Asploro Journal of Biomedical and Clinical Case Reports
(ISSN: 2582-0370)
Case Report
DOI: https://doi.org/10.36502/2023/ASJBCCR.6285

A Rare, Relapsing Kommerell Diverticulum – A Case Report

Alejandro Bautista-Pérez-Gavilán[1,2], Denya Sánchez-Quintero[2], Ana Gilabert-García[2], José Emiliano Ríos-Méndez[2], Santiago Saenz-Ancira[1], Carlos Alberto Villa-Ramirez[1,2], Andrea Ibarra-Moreno[1], Mariell Rodríguez-Salazar[1], Sofía Molina Castillo[1], Jorge Luis Bermudez-Gonzalez[1,2], Nilda Espinola-Zavaleta[1,3], Leonardo Proaño Bernal[1], Erick Alexanderson-Rosas[1,2iD*]

[1]*Department of Nuclear Cardiology, Instituto Nacional de Cardiología Ignacio Chávez, Mexico City, Mexico*
[2]*School of Medicine, Universidad Nacional Autónoma de México, Mexico City, Mexico*
[3]*Department of Echocardiography, Centro Médico ABC, I.A.P., Mexico City, Mexico*

Corresponding Author: **Erick Alexanderson-Rosas** ORCID iD
Address: National Institute of Cardiology Ignacio Chávez, Juan Badiano N°1, Colonia Sección XVI, Tlalpan, P.C. 14080, Mexico City, Mexico; Email: ealexanderson.edu@gmail.com
Received date: 19 January 2023; **Accepted date**: 06 February 2023; **Published date**: 13 February 2023

Citation: Bautista-Pérez-Gavilán A, Sánchez-Quintero D, Gilabert-García A, Ríos-Méndez JE, Saenz-Ancira S, Villa-Ramirez CA, Ibarra-Moreno A, Rodríguez-Salazar M, Castillo SM, Bermudez-Gonzalez JL, Espinola-Zavaleta N, Bernal LP, Alexanderson-Rosas E. A Rare, Relapsing Kommerell Diverticulum – A Case Report. Asp Biomed Clin Case Rep. 2023 Feb 13;6(1):23-29.

Abstract

Kommerell diverticulums are an uncommon congenital vascular disease involving an aberrant origin of the right or left subclavian artery and a dilation of its root. Here, we present the case of a 44-year-old woman with a relapse of a surgically repaired aberrant subclavian artery with a Kommerell diverticulum.

Keywords

Kommerell Diverticulum, Congenital Vascular Disease, Aortic Regurgitation, Vascular Surgery, Aortitis

Past Medical History

The patient had a history of hypothyroidism, asthma, migraine and poorly controlled hypertension. She had been admitted to the emergency room two years prior due to piercing precordial pain with an intensity of 10/10 AVS that irradiated all the way to the interscapular region. An aortic regurgitation murmur II/IV in intensity was heard. An angiotomography was performed, revealing an aberrant right subclavian artery with a KD along with signs of imminent rupture. The defects were surgically repaired and the patient remained asymptomatic during the following eighteen months.

History of Presentation

The patient was admitted to the emergency department due to acute onset intense chest pain. She had a history of a surgically-corrected aberrant right subclavian artery associated with a Kommerell diverticulum (KD). There were no complications during or after initial intervention, and she had been asymptomatic for eighteen months until that day. She was admitted to our center's emergency department for further evaluation.

Differential Diagnosis

Due to the patient's clinical manifestations and past medical history, there was a strong suspicion of a

