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Case Report

Right-sided Aortic Arch with Aberrant Left Subclavian Artery arising from Kommerell's Diverticulum: A case report ☆,☆☆

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ABSTRACT

Right-sided aortic arch, first documented by Fioratti and Aglietti in 1763, is a rare variant of the thoracic vascular anatomy present in about 0.1% of the adult population. In half of these cases the left subclavian artery is also aberrant. The aberrant left subclavian artery usually originates from a conical dilatation near its origin from the aorta also known as "Kommerell's diverticulum." Fewer than 80 of these cases have been reported in the literature as far as our web search is concerned. It is usually asymptomatic and diagnosed incidentally during adulthood. We are presenting a 56 years old male patient presented with right side chest and shoulder pain of 1 week duration. The pain exacerbated with motion of the right upper extremity and radiates to his lower back. However, he had no history of cough, shortness of breath, syncope, and dysphagia. The vital signs were in normal range. Pulmonary and cardiovascular exam were unremarkable. The complete blood count (CBC), electrocardiogram (EKG), and echocardiography showed no abnormality. In the adult population a right-sided aortic arch with an aberrant left subclavian artery arising from Kommerell's diverticulum is a rare occurrence often asymptomatic unless aneurysmal disease or compression of mediastinal structures ensues. Even though it is rare and at times an incidental finding, the condition is clinically relevant because of the morbidity caused by the complications. We report a case of Kommerell's Diverticulum of an aberrant left subclavian artery in an adult patient with a right-sided aortic arch. Right-sided aortic arch with aberrant left subclavian artery arising from Kommerell's Diverticulum is quite rare and may remain asymptomatic. On times it may cause symptoms in adulthood often as a result of

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early atherosclerotic changes of the anomalous vessels, dissection, or aneurysmal dilatation with compression of adjacent structures causing dysphagia, dyspnea, cough, or chest pain. Even though there are no general guidelines for the management of this condition patients need to be informed about the nature and possible outcomes of their condition. Close follow up of asymptomatic patients is one option of management until there are situations which require consideration of surgical intervention.

Introduction

Right-sided aortic arch, first documented by Fioratti and Aglietti in 1763, is a rare variant of the thoracic vascular anatomy present in about 0.1% of the adult population [1,2]. In half of these cases the left subclavian artery is also aberrant [2]. The aberrant left subclavian artery usually originates from a conical dilatation near its origin from the aorta also known as "Kommerell's diverticulum." Fewer than 80 of these cases have been reported in the literature as far as our web search is concerned. It is usually asymptomatic and diagnosed incidentally during adulthood [3,4].

Case presentation

A 56-years-old male patient presented with right side chest and shoulder pain of 1 week duration. The pain exacerbated with motion of the right upper extremity and radiates to his lower back. However, he had no history of cough, shortness of breath, syncope, and dysphagia. The vital signs were in normal range. Pulmonary and cardiovascular exam were unremarkable. The complete blood count (CBC), electrocardiogram (EKG), and echocardiography showed no abnormality.

Chest radiograph done to evaluate for the cause of the chest pain showed widening of the superior mediastinum, the tracheal was deviated to the left side and was indented on the right side. The trachea was also displaced anteriorly with posterior impression seen on the trachea just at the level of aortic arch on the lateral film. The lung parenchyma and pulmonary vascular pattern were normal. The outline of the descending thoracic aorta was seen slightly to the right of the spinal column (Figs. 1A and B).

With the above X-ray findings, right side aortic arch was diagnosed and further imaging with contrast enhanced chest computed tomography (CT) was done with reconstructed images in coronal and sagittal planes for detailed evaluation of the mediastinal vascular pattern. On the CT the ascending aorta was seen arching over the right main bronchus between the superior vena cava on the right and the trachea on the left

