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Aberrant Right Subclavian Artery and Stanford Type B Aortic Dissection

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Abstract An aberrant right subclavian artery (ARSA) is a rare developmental anomaly (0.4-1.8%) of the aorta in which the right subclavian artery arises from the aortic arch distal to the origin of the left subclavian artery often coursing behind the esophagus to reach the right arm. It courses behind the esophagus in about 80% of cases, between the esophagus and the trachea in 15%, and anterior to the trachea or mainstem bronchus in 5%. Patient with this anomaly rarely have symptoms (90-95%) but when symptomatic the ARSA give rise to symptoms of dysphagia lusoria, dyspnea and chronic cough. In a vast majority of patients ARSA is clinically silent till right radial angiography is performed. We are reporting a case of dissection of the retroesophageal right subclavian artery with extension to the descending thoracic aorta (Stanford Type B).

Keywords: *aberrant subclaviaartery, Standford type B aortic dissection, dyphagia lusoria, Kommerell's diverticulum, cardiovocal syndrome, Ortner's syndrome*

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1. Introduction

An Aberrant Right Subclavian Artery (ARSA) is a rare developmental anomaly (0.4-1.8%) of the aorta in which the right subclavian artery arises from the aortic arch distal to the origin of the left subclavian artery often coursing behind the esophagus to reach the right arm [1].

Hanuld in 1735 first described this arterial anomaly but its symptomatic form "dysphagia lusoria" in patients with dysphagia was described by Bayford in 1787[2,3].

A German Radiologist, Burkhard Friedrich Kommerell observed that the ARSA often arises from a dilated segment of the descending aorta and described it as remainder of the primitive right dorsal aorta. This vascular anomaly often seen with ARSA but not limited to it has been subsequently named the Lusorian artery or Kommerell's diverticulum [4].

Since the introduction of diagnostic trans-radial catheterization in 1989 and its use for coronary intervention in 1993, the trans-radial approach has become increasingly popular. However, a review of literature shows that successful right radial cardiac catheterization could be successfully performed in only 60% of cases of ARSA [5]. This is because the right trans-radial approach in patients with ARSA makes it difficult to approach the aorta root and requires the catheter to curve back into the ascending aorta.

We are reporting a case of dissection of the retroesophageal right subclavian artery traveling into the descending thoracic aorta (Stanford Type B) during right radial cardiac catherization performed on a patient presenting with non-ST elevation myocardial infarction.

2. Case

A 58-year-old female with no previous medical history came to the emergency department after feeling that "her heart abruptly began pounding" while at work. She stated that her work as an airline ground staff involved walking large distances every day and never had she experienced such symptoms before. Her symptoms had persisted for 3-4 hours and were now associated with a sensation of midchest tightness and a feeling of lightheadedness. She had been a lifetime non-smoker, occasionally drank a glass of wine and had never abused any illicit drugs. An electrocardiogram (ECG) demonstrated a narrow complex tachycardia with no discernable P waves, at a rate of 160 beats per minute. An attempt at closed nose blowing (Valsalva Maneuver) reverted her supraventricular tachycardia (SVT) back to normal sinus rhythm at a rate of 98 beats per minute. The palpitations resolved but she continued to have mild discomfort in the center of her chest. Her blood pressure was 143/90 mm of Hg, temperature 98.3^0F and she was saturating 99% breathing 18 breaths per minute of room air. Her cardiovascular

examination was noncontributory. She had no jugular venous distention, a regular heart rate with no murmurs or gallop on auscultation, equal air entry in her lungs and no lower extremity edema. Laboratory analysis revealed a troponin elevation of 1.39 ng/ml (normal <0.015 ng/ml). All other hematological and biochemical tests were within normal limits. Her elevated troponin was attributed to a Type II myocardial nnfarction from the SVT (likely atrioventricular nodal reentrant tachycardia). An electrophysiological (EP) study and a with possible ablation was planned. However because of the continuous chest tightness and elevation in the troponin, a cardiac catheterization was performed prior to the EP study to rule out coronary artery disease.

