal origin of the right SCA can be explained by the involution of the fourth vascular arch with the right dorsal aorta [16].

The diagnostic modalities available to visualize an AL include chest roentgenogram, barium esophagogram, CT, MRA, DSA and endoscopy. Because of new advances in CT technology, even small vascular structures can be visualized in detail. MDCTA is now an established diagnostic test in the evaluation of many vascular diseases [17]. We evaluated the characteristics of AL with MDCT and also compared the dysphagia score with various measurements of AL. To our

knowledge, no larger patient group has been reported previously in the literature. No prior studies have been performed by using 16-detector MDCT.

It is unclear when the dysphagia symptoms onset and in which circumstances the AL causes the symptoms. The majority of AL cases are usually discovered during investigations for unrelated symptoms [18]. Mediastinal abnormalities may be seen on chest X-ray. However, the findings are usually indirect and include limited data.

The barium esophagogram is a useful method to evaluate the possibility of DL, but the diagnosis can be easily missed unless most of the superior thoracic esophagus is carefully examined and if lateral or oblique views of the esophagus are not obtained. In a study including 43 patients with dysphagia symptoms and using barium esophagogram vascular pathology has been demonstrated only in a case (2%) and no pathology revealed in 13 cases (30%). The other reasons of

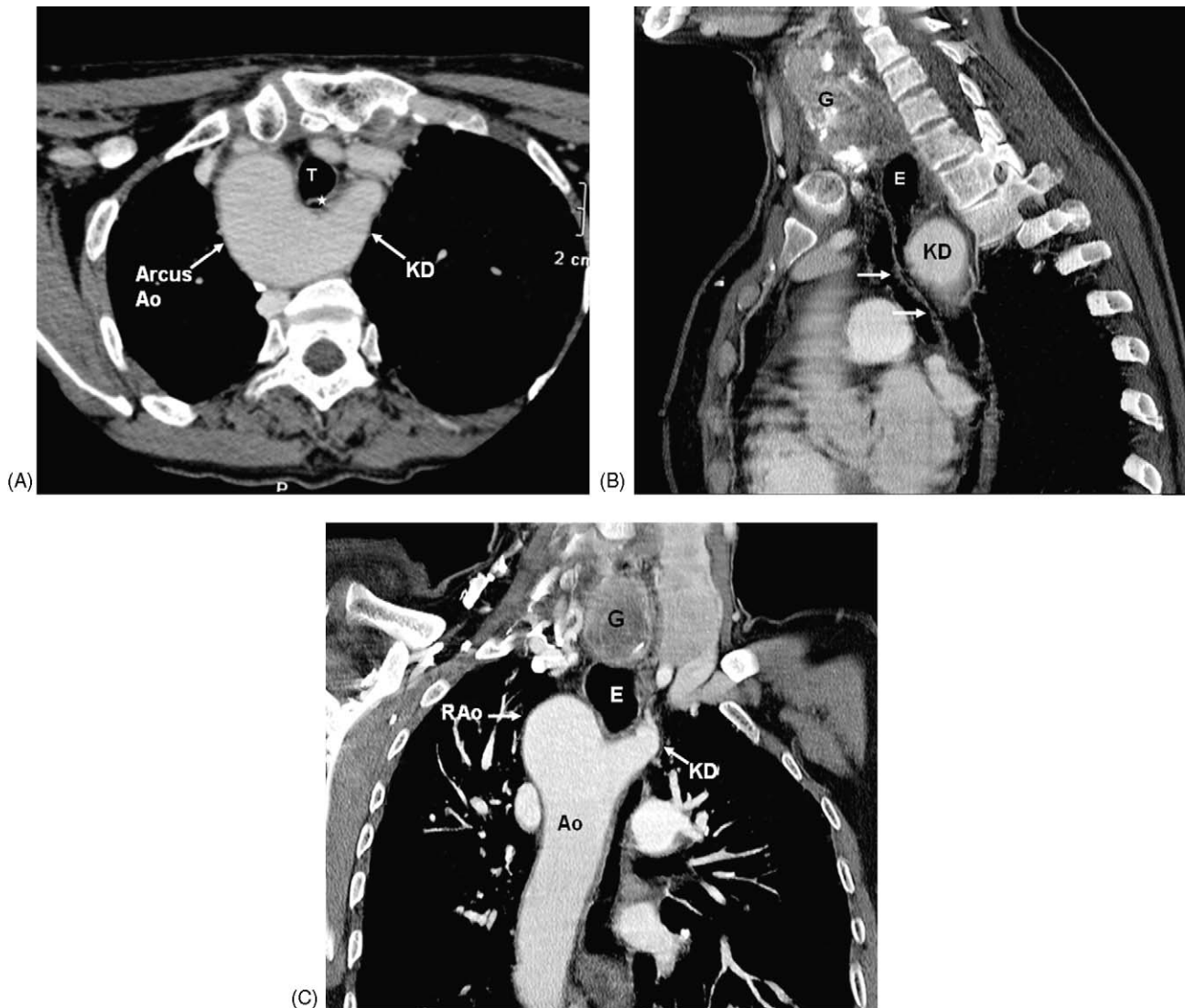


Fig. 3. A patient with right arcus aortae and left aberrant subclavian artery with Kommerell's diverticulum. (A) Axial MDCT image shows apparent compression of esophagus (white star). KD: Kommerell's diverticulum; T: trachea. Arcus Ao: aortic arch. (B) Sagittal MDCT image shows compression of esophagus (white arrows) by Kommerell's diverticulum (KD) placed between esophagus and dilatation of proximal esophagus (E); G: goiter. (C) Coronal MDCT image shows Kommerell's diverticulum (KD) extending from right to left side, right paravertebral course of thoracic aorta (Ao) and proximal esophageal dilatation (E). G: goiter; RAo: right aortic arch.

dysphagia in that study were esophageal dysfunction (25%), gastroesophageal reflux disease (19%), esophageal stricture (12%), pharyngeal dysfunction (5%), achalasia (5%), and esophagitis (2%) [19]. In our study, after exclusion of the cases with intrinsic esophageal pathology, we found vascular pathology in 15 of 38 cases (40%) with MDCTA. We also found that barium esophagogram was negative in 40% of the patient with AL. This result shows that barium esophagogram may underestimate a real incidence of vascular abnormalities. The findings of barium esophagogram are also nonspecific, because it only shows an extrinsic compression and cannot explain whether it is caused by DL.

Endoscopy may reveal pulsatile, shelf-like extrinsic compression in the posterior wall of the esophagus, with intact

