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Original Research Article

Right Sided Aortic Arch with Aberrant Left Subclavian Artery with Aneurysmal Dilatation Presenting with Dysphagia: A Rare Combination of Aortic Arch Anomaly: A Case Report

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Abstract:

This case report details of a 54-year-old male patient experiencing difficulty swallowing with intermittent chest pain and breathlessness, ultimately leading to the discovery of a right-sided aortic arch anomaly. Initial chest radiography revealed telltale signs of an enlarged right mediastinum and a right-sided aortic knob, prompting further investigation. Contrast-enhanced computed tomography (CECT) and CT Angiography confirmed the presence of a right-sided aortic arch coupled with an aberrant left subclavian artery arising from Kommerell's diverticulum and aneurysmal dilatation. Notably, this anatomical variant resulted in extrinsic compression of the esophagus, contributing to the patient's dysphagia symptoms. Discussion encompasses the classification of right-sided aortic arch anomalies, their association with vascular rings, and potential clinical manifestations arising from adjacent structure compression. Additionally, the historical significance and clinical implications of Kommerell's diverticulum, its prevalence, and typical anatomical locations are explored in depth. This case underscores the importance of recognizing and understanding rare anatomical variations in clinical practice, particularly when presenting with atypical symptoms such as dysphagia.

Keywords: Right sided Aortic arch, Left aberrant subclavian artery, CT Angiography, Kommerell's diverticulum, Dysphagia lusoria.

Introduction

A right-sided aortic arch is a rare anatomical variant found in approximately 0.1% of adults.[1,2] Different configurations can be found based on the supra-aortic branching patterns. In about half of these cases, there's an accompanying anomaly known as an aberrant left subclavian artery, occurring at a rate between 0.05% to 0.1%.

A right-sided aortic arch paired with an aberrant left subclavian artery is less common compared to its counterpart, where a left-sided aortic arch is coupled with an aberrant right subclavian artery, with a prevalence ranging from 0.5% to 2.0%. [3,4] The presence of a right-sided aortic arch signifies the persistence of the right fourth aortic arch during development, while the left counterpart regresses. In some instances, an aberrant left subclavian artery may originate from a structure called Kommerell's diverticulum. There is a significant association with Digeorge syndrome. Often, this condition is discovered incidentally in adulthood and typically does not cause symptoms. However, when associated with the other vascular anomalies like vascular ring, they may cause compression of the trachea and/or esophagus.

Case Report: A 54-year-old male patient, previously in good health, underwent a chest radiograph due to his difficulty in swallowing with intermittent chest pain and breathlessness.

The radiograph revealed a right-sided aortic knob and an enlarged right mediastinum. Additionally, the thoracic aorta appeared tortuous on the right side of the spine, while the usual contour of the thoracic aorta on the left side was not visible. Based on these findings from the chest radiograph, a diagnosis of right-sided aortic arch was established. The Chest X-ray was followed up with a Barium Swallow. The lateral view of barium swallow study revealed a smooth extrinsic compression of the posterior wall of the esophagus at the level of aortic arch and right indentation of

the esophagus on frontal view. Contrast-enhanced computed tomography (CECT) of the thorax and CT Angiography of Aorta was performed to further assess the condition of the thoracic aorta. The imaging confirmed the presence of a right-sided aortic arch, where the thoracic aorta descended along the right side of the spine before looping to the left and entering the aortic hiatus at its usual position with aneurysmal dilatation. Additionally, an aberrant left subclavian artery was observed, originating from a diverticulum of the aortic arch known as Kommerell's diverticulum. The branches of the right-sided aortic arch, in order from proximal to distal, included the left common carotid artery, right common carotid artery, right subclavian artery, and left subclavian artery. Kommerell's diverticulum was located posterior to the trachea and esophagus, resulting in compression of the esophagus posteriorly and on the right side. However, there was no evidence of compression of the trachea. The final diagnosis of right-sided aortic arch with aberrant left subclavian artery associated with Kommerell diverticulum resulting in dysphagia lusoria was formulated.