Case Report

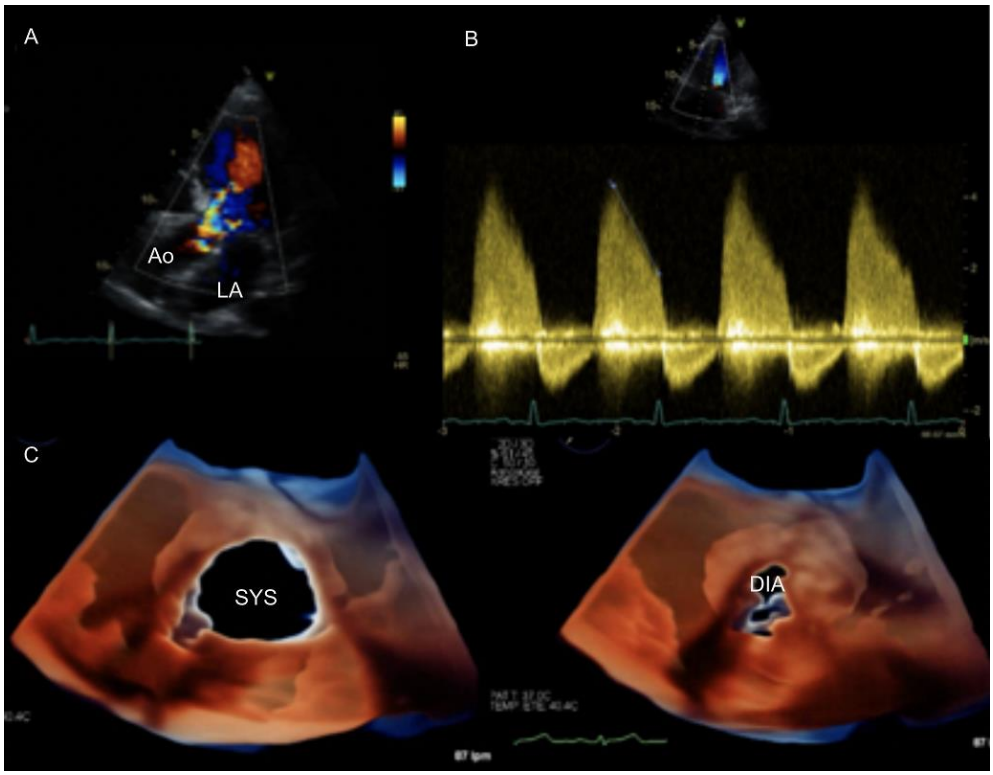

Fig-1: 2D TTE with continuous Doppler and 3D TEE with color

A: *Apical 5-chamber view showing severe aortic regurgitation with Doppler color.* ***B:*** *Continuous Doppler showing severe Aortic Insufficiency with pressure half time <200 ms.* ***C:*** *3D TEE with transillumination where the incomplete closure of the aortic valve is shown, confirming the diagnosis of aortic regurgitation. Ao: Aorta; DIA: Diastole; LA: Left Atrium; SYS: Systole.*