Her right radial artery was accessed using the modified Seldinger technique and a 6 French Glidesheath was placed. A J wire was advanced under fluoroscopic guidance to the level of the arch of the aorta. Even with deep inspiration the wire repeatedly selectively entered the descending aorta instead of the ascending aorta [Figure 1A]. Subsequent attempts to direct an angled guidewire into the ascending aorta resulted in the patient experiencing central crushing chest pain and all catheter and wire manipulations were stopped. Subclavian angiography during catheter withdrawal demonstrated dissection of an aberrantly origination right subclavian artery [Figure 1B]. The patient's pain resolved. Cardiothoracic surgery's guidance was obtained and the patient was taken immediately for a chest computed tomography (CT) scan.

The CT scan demonstrated dissection of the retroesophageal right subclavian artery [Figure 1C]. The dissection extended down the descending aorta and terminated in the iliac arteries with no obvious malperfusion [Figure 1D]. The Stanford B dissection was managed medically with an esmolol drip. A repeat CT scan 3 days after did not show any progression of the aortic dissection and the patient remained free of chest pain. She was discharged home on oral beta blockers with a plan for a short follow-up with cardiothoracic surgery and a repeat CT scan in 4 weeks.

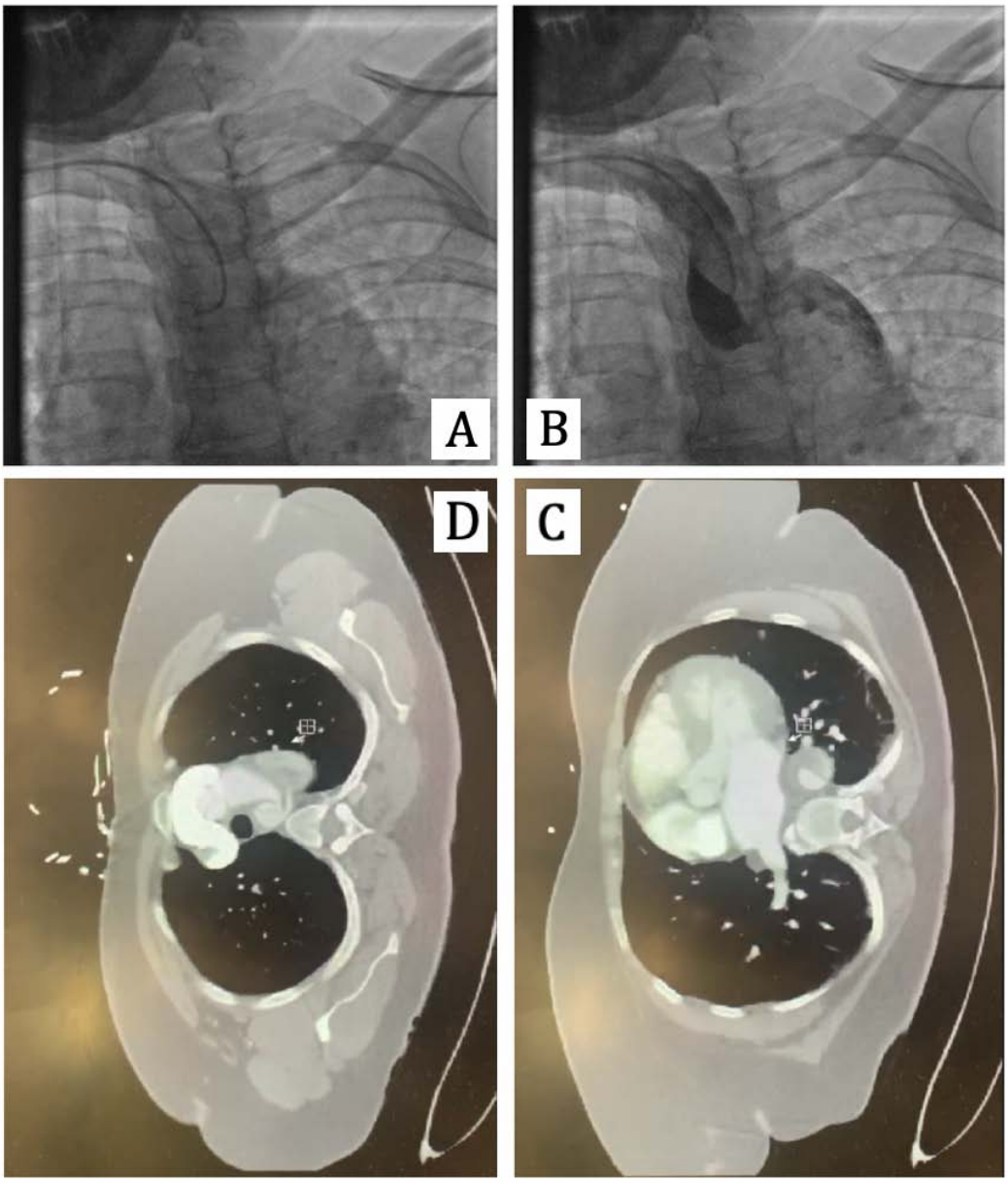

Figure 1. Clockwise from top left: **A.** JR catheter in the aberrant right subclavian artery. **B.** Dissection of the aberrantly originating right subclavian artery. **C.** Dissection extending down the descending aorta. **D.** Dissection of the retroesophageal right subclavian artery

3. Discussion

The aorta develops in the 3rd week of gestation [6]. Normally, the right aortic arch regresses caudal to the origin of the 7th intersegmental artery which subsequently gives rise to the right subclavian artery. In about 0.4-1.8% of individuals, the regression occurs between the right common carotid and the 7th intersegmental artery. This causes the right subclavian artery to be connected to the left dorsal aorta via the right dorsal aorta, which in normal development would have otherwise regressed. During the course of its development, the origin of the aberrant right subclavian migrates until it is distal to the origin of the left subclavian artery [1].