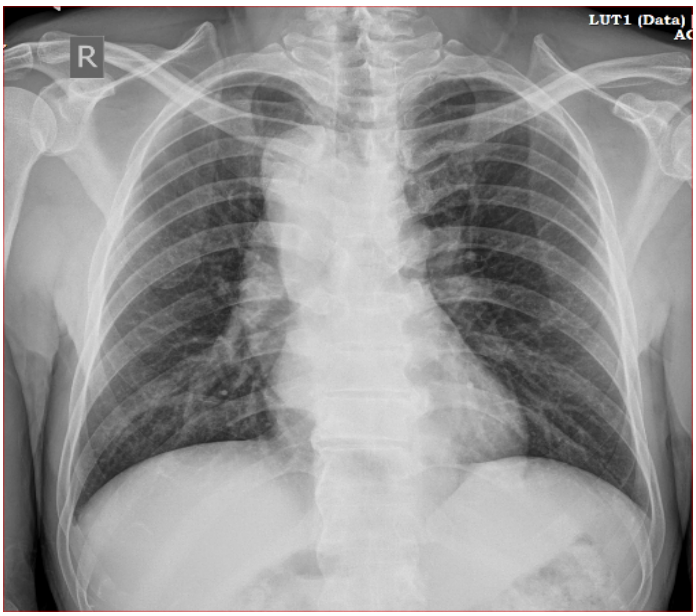

Figure 1: Chest X-ray PA view shows, Right-sided aortic knob and an enlarged right mediastinum

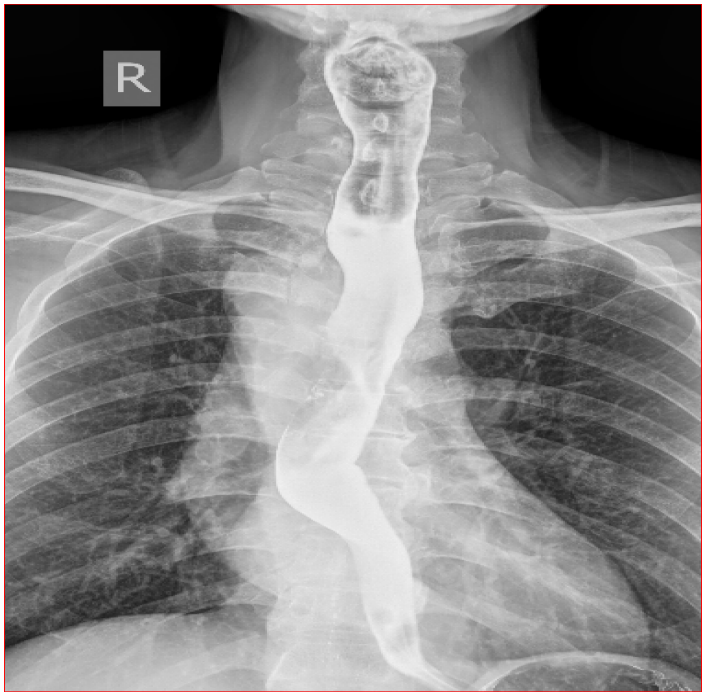

Figure 2: Barium swallow frontal view shows, Right indentation of the esophagus at the level of aortic arch

