No Pain and No Gain: Kommerell’s Diverticulum with a Right-Sided Aortic Arch, and an Aberrant Left Subclavian Artery Presenting in a Body Builder with Muscle Bulk Imbalance

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ABSTRACT: A 22-year-old right-handed male presented to the Sports Medicine clinic with concerns of upper extremity muscle asymmetry. Physical examination showed gross muscular asymmetry when comparing the left upper extremity to the right. Radial pulses were 2+ on the right and 1+ on the left. Due to concern for vascular anomaly, computed tomography angiography was performed which revealed a right-sided aortic arch with Kommerell’s diverticulum and aberrant left subclavian artery. The patient underwent a left carotid subclavian bypass successfully, but his recovery was complicated by an upper extremity deep venous thrombosis. He is currently on novel anticoagulant but has been released to normal activities and doing well. Kommerell’s Diverticulum (KD) is a rare congenital anomaly caused by a persistent remnant of the fourth primitive dorsal arch during embryological development. Although the prevalence of KD is rare, it is important to identify and diagnose this condition to provide definitive care.

KEYWORDS: Kommerell’s Diverticulum, sports medicine, vascular surgery

Introduction
Kommerell’s diverticulum (KD) is a rare congenital anomaly caused by a persistent remnant of the fourth primitive dorsal arch during embryological development. The most common KD is found with the diverticulum at the right aortic arch with an aberrant left subclavian artery. Other reported KD variations include diverticulums at the left aortic arch with an aberrant right subclavian artery, diverticulum located at the aortic-ductal junction, and a diverticulum at the left aortic arch with a right descending aorta and a right ligamentum arteriosum. Symptoms of individuals with KD include dysphagia, chest pain, and shortness of breath in children. Physical exam findings may reveal blood pressure differences in the upper extremities as well as asymmetric pulses. We detail the case of a bodybuilder who presented with an imbalance of muscle bulk, a physical exam finding not previously described in KD, to the upper extremities and will discuss the pathophysiology, diagnosis, and treatment of KD.

Case Report
A 22-year-old right-handed male presented to the Sports Medicine clinic with concerns of muscle asymmetry of his upper extremities. The patient previously played college football but currently works at a gym and lifts weights daily. He noticed that while lifting he is able to sustain muscular contraction of the right upper extremity for a longer period than his left. He has also noted greater muscle bulk of the right shoulder, biceps, and triceps on the right upper extremity when compared to the left. The patient notes that he has had acromioclavicular (AC) joint separations on both shoulders due to football but has healed well from each. He has no other past medical history aside from an appendectomy. The patient is a fraternal twin and was a known triplet, however, one fetus was lost in utero. His initial vital signs were T: 36.5°C HR: 64 BP: 138/65 mm Hg RR: 14.

Gross examination showed great asymmetry of muscle bulk on the right upper extremity compared to the left but with equal muscle bulk of the lower extremities. Cardiac exam revealed a regular rate and rhythm without rubs, murmurs, or gallops. The right radial pulse was 2+ and the left radial pulse was 1+. Dorsalis pedis pulses were 2+ bilaterally. Capillary refill was less than 3 seconds bilaterally. Strength was 5/5 in bilateral upper extremities. Sensation was intact to bilateral upper extremities. The patient’s shoulder exam was equal bilaterally and negative for Hawkins, Neer’s, and Speeds. No scapular winging was noted. Measuring the patients mid-humerus revealed a regular rate and rhythm without rubs, murmurs, or gallops. The right radial pulse was 2+ and the left radial pulse was 1+. Dorsalis pedis pulses were 2+ bilaterally. Capillary refill was less than 3 seconds bilaterally. Strength was 5/5 in bilateral upper extremities. Sensation was intact to bilateral upper extremities. The patient’s shoulder exam was equal bilaterally and negative for Hawkins, Neer’s, and Speeds. No scapular winging was noted. Measuring the patients mid-humerus showed the right arm to be approximately 4 cm larger in circumference compared to the left (Figure 1).