mucosa. As shown in our study, it will give indirect findings in such cases; however, the associated risk of perforation and hemorrhage in patients with vascular rings make this test not so popular [20,21].

MRI has the advantage of being noninvasive and a patient is spared the potential risk of intravenous contrast agents. However, MRI images are not as useful as MDCT due to cardiac and respiratory motion artifacts. In addition, it is not a preferred method due to its cost and prolonged scan time. MR angiography may show a vascular anomaly, but the information regarding nonvascular mediastinal structures is not enough [22].

Digital subtraction angiography gives valuable information regarding AL. It is an invasive procedure and, in contrast

to MDCT, has the disadvantage in showing extravascular structures such as esophagus. It has also been shown that the effective radiation doses in MDCT angiography studies are moderate and even lower in comparison with DSA in a comparable patient group [23].

Combining of MDCT with 3D volume rendering images provides some additional advantages including not only for the depiction of the thoracic vascular anomalies but also for the assessment of the diameter, angle and compressed area of esophagus and its relationship with the esophagus and other mediastinal structures [24]. The other advantages of MDCT compared with MRA and DSA are its noninvasiveness (only DSA) and easy application and short time requirement (both DSA and MRA).

In this study, we also determined three different types of vascular abnormalities of the aortic arch which were manifested as dysphagia lusoria, which are aberrant right SCA, right aortic arch and aberrant left SCA with Kommerell's diverticulum. Some of these cases were rare abnormalities and their MDCT images were fairly detailed and better than the previously published ones. It is possible to obtain similar images by MRA and DSA also, but, as we mentioned above, MDCT has provided us some additional information regarding esophagus such as compressed area.

We also compared dysphagia scores with the measurements provided by MDCT, including diameter and angle of AL and compressed area of esophagus. However, we could not determine any significant correlation among them. The results suggest that there may be other factors in the development of dysphagia symptoms in such cases. Because these parameters could easily be measured by this method, it may be repeated in selected cases and may help to determine whether any change in these parameters is related to dysphagia.

Although our study is a preliminary one, it has some limitations due to the small sample size (i.e. small number of patients), and it has indicated that MDCTA provides a high quality and accurate modality to visualize and diagnose intrathoracic vascular abnormalities. The origin and course of all anomalous AL can be demonstrated clearly with a high average diagnostic image quality. It seems that MDCT has the potential to serve as a reliable initial diagnostic modality to use in evaluation of dysphagia.

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