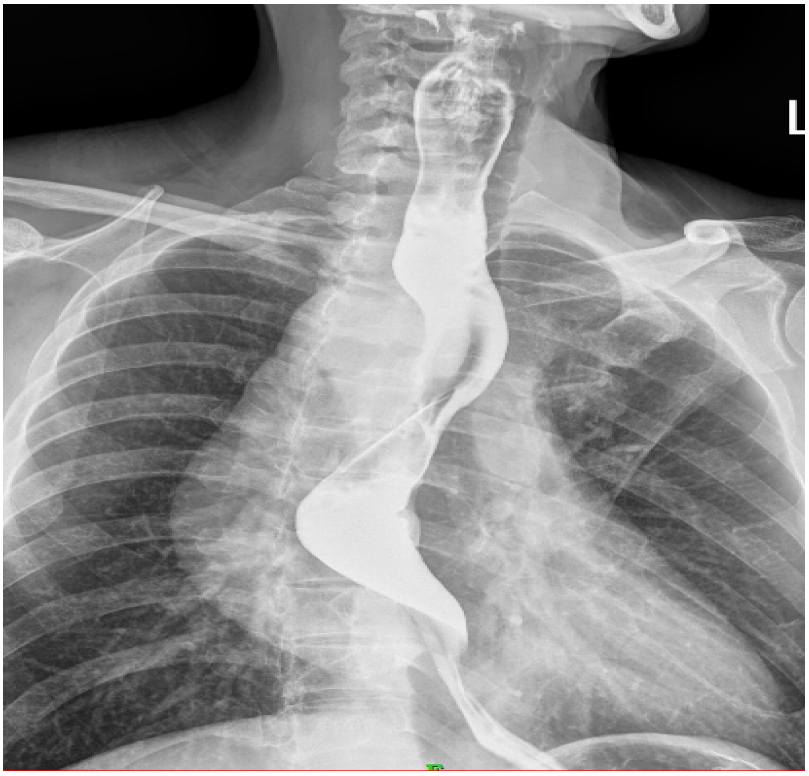

Figure 3: Barium swallow LAO view shows, a smooth extrinsic compression of the posterior wall of the esophagus at the level of aortic arch

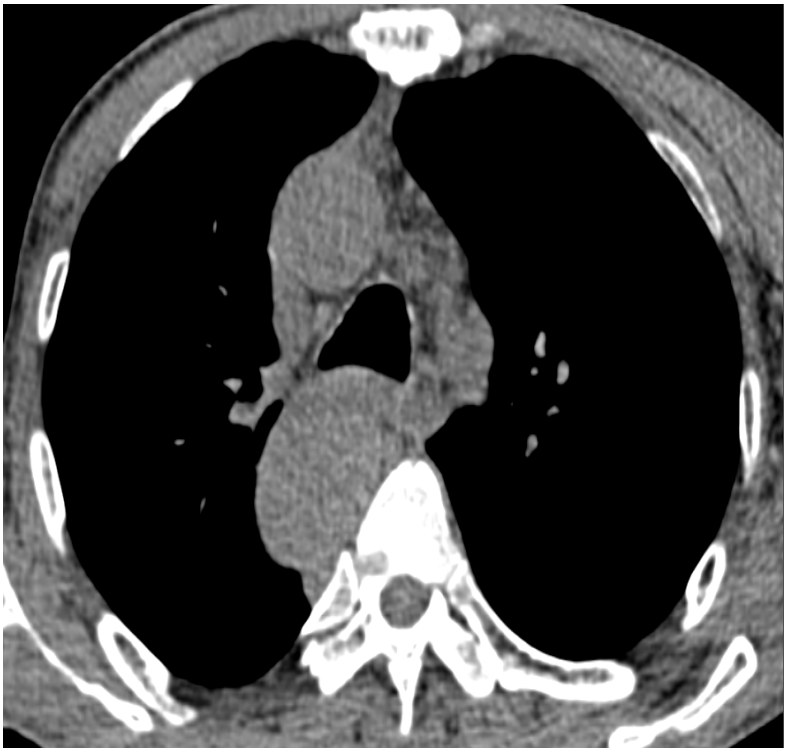

Figure 4: Axial CT Thorax shows, right-sided aortic arch and descending thoracic aorta with aneurysmal dilatation