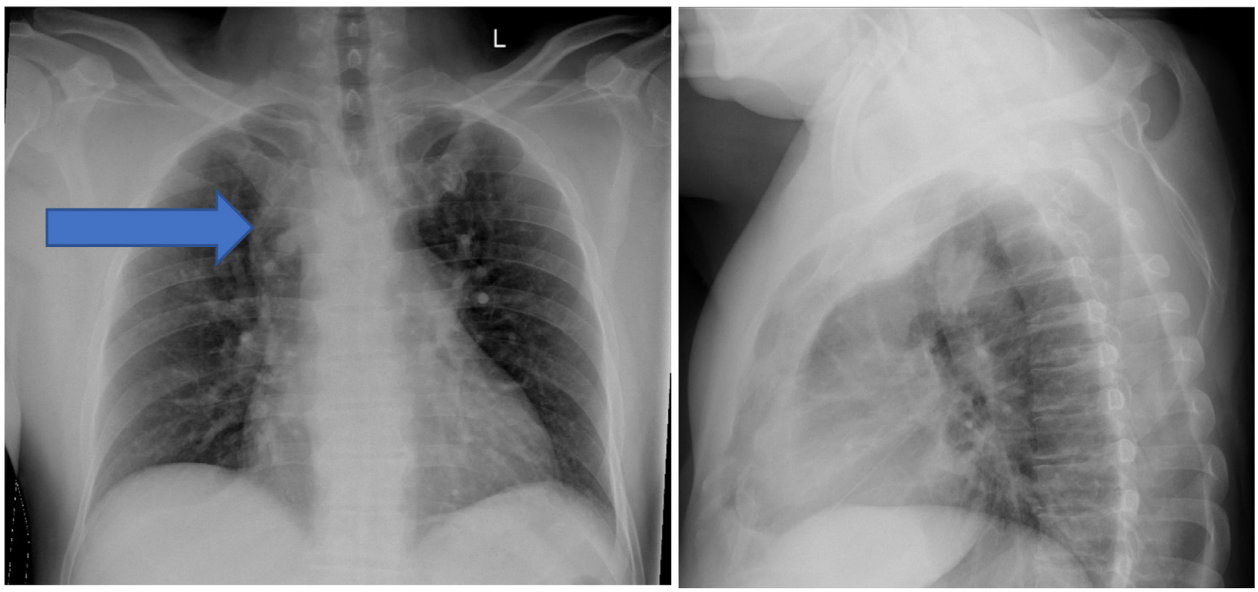

Fig. 1 – (A and B) Supine AP and Lateral CXR showing widening of the superior mediastinum, the tracheal deviated to the left side with indentation on the right side. The aortic Knob is seen on the right side. The lateral view shows the trachea displaced anteriorly with posterior impression seen on it just at the level of aortic arch. The outline of the descending thoracic aorta can be seen slightly to the right of the spinal column.

