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COMMENTARY

3D modeling and comparative analysis of the double arcus aorta case

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Abstract
We analyzed the double arch of a 51-year-old male patient who applied to the outpatient clinic with chest pain and shortness of breath and compared this rare case with the studies in the literature. Double aortic arch (DAA) is defined as a type of vascular ring malformation. The incidence of congenital heart diseases is less than 1%. DAA makes up 46–76% of all rings. We aimed to contribute to cardiac surgery by examining and modeling the diameters in the 2D and 3D images of the patient. For 3D modeling, an open-source software program ITK-SNAP 3.8 was used, which converts 2D images from MRI, CT, and ultrasound to 3D medical image volumes. CT images of the case taken from the SECTRA system of our hospital were uploaded to ITK-SNAP and segmentation was performed. With 3D modeling, a better understanding of the stenosis in the trachea and the double arch was achieved. The ascending aorta diameter was 30 mm. There were atherosclerotic changes in the aorta and its branches. The diameter of the right aortic arch was 22.2 mm, and the diameter of the left aortic arch was 14.5 mm. Trachea diameter was found to be 17 mm/13.2 mm. Esophageal diameter was 9.8 mm. The patient had no specific complaints and no medical or surgical treatment was recommended because his physical examination was normal. We think that a better understanding of such cases in 3D may contribute to cardiovascular surgery.

Keywords Double aortic arch · Variation · Congenital vascular malformation · ITK-SNAP · 3D modeling · Imaging

Introduction

Double aortic arch (DAA) is defined as a type of vascular ring malformation [1]. Its incidence is less than 1% of congenital heart diseases [2]. The Congenital Double Aortic Arch (DAA) which accounts for 46–76% of the complete rings is the most common vascular malformation in the congenital rings [3] [4]. Due to the persistence of the fourth aortic arches during embryonic development, both aortic arches are from the ascending aorta, bypassing the trachea and oesophagus, inflowing into the descending aorta [5]. DAA has no known ethnic or geographic disposition. DAA's mechanism is derived from a developmental anomaly of the original arterial arch in the embryonic term or chromosomal 22q11 microdeletion [6].

Tracheal and oesophageal compression is usually seen in patients with DAA. In the early stage, the symptoms of tracheal compression are more clear, such as repeated pulmonary infection, wheeze, shortness of breath, and so on. When eating liquid food, it is less hard to swallow due to esophageal stricture. The strain of swallowing is relatively obvious as times go on, so early intervention is very important [7].

By its anatomic characteristics, DAA has been divided into three types: large right posterior arch and small left anterior arch (right dominant aortic arch), which account for 75%; small right rear arch and large left front arch (left dominant aortic arch), account for nearly 20%; and equal diameter DAA (balanced type aortic arch), account for 5% [8].

Case report

A 51 year-old male patient was admitted to the Chest Diseases Polyclinic with complaints of chest pain and stinging in breathing. Computed tomography (CT) showed that

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the patient had a double aortic arch and the patient was referred to the Cardiovascular Surgery Policlinic.

In the echocardiography (ECHO) of the patient, there were no signs of heart failure, signs of cardiac hypertrophy were observed. Electrocardiography (ECG) was in sinus rhythm, there was no evidence of hypertension, and blood values were normal. The diameter of the right aortic arch was 22.2 mm, and the diameter of the left arch aorta was 14.5 mm. Ascending aorta diameter was 30 mm. There were atherosclerotic changes in the aorta and its branches (Fig. 1).

According to the literature, the diameter of the trachea taken from the CT image of a healthy person is anterior–posterior: 15 mm, right-left: 20 mm. measured [9]. In our case, the trachea diameter was found to be 17 mm/13.2 mm in the first image. It looks both narrowed and deformed in terms of aspect ratio (Fig. 2).

