Journal of Cardiology and Cardiovascular Research

Ohene BE, et al., 2021- J Cardiol Cardiovasc Res
Case Study

Diverticulum of Kommerell with Micro Aneurysm, an Aberrant Right Subclavian Artery and Ventricular Septal Defect in a 39 years Old Female: case report

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Accepted Date: 08-27-2021
Published Date: 09-27-2021

Abstract

Diverticulum of Kommerell is a congenital anomaly in the development of the primitive embryonic aortic vasculature resulting in either an aberrant left subclavian artery (ALSA) from a right-sided aortic arch (RSAA) or an aberrant right subclavian artery (ARSA) from a left-sided aortic arch (LSAA). Kommerell's diverticulum (KD), is extremely rare in the general population having prevalence of 0.7–2.0 %. KD with true aneurysmal dilatation like any vascular lesion of the thoracic aorta weakens the vasculature and increases the risk of aortic rupture. Timely surgical intervention is of paramount importance in most of these cases. Variable clinical presentations in patients with KD are either a result of compression of some mediastinal organs or strangulation by the vascular ring from the aberrant subclavian artery (ASA). We report a case of a 39 years old Chinese female with a rare co-existence of a huge KD with a micro aneurysm, an aberrant right subclavian artery (ARSA) and ventricular septal defect (VSD). She underwent a successful operation and all her pre-surgical symptoms disappeared.

Keywords: Diverticulum of Kommerell, Aberrant Right Subclavian Artery (ARSA); micro-aneurysm; Ventricular Septal Defect (VSD); coexistence.

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DOI: https://doi.org/10.37191/Mapsci-JCCR-2(3)-038

Introduction

Approximately 80 years ago, it was described as an aortic diverticulum from which an aberrant right subclavian artery originated later became known as KD and represents an aneurysmal dilatation occurring at the proximal descending aorta (of both left and right arch configuration) where an aberrant subclavian artery (ASCA) takeoff [1,2]. KD is a congenital anomaly in the development of the primitive embryonic aortic vasculature resulting in either an aberrant left subclavian artery (ALSA) from a right-sided aortic arch (RSAA) or an aberrant right subclavian artery (ARSA) from a left-sided aortic arch (LSAA) [3].

KD is uncommon in the general population, its prevalence is about 0.7–2.0% in individuals with left-sided aortic arch with aberrant right subclavian artery and 0.04–0.4% for right-sided aortic arch with aberrant left subclavian artery [2,4,5]. This condition could result in compression from aneurysmal dilatation and strangulation by vascular ring formed by the ASA. Clinical presentations which include dysphagia, mild intermittent asthma-like dyspnea, and hoarseness may be the results of esophagus, trachea, recurrent laryngeal nerve compression or strangulation and chest discomfort and angina-like exercise intolerance of an unknown origin [6].

In some cases, the takeoff of the ASAs is very small in luminal diameter resulting in decrease perfusion, blood pressure difference in upper extremity supplied by the ASA [7]. Like any other aneurysm, KD with frank ballooning of the aorta, increases the risk of aortic rupture and for that matter may require a drastic surgical intervention. This case is worth reporting because in light with our thorough review of literature the coexistence of KD with an aberrant right subclavian artery from left sided aortic arch and ventricular septal defect is yet to be described.

Case Report

This is a case of a 39 years old female patient, who presented to the cardiovascular surgery Out-patient clinic with 5 years history of syncope, intermittent chest pain, and dyspnea on exertion, mild dysphagia for solids accompanied with weight loss of unclear origin. Pulse and BP were slightly different in the upper extremities (right arm lower that the left arm) with intermittent weakness of the right arm. Her lungs were clear on auscultation bilaterally with normal vesicular breath sounds, her EKG was unremarkable. Other physical examinations were non-specific. Contrast enhanced Computer tomography of the aorta demonstrated a huge (>55mm in diameter) saccular aneurysm of the proximal descending thoracic aorta. The 3-D reconstructed image is shown in Figure 1 and 2. Computed tomography of the heart revealed a small ventricular septal defect (VSD) less than 4mm) label “D” in Figure 3.