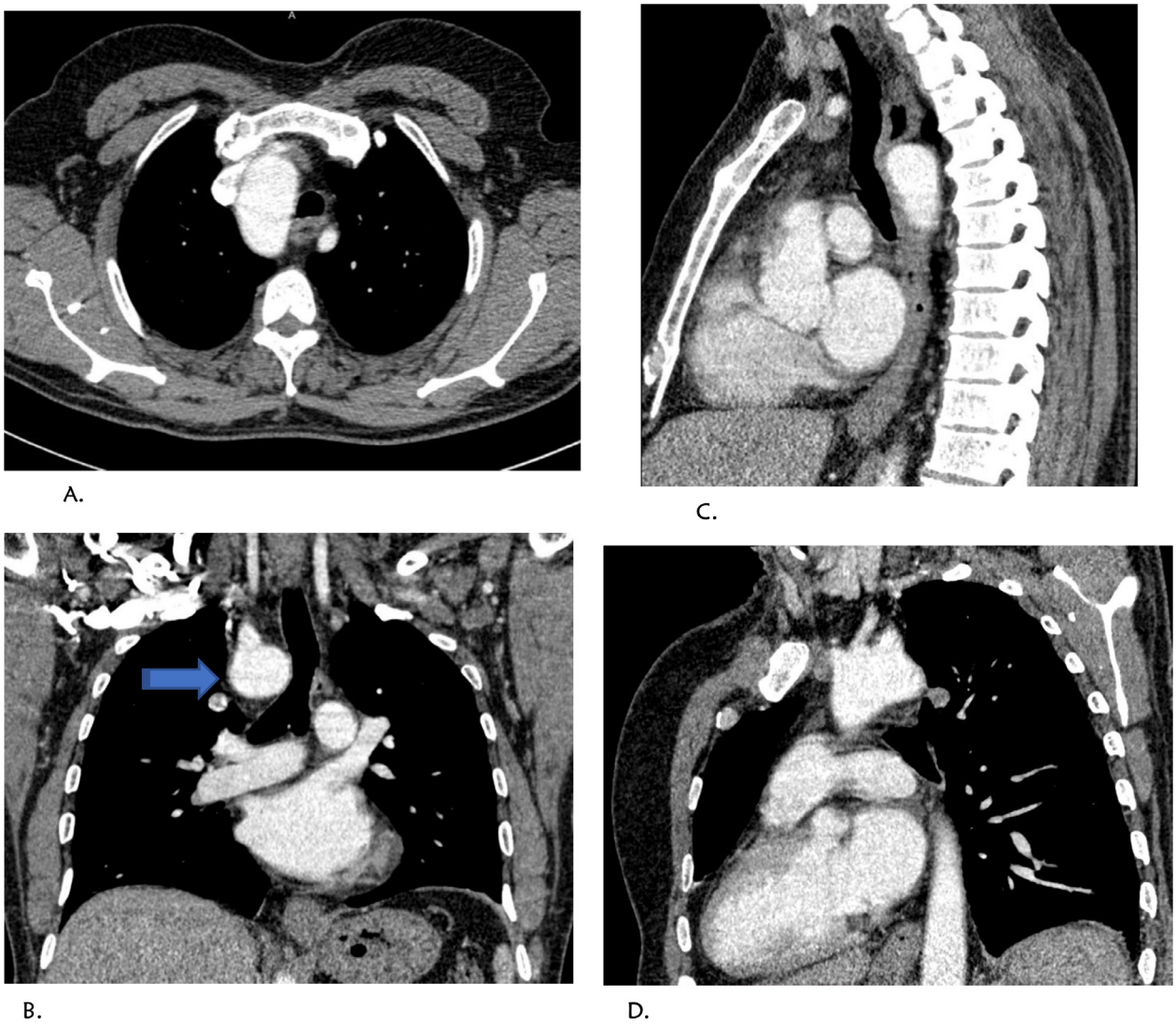

Fig. 2 – (A) Axial (B) Coronal and (C) Sagittal CECT images showing the ascending aorta arching over the right main bronchus between the superior vena cava on the right and the trachea on the left indenting the trachea from the right side and pushing it off the midline slightly to the left side. (D) sagittal image, shows the major branches arising, left common carotid artery (LCCA), right common carotid artery (RCCA), and the right subclavian artery (RSCA); in that order from proximal to distal.

indenting the trachea from the right side and pushing it off the midline slightly to the left side (Fig. 2). It gives of the left common carotid artery (LCCA), right common carotid artery (RCCA), and the right subclavian artery (RSCA); in that order from proximal to distal (Fig. 2D).

The left subclavian artery was seen arising near the distal segment of the aortic arch having a bulbous configuration at its origin. It then crosses the midline to the left side posterior to the trachea and esophagus indenting the esophagus and the trachea posteriorly (Fig. 3).

The conical dilatation of the aberrant left subclavian artery (ALSCA) at its origin, also known as "Kommerell's diverticulum" has antero-posterior diameter of 24.6 mm, as shown in Fig. 4. The thoracic aorta initially descends slightly to the right of the spinal column and the thoracic esophagus pushing the azygous arch slightly to the right. It then gradually moves to the left side before exiting through the diaphragmatic hiatus. The maximum diameter of the descending thoracic aorta adjacent to the diverticulum was 42.2 mm. The vertebral arteries were seen arising for the subclavian arteries on each side. Apart from a solitary thin-walled pulmonary cyst in the anterior segment of the left upper lobe with a diameter of 1.9 cm, the patient had no pulmonary findings.

Finally, the diagnosis of right-side aortic arch with aberrant left subclavian artery and Kommerell's diverticulum was made. Considering the risk of acute complications, the patient was advised to avoid strenuous exercise to reduce vascular strain on the diverticulum and was well informed about the possible symptoms of the condition. There was no immediate interventional procedure planned for him however for