In healthy individuals, the esophagus diameter is between 10–20 mm on average [10]. A minimum of 14 mm is seen in anatomical stenosis. Although the place where we measured did not coincide with anatomical stenosis, the esophagus diameter seems to have decreased to 9.8 mm (Fig. 3) In

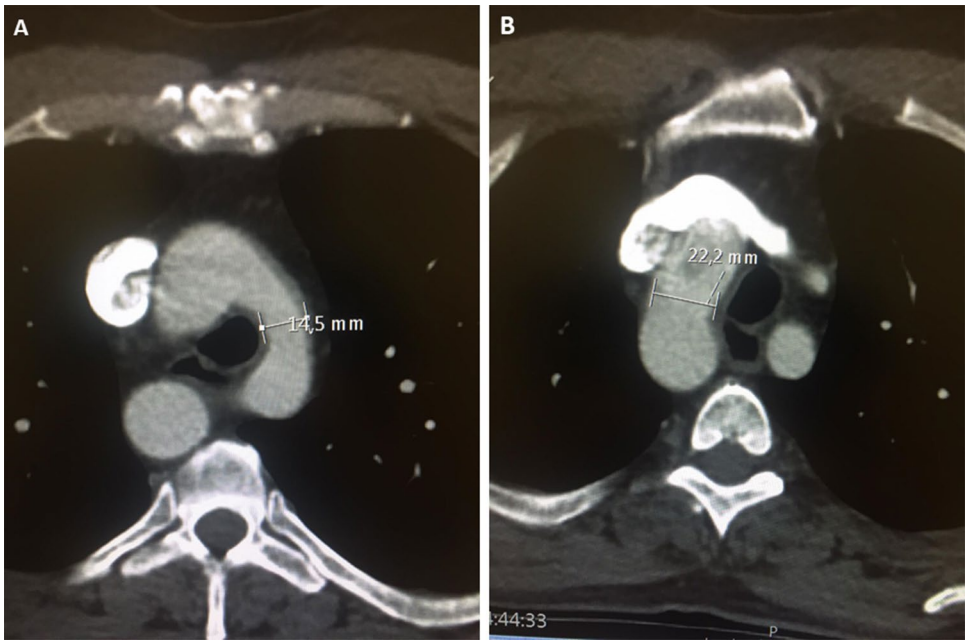

Fig. 1 Measurement of the diameters of the double aortic arch. **A** Minor arc diameter measured 14.5 mm, **B** Major arc diameter measured 22.2 mm

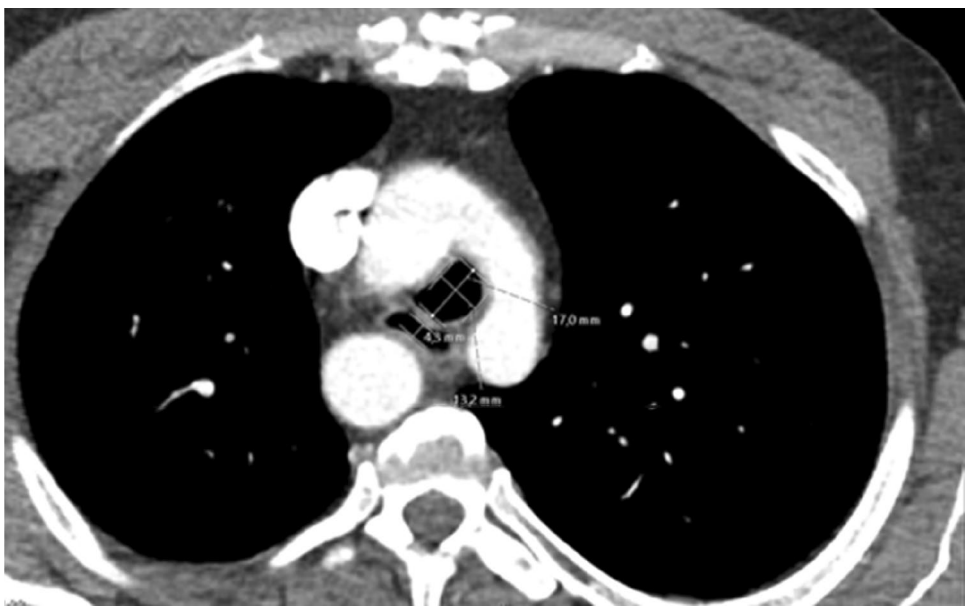

Fig. 2 Section was taken from the axial ct image of the case. Measurement of trachea diameters narrowed by the double arch (13.2 mm–17 mm)

addition, the short diameter of the esophagus decreased to 4.3 mm at the narrowing of the 2nd arcus aorta (Fig. 2).

The patient had no specific complaints and medical treatment was not initiated because his physical examination was normal.