Transthoracic echocardiography (TTE) studies were done to ascertain the hemodynamic significance of the defects, that is the degree of left to right shunting of blood across the 3-4 mm defect. Anesthesia was done under left-sided double lumen endotracheal intubation [8]. The aneurysm and the proximal descending thoracic aorta

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were exposed through a left lateral thoracotomy at the fourth intercostal space, as this was recommended to provide a better view of aneurysms at this anatomic location [9]. Partial cardiopulmonary bypass was established via femoral artery and femoral vein cannulation with a mild hypothermia (33.6 degrees Celsius). Size 16mm artificial Hemashield vascular graft was anastomosed (full thickness suture). Thus, partial arch replacement and reconstruction of the right aberrant subclavian artery (ARSA) were accomplished. The VSD was not repaired as it is less than 4mm and was not hemodynamically insignificant. The patient made a remarkable recovery and was discharged 9 days after the operation. Figure 4 and 5 represent a 3-D CT reconstruction of her aorta after surgery.

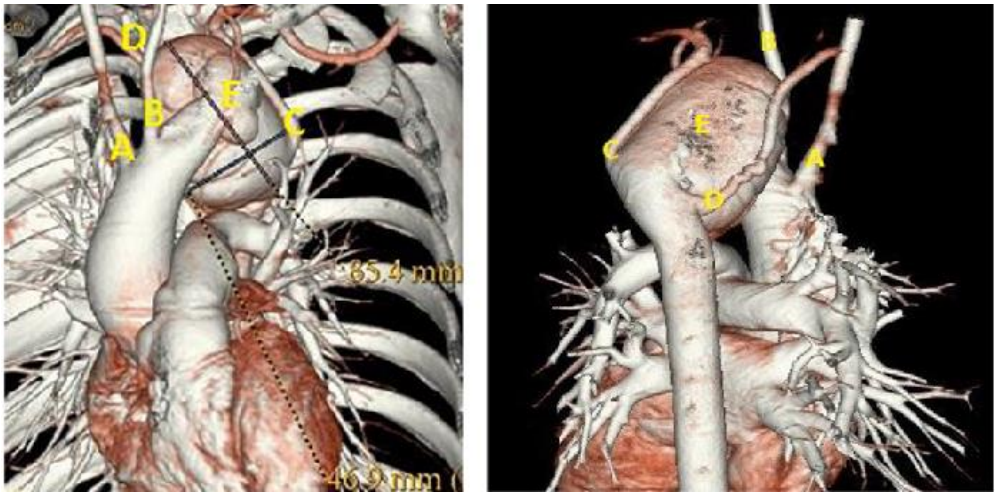

Figure 1 and 2: 3-D reconstructed CT showing antero-posterior and posterior-anterior views respectively, a huge saccular aneurysm of the proximal descending thoracic aorta (KD with ARSA from a left-sided aortic arch.), A- right common carotid artery; B-left common carotid artery; C-left subclavian artery; D- the aberrant right subclavian artery; E-micro-aneurysm on the KD.

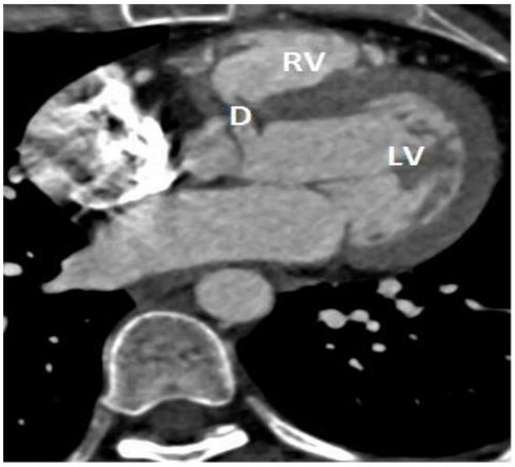

Figure 3: CT of the heart. RV: right ventricle; LV-left ventricle; D-ventricular septal defect (VSD); AA – Ascending aorta; DD-descending.

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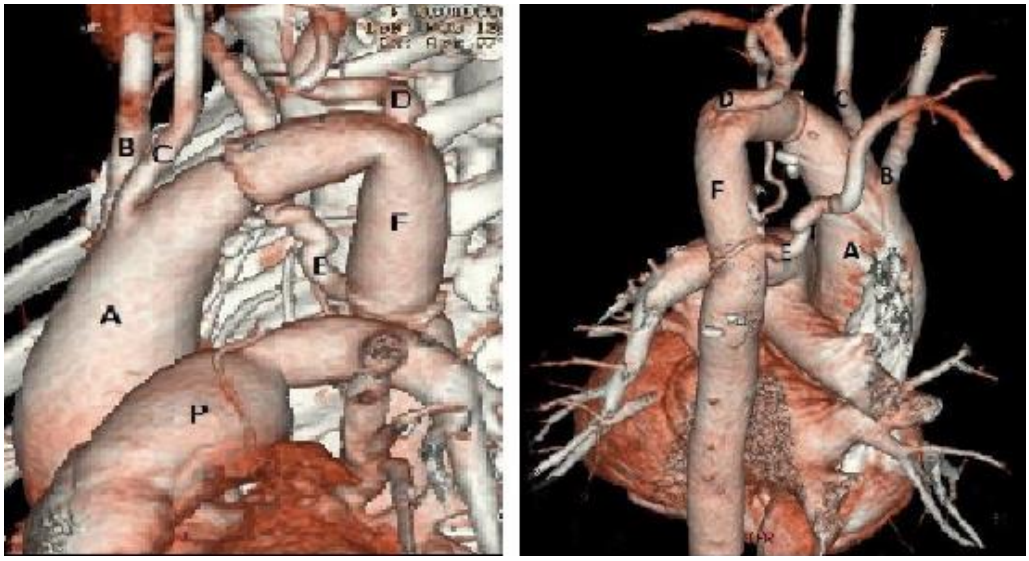