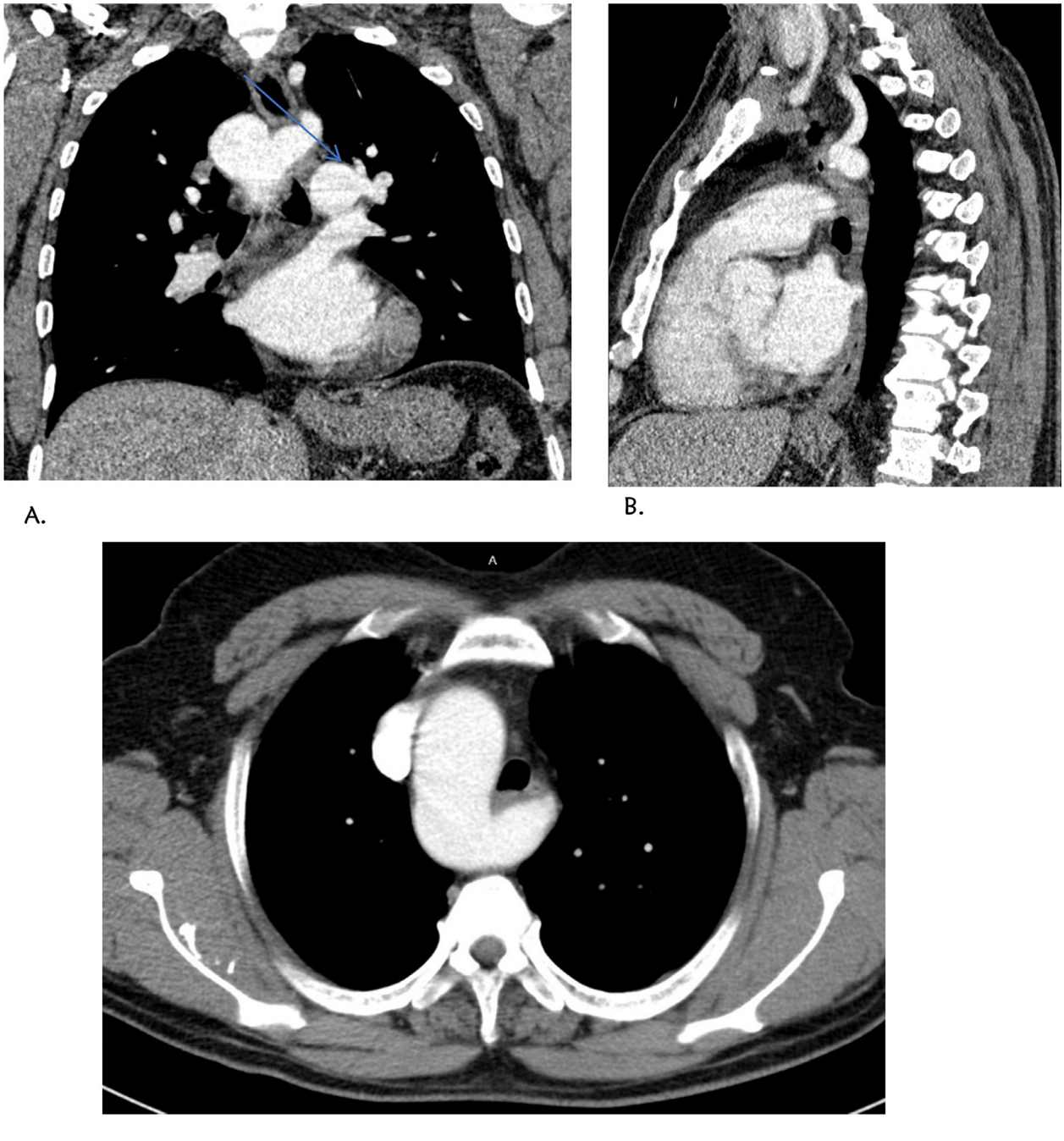

Fig. 3 – (A) Coronal (B) Sagittal and (C) Axial CECT images showing the aberrant left subclavian artery is arising near the distal segment of the arch having a bulbous configuration at its origin (KD).

possible early diagnosis of complications follow-up with serial CTA was planned after a year.

Discussion

Aberrant left subclavian artery arising from Kommerell's diverticulum in right sided aortic arch is quite a rare congenital vascular variant, and also underreported in the literatures [3]. Normal vascular anatomy of left sided aortic arch occurs when the left fourth embryologic aortic arch persists and the right involutes [5]. Right-sided aortic arch (RAA) is a type of aortic arch variant characterized by the aortic arch coursing to the right of the trachea. It was first documented by Fioratti and Aglietti in 1763 [1]. It is the result of an abnormal organogenesis of primitive aortic arches. Between the fourth and fifth weeks of embryonic life, blood leaves the heart by a single