The aberrant right subclavian artery (ARSA) courses behind the esophagus in about 80% of cases, between the esophagus and the trachea in 15%, and anterior to the trachea or mainstem bronchus in 5% [7]. ARSA is seen more often in women and has a female to male prevalence of 3:1 in the general population [8]. When present, ARSA may be associated with other vascular anomalies like truncus bicaroticus (19.2%), Kommerell's diverticulum (14.9%), aneurysm of the artery itself (12.8%), and a right sided aortic arch (9.2%). Prevalence of this anomaly is higher (26-34%) in patients with Down syndrome and other congenital abnormalities. Patients with this anomaly rarely have symptoms (90-95%) but when symptomatic the ARSA give rise to symptoms of dysphagia lusoria, dyspnea and chronic cough [1,7]. Rarely, the ARSA may lead to Ortner's syndrome (cardiovocal syndrome) by causing compression of the recurrent laryngeal nerve leading to stridor [9].

A study of 10,260 aortic CT angiography records of 6,804 Chinese patients reported higher rates of aortic dissection or coarctation in patients with ARSA when compared to the control group (52.6% vs 36.8%, p=0.006; 5.1% vs 0.9%, p=0.007, respectively) [10].

Iatrogenic aortic dissection when occurring in the context of arteria lusoria tend to be typically be Stanford B in variety and extend down the descending aorta given the location of the origin of the artery in the descending aorta distal to the left subclavian artery [11].

It is often difficult to identify ARSA on the postero-anterior projection during conventional angiography. The guidewire repeatedly entering the descending aorta during the right radial approach should alert the operator to the possibility of this vascular anomaly. This should prompt an oblique view of the right subclavian artery angiogram which would then better demonstrate the origin of the ARSA distal to the left subclavian artery [12,13].

4. Conclusion

In a vast majority of patients, ARSA is clinically silent. However, it can lead to symptoms of dysphagia lusoria or a cardiovocal syndrome due to compression of the esophagus or the recurrent laryngeal nerve respectively. Aortic dissection or coartation can also be seen among ARSA patients. Our case highlights the importance of considering this rare but not infrequent coronary anomaly when the J wire selectively enters the descending aorta during right radial catheterization. In such a case, catheterization of the ascending aorta may be difficult or even impossible (7.1%) [12]. Unsuccessful attempts at navigating the wire into the ascending aorta pose the risk of trauma to the endothelial lining and potential adverse outcomes like aortic dissection [14] and intramural hematoma formation [15]. They should prompt the angiographer to obtain a subclavian angiogram with oblique views and if unsuccessful consider accessing the ascending aorta either from the left radial or the femoral approach.

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