For 3D modeling, a software program called ITK-SNAP, which converts 2D images in MRI, CT, and ultrasound into 3D medical image volumes, was used [11].

ITK-SNAP version 3.8 is an open-source toolkit built on the ITK C++ classes, widely used in academic and industrial environments [12]. CT images of the case taken from the SECTRA system of our hospital were uploaded to ITK-SNAP and segmentation was performed. With 3D modeling, a better understanding of the stenosis in the trachea and the double aortic arch was provided (Figs. 4, 5).

Discussion

Usually, in embryonic growing, the fourth left arch develops into the aortic arch, the fourth right arch develops into the innominate artery, the left dorsal aorta develops into the descending thoracic aorta, and the dorsal intersegmental arteries bilaterally develop into the subclavian arteries [13]. DAA is formed when both fourth arches and dorsal aortas leave a presence around the trachea and oesophagus. The right aortic arch is always larger and placed higher than the left aortic arch, which might be degraded or hypoplastic [1]. In this case, both aortic arches reunite to form the descending aorta placed in front of the spine, with the right aorta being larger than the left aorta [14].

The true happening of DAA in adults is unknown, but it is reported to be highly rare because it tends to cause hard respiratory symptoms and results in early diagnosis and correction in infancy or childhood [15].

DAA is the most usual type of complete vascular ring, it surrounds the trachea and esophagus, compresses them, and causes different degrees of tracheal stenosis, and esophageal obstruction. For patients with intractable wheeze, and recurrent lung infections, especially those with eating obstruction, the vascular ring may be suspected highly [6].

In summary, DAA is a rare congenital vascular malformation that can be easily misdiagnosed. ECHO combined with CT and airway reconstruction can effectively diagnose. After the diagnosis is made, it can be questioned whether there is compression in the trachea and whether surgical intervention is required. However, in patients with chronic wheezing, shortness of breath, recurrent cough, dysphagia, timely diagnosis, and surgical treatment are correct. Since the degree of tracheal stenosis caused by DAA is less than other vascular rings such as a pulmonary sling, it can be treated temporarily without simultaneous surgery and it is possible to return to normal with its growth and development [16].

Vascular rings account for less than 1% of all congenital cardiovascular anomalies. DAA is a congenital vascular malformation that can be easily misdiagnosed, and it is the most common type of vascular ring. DAA; is examined in 3 groups right dominant, left dominant, and balanced aortic arch. In 75% of the cases, the right arch, in 20% of the left arch dominant, and 5% of the cases, both arch dominance are equal.

Alsenaidi et al. stated in their study in 2006 that 71% of 81 cases with DAA had right arch dominance, 20% left arch dominance and 9% both arches were balanced [17]. Yang

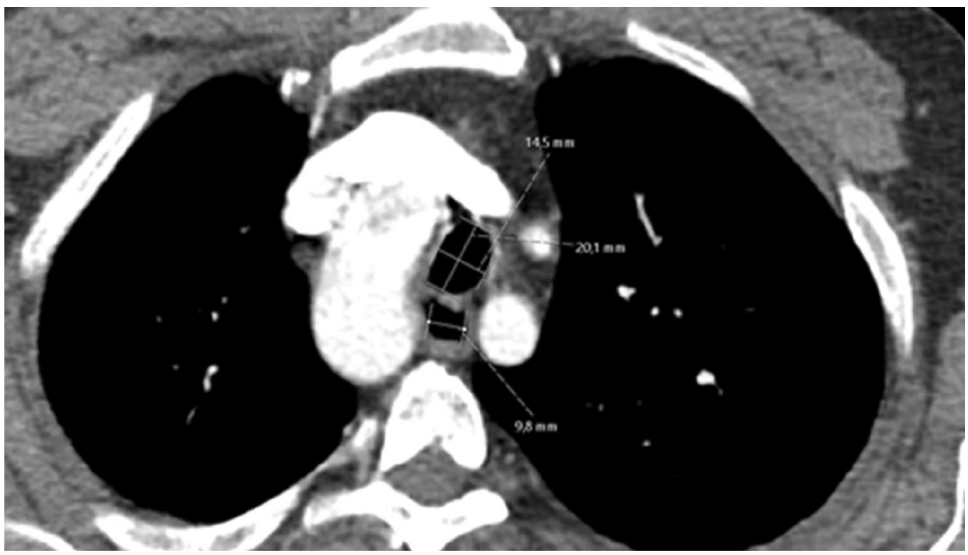

Fig. 3 Section was taken from the axial ct image of the case. Measurement of the diameter of the esophagus narrowed by the double arch (9.8 mm)