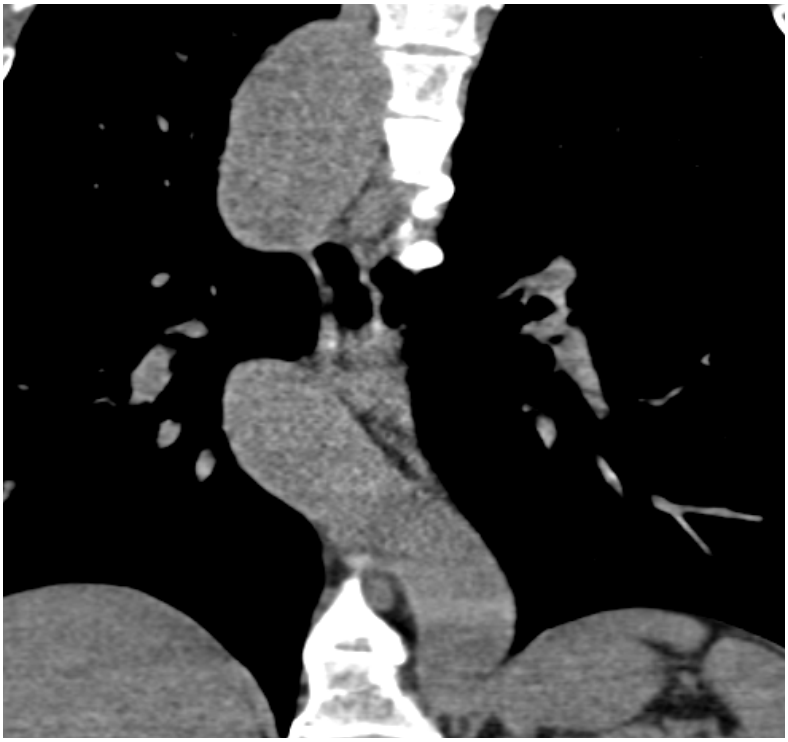

Figure 5: Coronal CT Thorax shows, right-sided aortic arch, where the thoracic aorta descended along the right side of the spine before looping to the left and entering the aortic hiatus at its usual position with aneurysmal dilatation

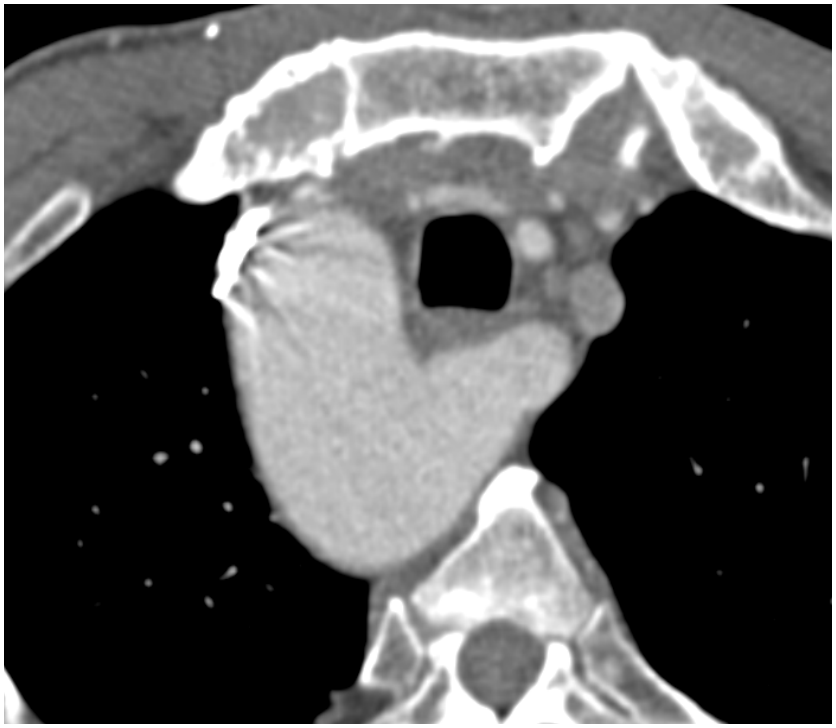

Figure 6: Axial CT Angiography of aorta shows, right sided aortic arch with aneurysmal dilatation