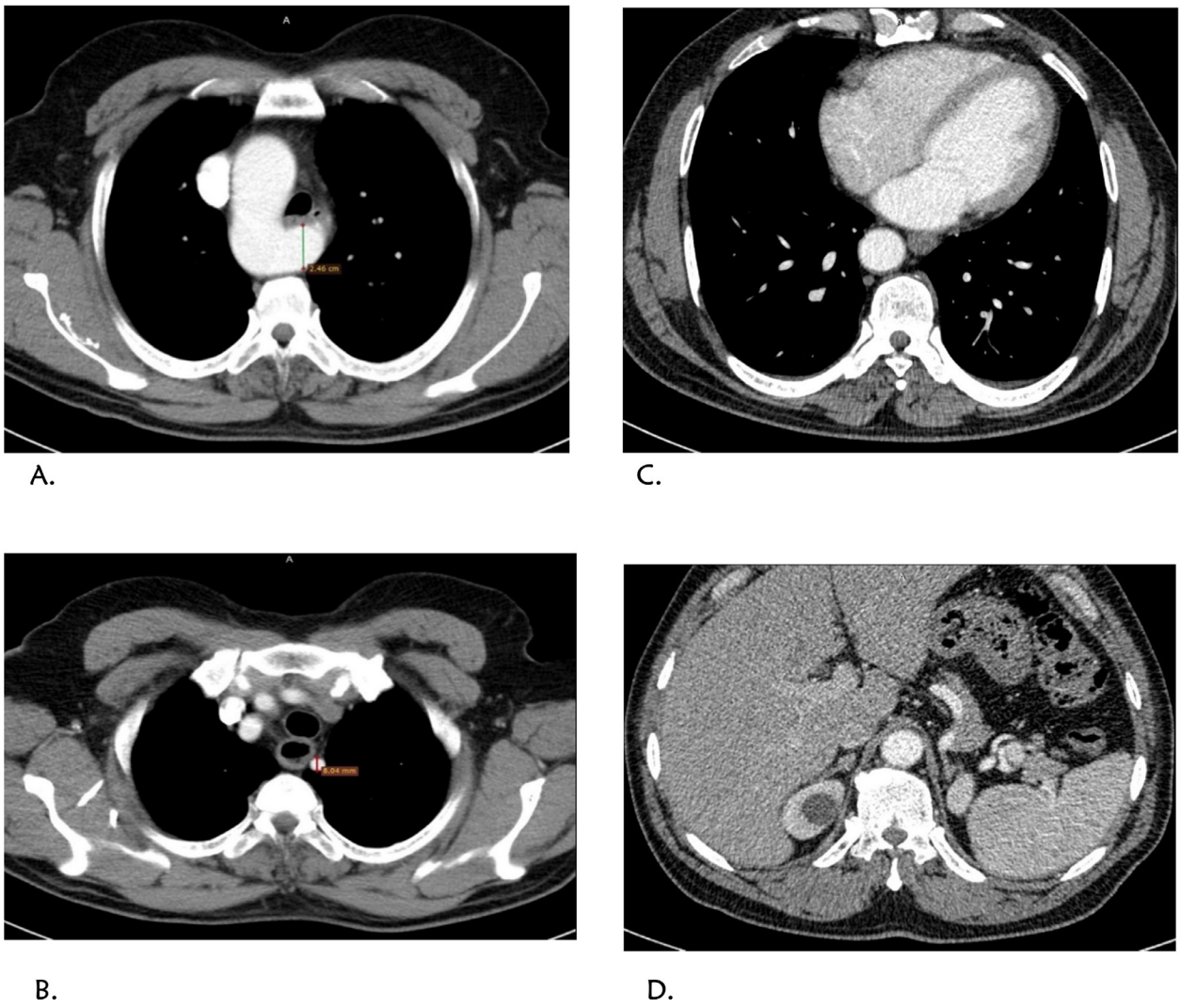

Fig. 4 – (A-D) Axial CECT images. the diameter of the Kommerell's diverticulum seen in (A). The major aortic branches, the left common carotid artery (LCCA), right common carotid artery (RCCA), the right subclavian artery (RSCA) and the aberrant left subclavian artery (ALSCA) are seen distal to their origin in (B). The descending thoracic aorta on the right side of the spinal column gradually moves to the left side as it exits through the diaphragmatic hiatus, an Incidental right renal cyst is also seen in (C) and (D).

vessel, the truncus arteriosus, which divides into 2 branches, the ventral aortae. These are connected with the paired dorsal aortae by 6 branchial vessels, called aortic arches, as shown in Fig. 5.