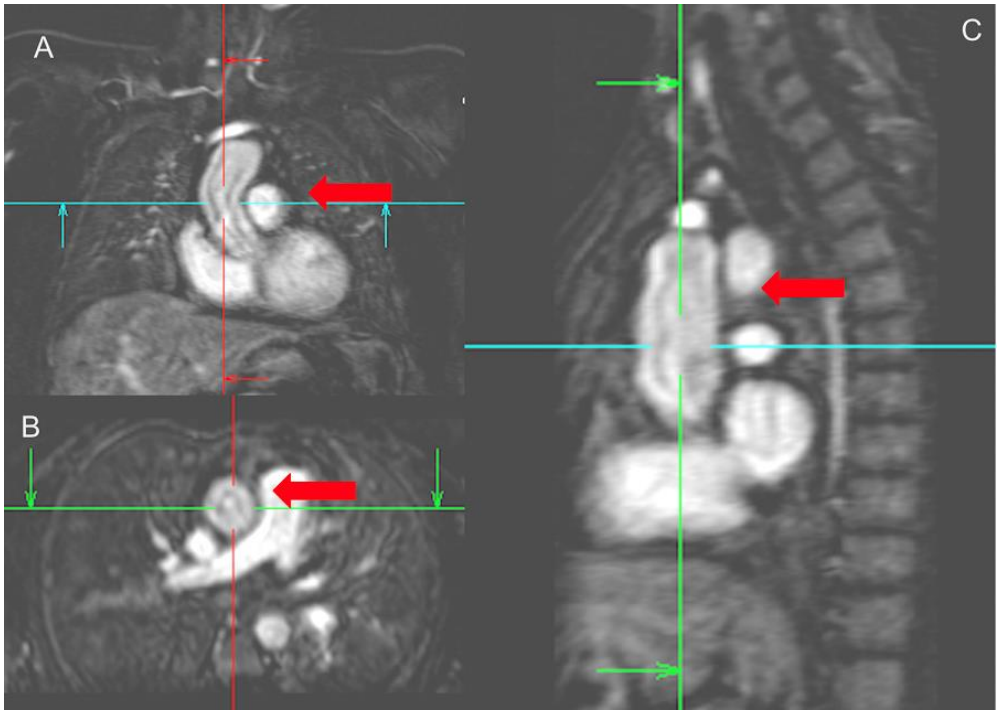

Fig-2: Cardiac MRI study where the recurrent Kommerel's diverticulum was first seen

A: *Cardiac MRI study showing Gadolinium enhancement in the Ascending Aorta* ***B:*** *Axial slice.* ***C:*** *Sagittal slice. Note the correlation between the increased uptake of Gadolinium in the study shown above and the increased uptake of FDG-18 in the subsequent PET study (See:* ***Fig-3****).*