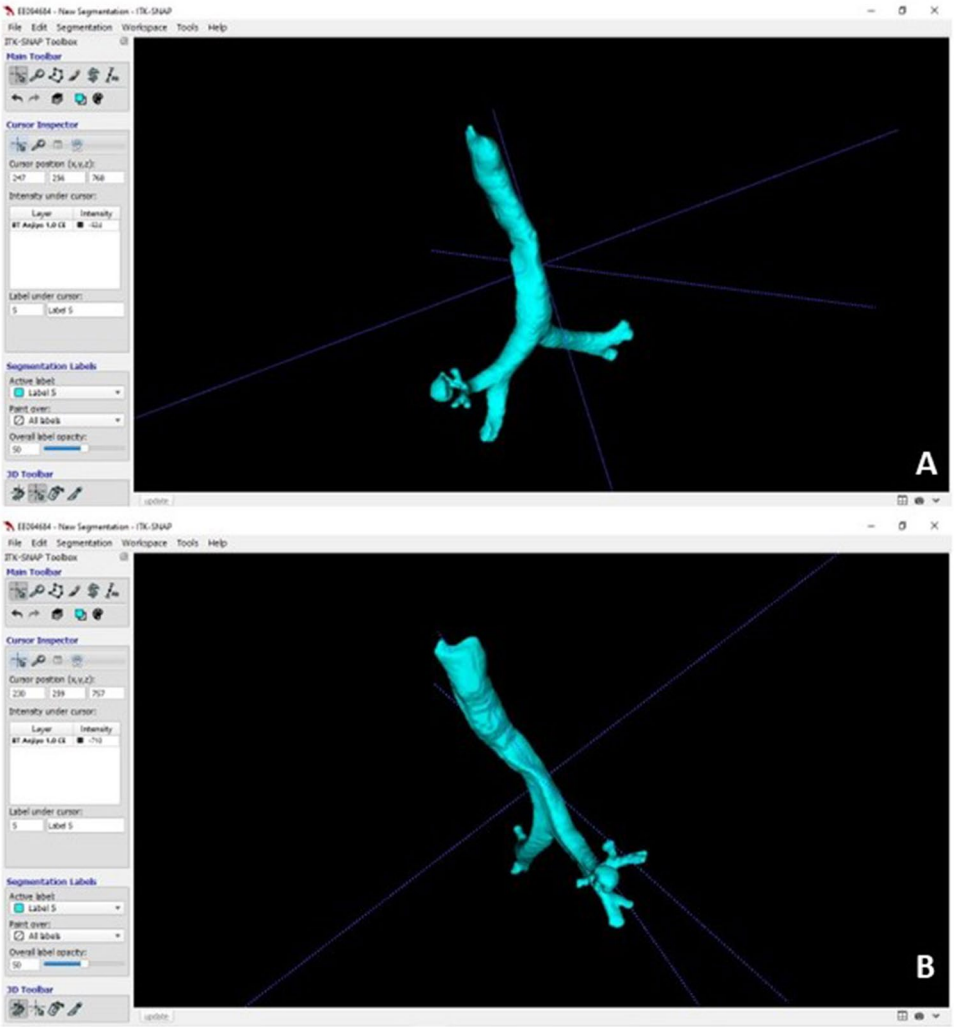

Fig. 4 3D trachea image of the CT image of the case created with the ITK-SNAP application, **A** front view, **B** bottom left view

et al. detected DAA with right dominance in a 3-month-old baby girl in 2019 [7]. In 2019, Zeigler et al. Reported a case of an 80-year-old female with a diagnosis of right-dominant DAA [18]. Again in 2019, Tsukioka et al. Reported a 71 year-old female case with a diagnosis of right dominant DAA [19].

In the Turkish population; Atay et al. Identified three cases in 4 years in their study in 2001, and they reported right arch dominance in two and equally dominance in one [20]. In 2012, Paç et al. Reported a 7 year-old girl with a left dominance of DAA [21]. Şen et al. Reported a right dominant DAA in a 30 year-old male patient in 2018. In our case, the right dominant arch was detected [22].