Segments of the first 3 arches, together with their dorsal and ventral aortic connections, form the carotid system. A segment of the right ventral aorta, the right fourth arch, and a portion of the right dorsal aorta develop into the right subclavian artery and the innominate artery. The left fourth arch persists as the adult aortic arch, and with the anlagen of the seventh dorsal intersegmental artery it forms the left subclavian artery. The fifth arches are both resorbed, and the sixth arches form the pulmonary artery and the truncus arteriosus (Fig. 6). The right-sided aortic arch results from persistence of the right fourth aortic arch and involution of the left.

Several classifications of these anomalies have been proposed [6–8]. It was classified in 1948 by Edward [9] into 3 main types of right-sided aortic arch: type I, with mirror-image branching of the major arteries; type II, with an aberrant subclavian artery with or without a Kommerell diverticulum; and type III, with isolation of the subclavian artery (where the left subclavian artery does not attach to the aorta but is connected to the pulmonary artery through the ductus arteriosus) as shown in Fig. 7A-C.

Type - 1 and 2 form 98% of the right-sided aortic arch cases [10]. A RAA with an ALSA [Type-2] is the most common type [11–13]. In this anomaly, the first branch arising from the

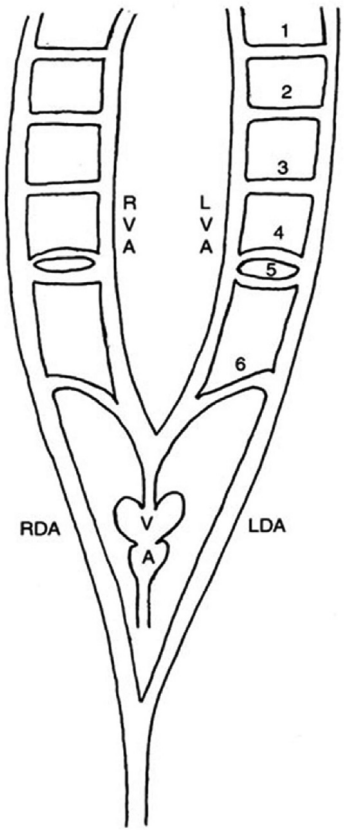

Fig. 5 – Schematic drawing shows circulation in 4-week human embryo. 1-6, Primitive aortic arches; RVA, right ventral aorta; LVA, left ventral aorta; RDA, right dorsal aorta; LDA, left dorsal aorta; V, ventricle; A, atrium. [Courtesy of 4].

aortic arch is the left carotid artery, which is followed by the right carotid artery, right subclavian arteries and ALSA in order (Fig. 7B). The descending aorta is usually on the right side or near the midline. An ALSA may arise from a remnant of the left dorsal aortic root (Kommerell's diverticulum), named after Burckhard F Kommerell, a German radiologist (1901-1990) who first described it in 1936 while studying a barium swallow and discovering a pulsatile mass posterior to the esophagus causing its compression [14]. This occurs as a result of the reabsorption of the left fourth aortic arch proximal to the origin of the left subclavian artery [15].

This anomaly rarely produces symptoms and is usually an incidental radiological finding, our case is diagnosed incidentally while he is on work up for shoulder and chest pain. Meanwhile symptoms like cough, shortness of breath, syncope and dysphagia associated with compression of the trachea or the esophagus or subclavian steal phenomenon are absent and evidence for aortic dissection and myocardial cause of chest pain were lacking. Symptoms of KD in adulthood are more often the result of early atherosclerotic changes of the anomalous vessel, dissection, or compression of surrounding structures causing dysphagia, dyspnea, stridor, wheezing, cough, choking spells, recurrent pneumonia, obstructive emphysema, or chest pain [16].

Rarely, a right arch with an ALSA forms a complete vascular ring, left pulmonary artery and left ductus arteriosus. The complications associated with KD are diverticular rupture (4%), aortic dissection (11%), aortic aneurysm rupture, or ASA aneurysm rupture [17]. Given the rarity of this condition, there are no general guidelines for the management of this condition. It is generally accepted to consider surgical intervention when the diameter of the diverticulum orifice exceeds over 30 mm, and/or the diameter of the descending aorta adjacent to the diverticulum exceeds over 50 mm [18].