Figure 4 and 5: Antero-posterior and posterior-anterior views of 3 D reconstruction of her computed tomography of the new aorta. 90 days after surgery. A- ascending aorta; B-right common carotid artery; C-left common carotid artery; D- left subclavian artery; E- aberrant right subclavian artery; F- the reconstructed vascular graft; P- pulmonary artery.

Discussion

Worldwide the average life expectancy has gained an increase however the all-cause mortality due to aortic aneurysm has risen in the last 2 decades [10]. The incidence of thoracic aneurysm in the general population is about 10/100000 and over 50 percent of the cases of thoracic aneurysm encountered in clinical practice were asymptomatic [11]. Surgical indication for aneurysm in the region of the thoracic aorta is recommended at 5.5cm and 6.5cm for ascending and descending aorta respectively and for symptomatic aneurysms were to be extirpated regardless of size [12]. Limited data available suggests that survival with thoracic aorta aneurysm is at best equivalent to that observed for abdominal aortic aneurysm [10] not to mention KD having a higher risk and rate of rupture [13]. Aneurysm of the thoracic aorta grows at an average rate of 0.10cm per year (0.07 for ascending aorta and 0.19 for descending aorta). Critical sizes for natural complications of aortic aneurysm (rupture or dissection) were found at 6.0cm for the ascending aorta and 7.0cm for the descending aorta with the risk of rupture or dissection of ascending and thoracic aorta was reported to be 31% and 43% at specific dimensions [12]. These were outcome of pooled statistical data and may not apply for symptomatic KD, therefore surgical intervention in these types of aneurysms could be considered at lower diameters. In the studies of KD, it was suggested that surgical intervention could be taken at the diameter of 3cm at the level of the orifice of the diverticulum as the threshold for surgery in low-risk patient [14]. A recommendation of surgery represents a balance, weighing estimates of natural history of thoracic aorta aneurysm and rupture risk against operative mortality not forgetting that symptoms presented by the patient is a very

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critical factor. This risk is related to the site, etiology, size, and expansion rate of the aneurysm [15].

Surgical interventions in asymptomatic and symptomatic patients with aneurysm and for that matter this patient was to provide a prophylactic surgical treatment against rupture of the aneurysm, relieve compression of high mediastinal structures responsible for clinical symptoms presented by patients (mild dysphagia for solids in our patient), keep cerebral perfusion and blood flow to the upper extremities (syncope and transient ischemic attacks, intermittent non palpable pulse in the right arm, blood pressure slightly lower in the right arm) steady, and ensure physiologic laminal flow through the aorta.

All the symptoms presented by this 39-year-old female which included personality change and inexplicable forgetfulness a reason which necessitated a medical consultation "faded out" within three months after her operation. We therefore wish to ascribe "Kommerell's diverticula Arch aneurysm steal syndrome" to the symptoms presented by this patient as there is no other clear explanation for her phenomenon.

Conclusion

Patients with congenital anomalies of the aortic arch may have other co-existing vascular and intra cardiac defects with diverse clinical presentations. Aberrant subclavian arteries and Kommerell's diverticulum can be repaired safely under elective conditions with mild hypothermia (33.6 ^{o}C). A left thoracotomy at the 4th ICS provides excellent exposure of aneurysms of the proximal descending thoracic aorta.

Competing interests

None declared

Acknowledgement

We wish to express our sincere gratitude to the entire theatre and the cardiac intensive care unit nursing staff of the 1st affiliated hospital, Dalian medical university for their immense support during the perioperative period.

Funding

This work was supported by grants from the Science and Technology Planning Project of Liaoning province, China (2015020303).

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