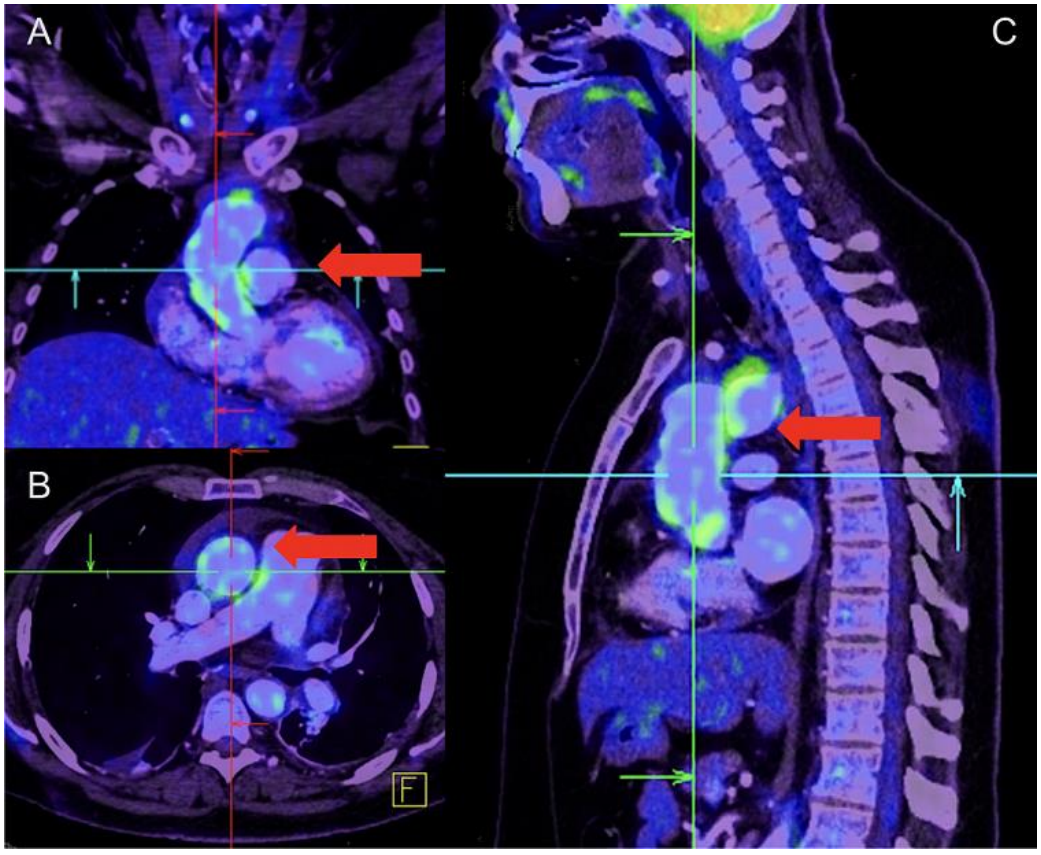

Fig-3: PET Study with 18-FDG

This study shows an abnormally high metabolic activity at the ascending aortic wall, thus confirming the diagnosis of aortitis and a recurrent KD. ***A:*** *Coronal slice with diffuse uptake in the aorta.* ***B:*** *Axial slice.* ***C:*** *Sagittal slice. Note the correlation between the increased uptake of Gadolinium in the Cardiac MRI (See:* ***Fig-2****) and the increased uptake of FDG-18 in the subsequent PET study shown above.*

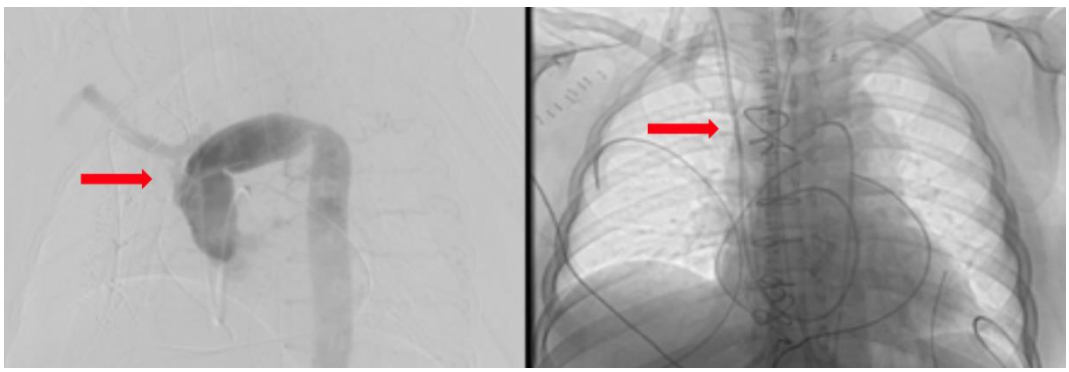

Fig-4: Aortogram

Demonstration of the recurrent KD in the proximal segment of the aberrant Right Subclavian Artery.

relapsing KD. Other diagnoses that could possibly explain the acute and intense chest pain referred by the patient such as angina or acute myocardial infarction were ruled out after performing transthoracic echocardiography (TTE); which revealed severe aortic regurgitation (**Fig-1a** and **Fig-1b**). Follow-up with transesophageal echocardiography (TEE) also showed severe aortic insufficiency (Carpenter type 1) with important dilation of the aortic ring (**Fig-1c**).

Investigations

A magnetic resonance imaging study (MRI) was performed, demonstrating a newly formed fusiform aneurysm at the location of the previous KD measuring 59 mm in diameter (**Fig-2**). The study also showed

early gadolinium enhancement, suggesting acute inflammation in the ascending aorta. A Positron Emission Tomography (PET) study with 18-fluorodeoxyglucose (18-FDG) was performed, exhibiting abnormally high metabolic activity in the wall of the ascending aorta, thus confirming the diagnoses of aortitis and a relapsing KD (**Fig-3**). As a complementary study, an aortogram was performed, showing clear evidence of the relapsing KD in the proximal segment of the aberrant right subclavian artery (**Fig-4**).

Management

Due to the patient's clinical presentation and current findings, surgical intervention was performed. Exclusion of the aneurysm, three-staged revascularization of the supra-aortic vascular branches, and aortic valve replacement with a bioprosthetic valve were carried out successfully and the patient recovered during the following months.

Discussion

KDs are uncommon congenital vascular abnormalities characterized by a dilation of the proximal segment of an aberrant right or left subclavian artery [1]. This patient had an aberrant right subclavian artery, which occurs when the right portion of a double aortic arch involutes into the segment between the right common carotid artery and the right subclavian artery. This results in the right subclavian artery arising as the fourth and last great branch of an otherwise normal aorta and subsequently passing to the right side of the body; behind and in close relation to the esophagus and trachea (**Fig-5**). We now know that even if 80% of the cases are retroesophageal, some of them can be located between the esophagus and the trachea, or even anterior to the trachea [2]. Additionally, around half of the patients with this condition have a higher risk for the development of aortic aneurysms, ruptures or dissections than normal individuals [3].