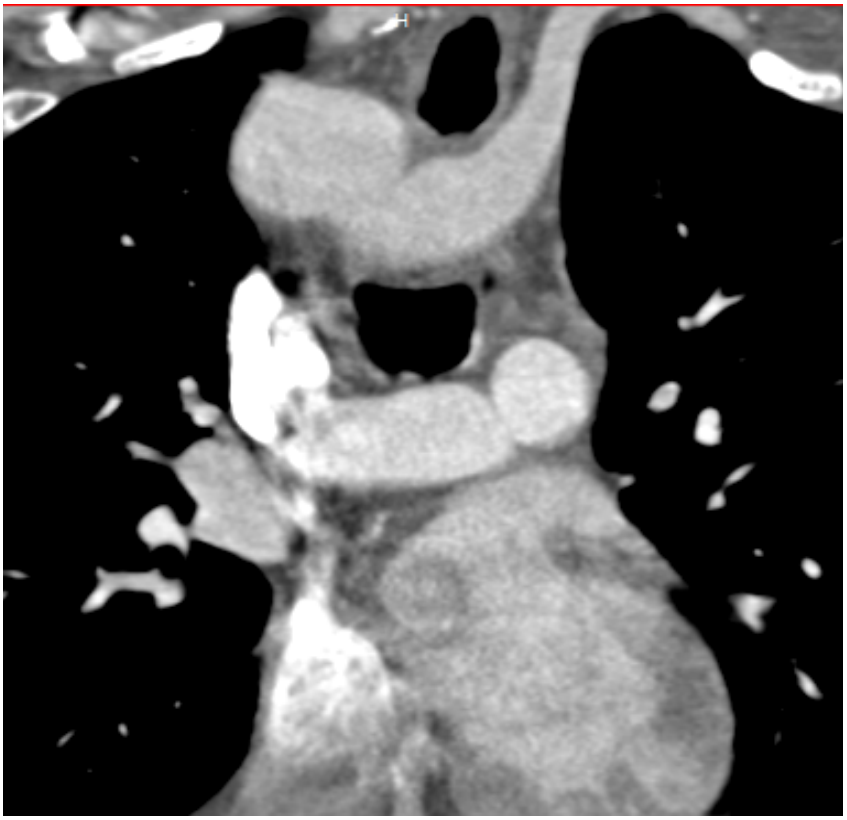

Figure 7: Coronal CT Angiography of aorta shows, abberant left subclavian artery with kommerell diverticulum

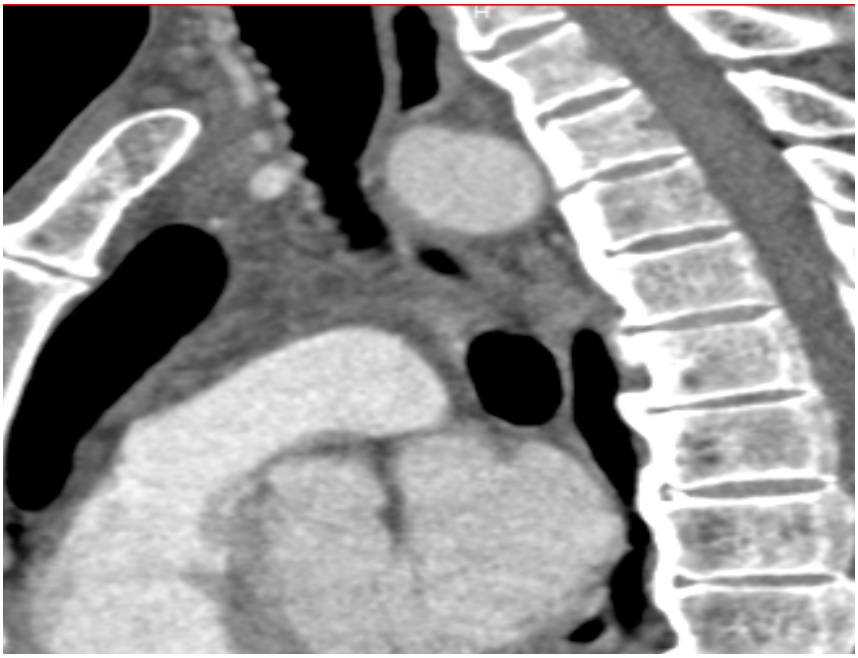

Figure 8: Sagittal CT Angiography of aorta shows, posterior esophageal compression by abberant left subclavian artery