After exam, differential diagnosis included thoracic outlet syndrome, cervical radiculopathy, coarctation of the aorta, Kommerell’s diverticulum with left subclavian artery stenosis, and quadrilateral space syndrome. The initial concern, given the atrophy of the left chest and upper extremity, radiculopathy, or cervical stenosis could not be disregarded.
Plain films of the left shoulder showed no abnormalities. Venous duplex of the left upper extremity showed patency of arteries and venous structures. Computed tomography angiography (CTA) imaging was obtained on an outpatient basis to further evaluate the patient’s condition and showed a right-sided aortic arch with Kommerell’s diverticulum and aberrant left subclavian artery. Significant stenosis at the origin of the left subclavian artery was also noted. The patient was instructed to restrict weightlifting to light workouts only (Figure 2).

Given these findings, the patient was referred to vascular surgery and underwent further diagnostic imaging and presurgical evaluation with an aortic arch angiogram. This confirmed a right sided aortic arch and aberrant left subclavian artery takeoff from the descending thoracic aorta with diverticulum and stenosis just distal to the takeoff of the artery. The patient underwent surgical fixation with vascular surgery undergoing resection of the Kommerell diverticulum and carotid subclavian bypass with an 8 mm Dacron graft. His recovery was complicated by a deep venous thrombosis to the subclavian vein. He is now on a novel anticoagulant and has started a light exercise program without further complications.

**Discussion**

The prevalence of KD with right aortic arch with aberrant left subclavian artery has been reported to be 0.04% to 0.4%. Of this population, right aortic arch with aberrant left subclavian artery is twice as common in men than in females with females more often presenting with left aortic arch with aberrant right subclavian artery. KD anomaly was first discovered by a German radiologist, Burckhard F. Kommerell, in 1936. Most commonly, children will present with breathing difficulties secondary to the development of the trachea and malleable tracheal rings. Case reports of wheezing secondarily to tracheal narrowing has been reported as well as hoarseness secondary to recurrent laryngeal nerve has been detailed. Adults will more commonly present with dysphagia, chest pain, or blood pressure differences in the upper limbs. Often, KDs are found in asymptomatic patients on CT imaging.

Surgical intervention is recommended for symptomatic patient’s, however, there is no guideline for asymptomatic KDs. Open surgical treatments include reconstruction of the aberrant subclavian artery, resection of the KD with division of ligamentum and translocation of the subclavian artery (pediatric patients), and graft replacement and reconstruction of the aberrant subclavian artery. Endovascular repair has also been reported with good success including hybrid endovascular repair with mini upper sternotomy and total endovascular aortic repair (TEVAR), total debranching of supra-aortic vessels via midsternotomy and TEVAR among others. Total endovascular repair with TEVAR has also been reported.
Reported outcomes of the above procedures are excellent with 96% of patients reported symptom relief and an operative mortality of 0% at 30 days.\textsuperscript{7} However, in one retrospective study, adverse outcomes of the procedures were significant for stroke, tracheostomy, and bleeding.\textsuperscript{7} Looking at both open and endovascular surgery 8.0% of patients had a stroke, 5.0% received a tracheostomy, and 5.0% had bleeding.\textsuperscript{7} Late all-cause mortality was seen in 11.0% of patients.\textsuperscript{7} It is important to note that given the abnormal forces through a KD there is a high risk of rupture and dissection.\textsuperscript{1} Given these risks, many experts recommend early surgical intervention when compared to standard thoracic aortic aneurysms even given the risks of surgery.\textsuperscript{1}

**Author Contributions**
All authors contributed to the case report conception. Material preparation, data collection and analysis were performed by Scott Meester, Diego Riveros, Aaron J. Monseau, and Brenden J. Balcik. The first draft of the manuscript was written by Scott Meester and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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