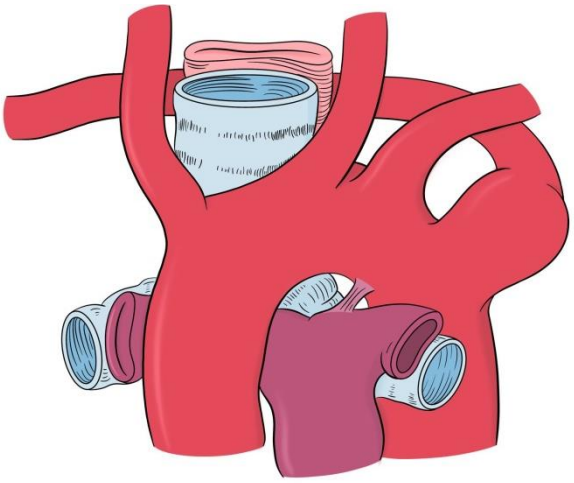

Fig-5: Aberrant right subclavian origin with a KD

An aberrant right subclavian artery with an aneurysmal dilation without aortic involvement, as described by Kiefer et al. [9]. A KD results from aneurysmal degenerative changes involving the thoracic aorta. [Image of own authorship]

Until nowadays, KDs were believed to be the result of a failure of regression of the primitive fourth dorsal arch that gives way to an abnormal remnant of this embryonic structure [4]. Nonetheless, there is now an ongoing debate, for in new literature, KDs are described as degenerative aneurysmal changes in the root of the aberrant right subclavian artery and not a congenital anomaly [5]. This condition was first described in 1936 by Burckrad Kommerell, a German radiologist that identified the anomaly after performing an esophagogram to a patient that exhibited a compressive and pulsatile mass posterior to the esophagus [1].

This condition has a prevalence of approximately 0.7-2% when found with an aberrant right subclavian artery, and of 0.04-0.4% when related to an aberrant left subclavian artery [6], the first type being the fourth most common congenital anomaly of the aortic arch [7]. Moreover, around 40% of the patients with an aberrant right subclavian artery present with a KD [8]. Most patients with KDs are asymptomatic, however, some of them (approximately 5%) can present a wide variety of clinical manifestations such as chest pain, coughing, dysphagia, exertional dyspnea, a difference in blood pressure between both upper limbs, and ischemia of the upper limb of the affected side due to occlusive complications [9]. In the most severe of scenarios, a spontaneous rupture or dissection of the diverticulum may occur, affecting around 6-19% of the cases with a 50% mortality rate. Another relevant but very rare complication is the formation of an esophageal or tracheal fistula, generated by either constant compression of the aberrant subclavian artery or procedures such as

placement of endotracheal or nasogastric tubes. Nonetheless, the most common iatrogenic complication with these patients is puncture or laceration of the aberrant subclavian artery during central venous catheterization [2]. Finally, aortitis is also a very unusual complication of this pathology, but it can lead to potentially adverse situations, just as the recurrence of the aneurysm after previous surgical correction [10], as exemplified in the present case.

Currently, the best tools available for the diagnosis of KDs are MRI and computed tomography (CT), since they have the ability to accurately observe and characterize vascular anomalies, their relationship to nearby organs or structures, and possible coexisting malformations [11]. Additionally, incidental findings in other diagnostic modalities such as chest x-ray or esophagogram in conjunction with symptoms compatible with esophageal compression should lead to the diagnostic suspicion of a KD [12]

There are currently no specific guidelines regarding the treatment of this condition. Nonetheless, surgical interventions should be considered in symptomatic patients or in those with a diverticulum diameter bigger than 30 mm [13]. Surgical approach should be determined by the anatomic features of the KD, clinical presentation, comorbidities, past medical history and hemodynamic state to ensure an optimal treatment. Treatment must address the aneurysm as well as revascularization of the upper extremity [14].