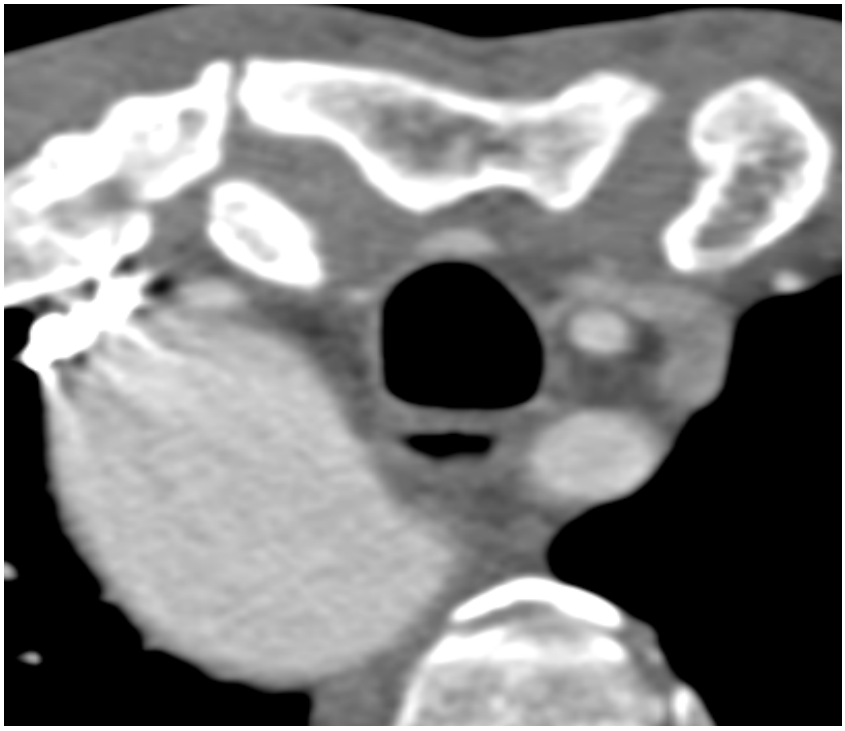

Figure 9: Axial CT Angiography of aorta shows, posterior esophageal compression by abberant left subclavian artery

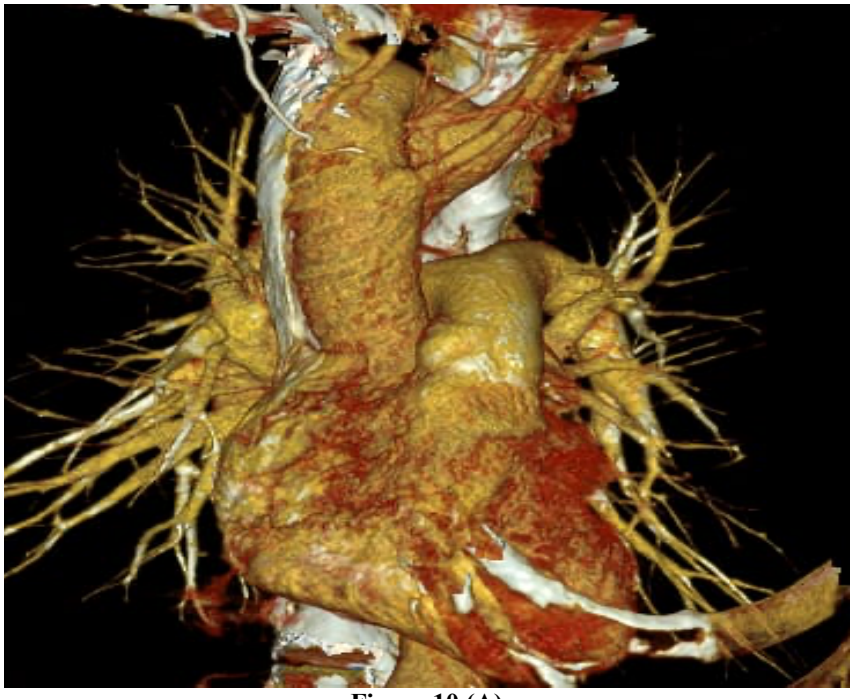

Figure 10 (A):

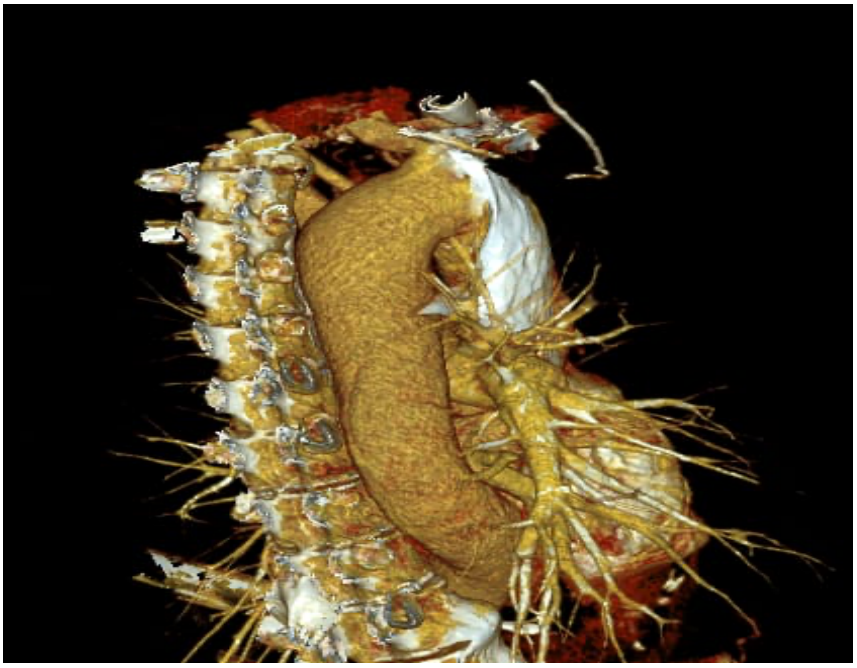

Figure 10 (B):