Conclusion

Currently, our patient does not have any compression symptoms. However, heart failure due to cardiomegaly may occur in the future in our patient, whose follow-up did not have any problems. If pressure increases due to anatomical variation cause enlargement of the aorta, rupture, dissection, and compression symptoms that may develop in the patient. To prevent complications that may develop in our patients, we think that it will be useful to monitor our patient's cardiac functions with regular ECHO follow-ups and to evaluate them with detailed contrast-enhanced CT if functions are observed. We think that we can contribute to the literature by sharing this rare case. The ITK-SNAP 3D model obtained from the CT images of this case will help cardiovascular surgery to better understand the case.

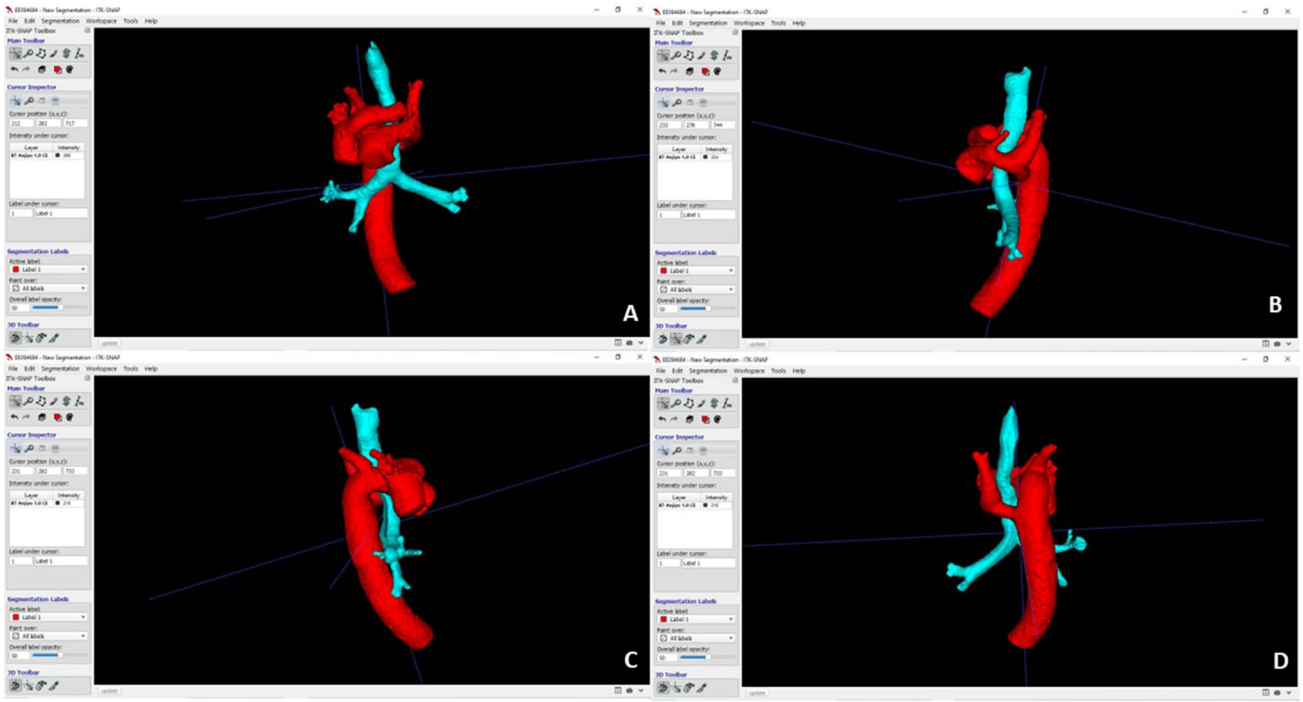

Fig. 5 3-dimensional trachea and double aortic arch view of the CT image of the case created with the ITK-SNAP application, **A** anterior view, **B** right view, **C** left view, **D** posterior view

Author contributions The design of the study was carried out by SA. The article was written by RD. Data collection and analysis was done by ME. 3D modeling and creation of figures was done by RD. Final reading and review done by SA. All authors have read and approved the final version of the article.

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Declarations

Conflict of interest The authors have no conflict of interest to declare regarding the materials or methods used in this study or the findings presented in this paper.

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