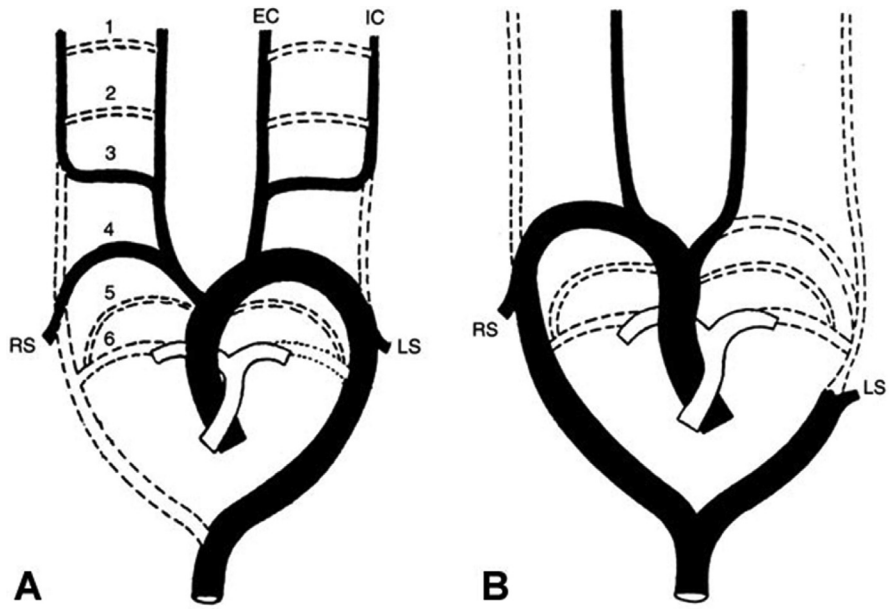

Fig. 6 – Development of left-sided aortic arch (A). Development of right-sided aortic arch with aberrant left subclavian artery (B). 1-6, Primitive aortic arches; RS, right subclavian artery; LS, left subclavian artery; EC, external carotid artery; IC, internal carotid artery; C, common carotid artery. [Courtesy of 4].

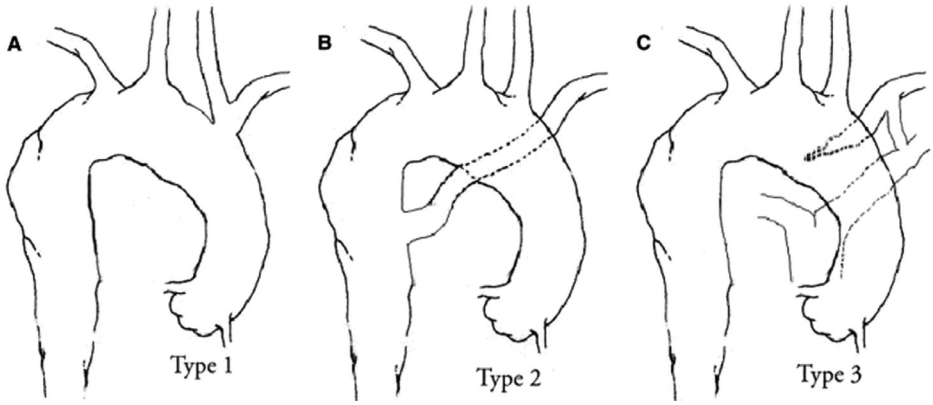

Fig. 7 – Classification of right-sided arch anomalies. (A) Type I, with mirror image arch branches. (B) Type II, with aberrant left subclavian artery. (C) Type III, with isolated left subclavian artery. [Courtesy of 6].

Conclusion

Right-sided aortic arch with aberrant left subclavian artery arising from Kommerell's Diverticulum is quite rare and may remain asymptomatic. On times it may cause symptoms in adulthood often as a result of early atherosclerotic changes of the anomalous vessels, dissection, or aneurysmal dilatation with compression of adjacent structures causing dysphagia, dyspnea, cough, or chest pain. Even though there are no general guidelines for the management of this condition patients need to be informed about the nature and possible outcomes of their condition. Close follow up of asymptomatic patients is one option of management until there are situations which require consideration of surgical intervention.

Patient consent

Patient written informed consent is obtained for publication from the patient.

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