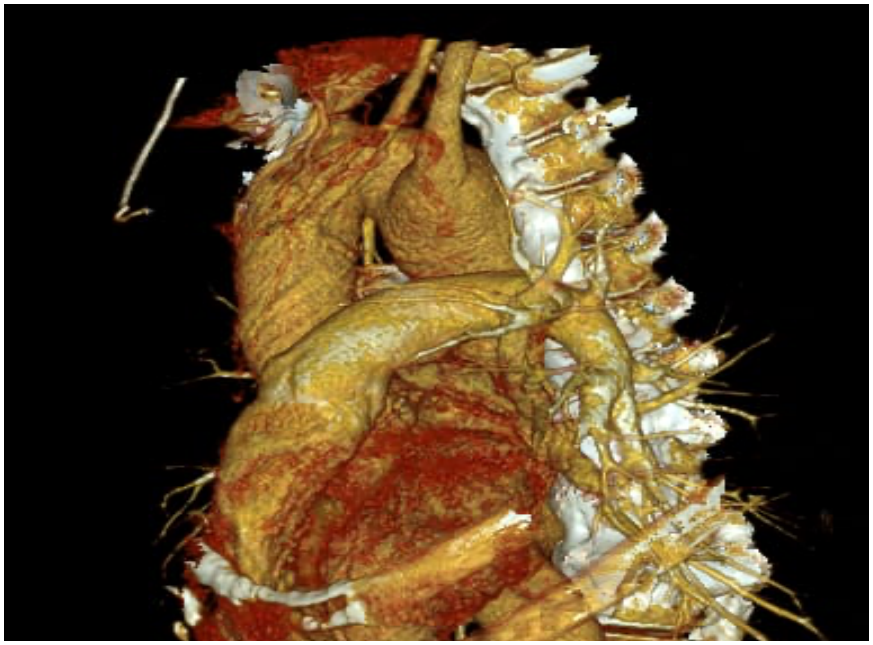

Figure 10 (c):

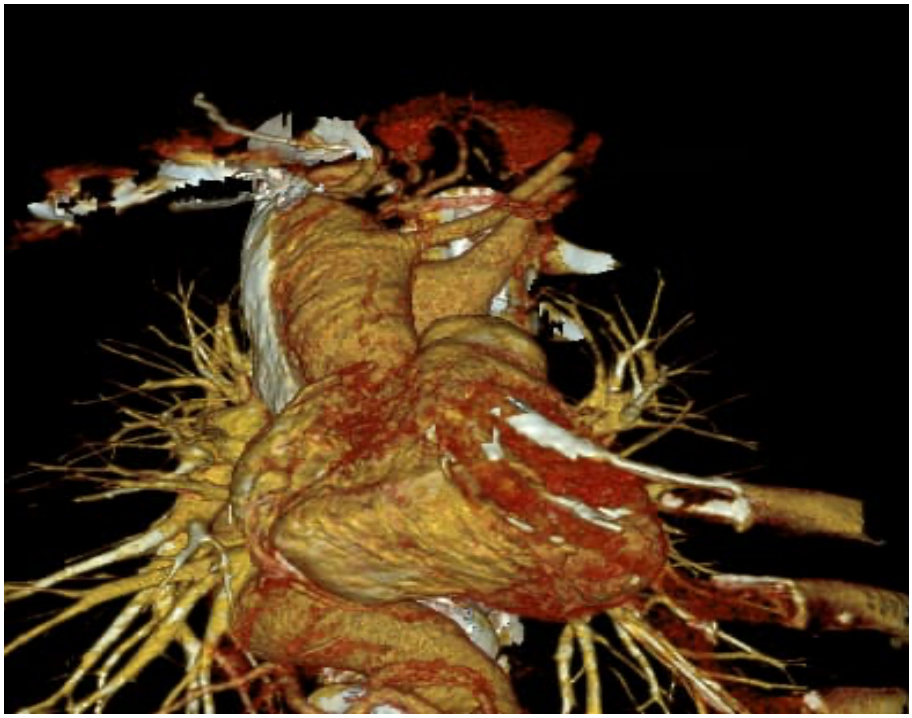

Figure 10 (D):

Fig 10: A, B, C, D 3D CT Angiography of aorta shows, right sided aortic arch with aneurysmal dilatation & abberant left subclavian artery from kommerell diverticulum.