Surgical repair is performed with the patient in supine position, with the right hemithorax elevated. The great blood vessels and the distal part of the aberrant left subclavian artery are exposed through a median sternotomy. Cardiopulmonary bypass is performed through the femoral artery and/or cannulation of the ascending aorta and bicaval drainage with pulmonary artery ventilation. Then, reconstruction of the aortic arch is performed in the following order: right subclavian artery, right common carotid artery and finally the left common carotid artery. After reconstruction, thoracotomy is performed in the 3rd, 4th or 5th intercostal space through which the diverticulum is exposed. After cross-clamping, the descending aorta is transected and anastomosed to the distal end of the graft. Finally, the graft is anastomosed with the ascending aorta [15].

Open surgical repair is associated with high morbidity as a major thoracic surgery. The procedure involves resection of the diverticulum and revascularization of the aberrant subclavian artery, which may be performed through a thoracotomy or median sternotomy [16].

Revascularization with the aortic arch can be performed through a sternotomy or extra-anatomically through a supraclavicular approach. Hybrid repair may be performed using an aortic endograft and extra-anatomic bypass in order to reduce morbidity associated with thoracotomy or sternotomy and aortic cross-clamping [17].

Many surgical approaches for repair of a KD have been described. We have briefly summarized open, hybrid and endovascular techniques in **Table-1**.

Table-1: Brief summary of the characteristics of the surgical techniques used to repair KDs

Technique Name	Open Surgical Repair	Hybrid Repair	Endovascular Repair
Approach	Right thoracotomy/Median sternotomy/Supraclavicular	Median sternotomy/Cervical/Endovascular	Endovascular
Description	Resection of the KD with revascularization of the ASA.	A number of techniques: ETEC, FET, TESG-CD.	Pure endovascular stent-grafting, repair and revascularization.
Grafts	None	Endograft with extra-anatomic bypass.	Chimney or Snorkel stent-grafts; branched thoracic endografts
Stages	One- or two-staged	Single-staged	Single-staged
Usage	More frequent	Less frequent	Rare

KD: Kommerell diverticulum; ASA: aberrant subclavian artery; ETEC: elephant trunk with endovascular completion; FET: frozen elephant trunk; TESG-CD: thoracic endovascular stent-grafting and cervical debranching [15,16]

Follow-Up

After surgery, the patient was subsequently admitted to the intensive care unit for three days, where she was intubated and under close surveillance. She improved steadily during the following weeks, prompting hospital discharge. She remained asymptomatic and recovered steadily during the following months.

Conclusions

KDs are uncommon congenital heart diseases involving the aortic arch and should be suspected in patients with symptoms compatible with esophageal compression and/or chest pain. Multimodal approach to this condition is vital to ensure the best outcomes for patients.

Conflict of Interest

The authors have read and approved the final version of the manuscript. The authors have no conflicts of interest to declare.

References

[1] Torres-Martel JM, Izaguirre-Guajardo G, Ramírez-Portillo CI. Divertículo de Kommerell (DK) [Kommerell's Diverticulum (KD)]. Gac Med Mex. 2016 May-Jun;152(3):424-28. Spanish. [**PMID**: 27335200]

[2] Yang C, Shu C, Li M, Li Q, Kopp R. Aberrant subclavian artery pathologies and Kommerell's diverticulum: a review and analysis of published endovascular/hybrid treatment options. J Endovasc Ther. 2012 Jun;19(3):373-82. [**PMID**: 22788890]

[3] Cinà CS, Althani H, Pasenau J, Abouzahr L. Kommerell's diverticulum and right-sided aortic arch: a cohort study and review of the literature. J Vasc Surg. 2004 Jan;39(1):131-39. [**PMID**: 14718830]

[4] Edwards JE. Anomalies of the derivatives of the aortic arch system. Med Clin North Am. 1948 Jul;32:925-49. [**PMID**: 18877614]

[5] Timaran CH. Upper extremity aneurysm. In: Rutherford JB, Cronenwett JL, Johnston W, eds. Rutherford's Vascular Surgery. 7st ed. Philadelphia: WB Saunders; 2010.