Discussion

The first documentation of right-sided aortic arch dates back to 1763 by Fioratti and Aglietti.[5] Since then, several classifications have been proposed by Edward in 1948, Felson and Palayew in 1963, and Steward et al. in 1964.[6] The classification system categorizes right-sided aortic arch into three types.

Type 1 features major arteries branching from the arch in the following sequence: left innominate artery, followed by the right common carotid artery, and right subclavian artery. These branches mirror the typical arrangement seen in a left-sided aortic arch.[2,7]

In Type 2 of the classification, the right-sided aortic arch is accompanied by an aberrant left subclavian artery, which aligns with the presentation in this case. Conversely, Type 3 involves an isolated left subclavian artery that doesn't connect to the aorta; instead, it links to the pulmonary artery via the ductus arteriosus.

Type 1 and Type 2 collectively make up about 98% of cases of right-sided aortic arch, while Type 3 is exceedingly rare.[8]

Literature documents approximately 50 instances of right-sided aortic arch with an aberrant left subclavian artery.[3] This anomaly originates from the persistence of the right fourth aortic arch during development, alongside the regression of the embryonic left fourth arch situated between the left common carotid artery and the left subclavian artery.[9,10] Consistent with this anomaly, as observed in our case, the initial branch emerging from the arch is the left common carotid artery, succeeded by the right common carotid artery, the right subclavian artery, and finally, the left subclavian artery.[7,11]

According to literature findings, a right aortic arch with an aberrant left subclavian artery typically accompanies a left ductus arteriosus.[8,14] This configuration forms a vascular ring, where the left ductus arteriosus connects the left pulmonary artery to the base of the aberrant left subclavian artery. This vascular ring can lead to compression of the trachea. However, the absence of tracheal compression or deformity may indicate the absence of a ring or suggest that the ring does not necessitate treatment.[15]

This anomaly ranks among the three most common causes of vascular rings. However, in most cases, the vascular ring is loosely structured and does not induce compression. Patients with a right-sided aortic arch and an aberrant subclavian artery typically do not display symptoms, and there isn't a significant association with cardiac anomalies. Any symptoms that do arise are often related to the presence of the vascular ring. However, more commonly, symptoms stem from atherosclerotic changes in the anomalous vessels, dissection, or aneurysm formation. These conditions can lead to compression of nearby structures, resulting in symptoms such as dysphagia (known as dysphagia

lusoria) and dyspnea.[16,17] In the case described, the patient had symptoms of dysphagia.

The aberrant left subclavian artery typically originates from a structure called Kommerell's diverticulum. This diverticulum is characterized as a conical dilation of the proximal portion of the aberrant subclavian artery near its point of origin from the aorta. [4,11] It's also referred to as "lusoria diverticulum," "remnant diverticulum."[16] Initially it was describe by Burckhard Friedrich Kommerell in 1936, [18] it was observed in a case involving an aberrant right subclavian artery (a remnant of the right fourth aortic arch) associated with a left aortic arch. Despite its historical association with the right subclavian artery, it's commonly found in cases of aberrant left subclavian artery, representing the remnant of the left fourth aortic arch. Kommerell's diverticulum is typically located behind the esophagus in 80% of cases, between the trachea and esophagus in 15%, and behind the trachea in 5%.

Conclusion:

Right-sided aortic arch is a very rare anatomical variant present in 0.1% of adult population. It may be associated with an aberrant left subclavian artery, and half of the cases causing dyspnea or dysphagia [19].

The embryologic origin of aorta start from aortic arches which develop from the aortic sac and proceed to course into the pharyngeal arches. The aortic arches or the pharyngeal arch arteries or branchial arches develop from the aortic sac, with a pair of branches (right and left) travelling within each pharyngeal arch and ending in the dorsal aorta. The right aortic arch develops due to the persistence of the right dorsal aorta and disappearance of the left dorsal aorta.

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