[6] Lee MW, Kim GH. Kommerell's diverticulum: a rare cause of esophageal subepithelial lesion. Korean J Intern Med. 2019 Nov;34(6):1392-93. [**PMID**: 29761792]

[7] Kaufman JA. Thoracic aorta. In: Kaufman JA, Lee MJ, eds. Vascular and Interventional Radiology: The Requisites. 1st ed. Philadelphia: Mosby; 2003:219–45.

[8] Kadir S. Regional anatomy of the thoracic aorta. In: Kadir S, ed. Atlas of Normal and Variant Angiographic Anatomy. Philadelphia: WB Saunders; 1991:19–54.

[9] Freed K, Low VH. The aberrant subclavian artery. AJR Am J Roentgenol. 1997 Feb;168(2):481-84. [**PMID**: 9016231]

[10] Ichikawa T, Koizumi J, Tanno K, Okochi T, Nomura T, Shimura S, Imai Y. Kommerell Diverticulum in Adults: Evaluation of Routine CT Examinations. Tokai J Exp Clin Med. 2016 Jun 20;41(2):65-69. [**PMID**: 27344995]

[11] Malik TH, Bruce IA, Kaushik V, Willatt DJ, Wright NB, Rothera MP. The role of magnetic resonance imaging in the assessment of suspected extrinsic tracheobronchial compression due to vascular anomalies. Arch Dis Child. 2006 Jan;91(1):52-55. [**PMID**: 15871985]

[12] Tanaka A, Milner R, Ota T. Kommerell's diverticulum in the current era: a comprehensive review. Gen Thorac Cardiovasc Surg. 2015 May;63(5):245-59. [**PMID**: 25636900]

[13] Van Rosendael PJ, Stöger JL, Kiès P, Vliegen HW, Hazekamp MG, Koolbergen DR, Lamb HJ, Jongbloed MRM, Egorova AD. The Clinical Spectrum of Kommerell's Diverticulum in Adults with a Right-Sided Aortic Arch: A Case Series and Literature Overview. J Cardiovasc Dev Dis. 2021 Feb 26;8(3):25. [**PMID**: 33652796]

[14] Morishita A, Tomioka H, Katahira S, Hoshino T, Hanzawa K. Surgical Treatment for Kommerell's Diverticulum Associated with a Right-Sided Aortic Arch and an Aberrant Left Subclavian Artery: Endovascular or Hybrid. Ann Vasc Dis. 2019 Jun 25;12(2):228-32. [**PMID**: 31275480]

[15] Tsukui H, Aomi S, Yamazaki K. Surgical strategy for Kommerell's diverticulum: total arch replacement. J Thorac Cardiovasc Surg. 2014 Oct;148(4):1423-27. [**PMID**: 24365271]

[16] Erben Y, Brownstein AJ, Velasquez CA, Li Y, Rizzo JA, Mojibian H, Tanweer M, Zafar MA, Ziganshin BA, Elefteriades JA. Natural history and management of Kommerell's diverticulum in a single tertiary referral center. J Vasc Surg. 2020 Jun;71(6):2004-11. [**PMID**: 31708305]

[17] Vucemilo I, Harlock JA, Qadura M, Guirgis M, Gowing RN, Tittley JG. Hybrid repair of symptomatic aberrant right subclavian artery and Kommerell's diverticulum. Ann Vasc Surg. 2014 Feb;28(2):411-20. [**PMID**: 24200134]

[18] Idrees J, Keshavamurthy S, Subramanian S, Clair DG, Svensson LG, Roselli EE. Hybrid repair of Kommerell diverticulum. J Thorac Cardiovasc Surg. 2014 Mar;147(3):973-76. Erratum in: J Thorac Cardiovasc Surg. 2016 Jul;152(1):291. Idrees, Jahanzaib J [corrected to Idrees, Jay J]. [**PMID**: 23535153]