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## **Case Report**

Subclavian Steal Syndrome Due to a Subtotal-Occlusion of an Aberrant Right Subclavian (Lusorian) Artery-A Case Report

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# Abstract

Despite its rare incidence, an occlusion or near-occlusion of an aberrant right subclavian artery (lusorian artery) presents a challenging pathology to treat. While open surgical approaches have been regarded as first-line treatment for a long time, endovascular solutions have proven over the last years to be a less invasive yet effective alternative, like in many other vascular conditions.

We report on a 47-year-old female complaining of dizziness and vertigo, together with a weakness in the right arm aggravated by manual activities.

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Other than obesity (BMI of 34 kg/m²) and smoking (45 Pack years), no other relevant history was present. A series of intensive neurological diagnostics revealed no pathological findings. However, duplex ultrasound of the neck vessels showed a retrograde flow in the right vertebral artery secondary to a subtotal occlusion of the right Subclavian Artery (SA). Complimentary CT-angiography additionally revealed an aberrant origin of the SA from the descending aorta.

Due to the location of the lesion and the expected technical difficulties in obese patients, we opted for an interventional endovascular angioplasty. Through this case report we aim to present the role of endovascular interventions in the treatment of occlusions and stenoses in the context of an abnormal anatomy.

**Keywords:** Aberrant right subclavian artery; Endovascular angioplasty; Subclavian steal syndrome

### **Background**

The right aberrant subclavian artery, also known as lusorian artery, is estimated to be the most common embryological anomaly of the aortic arch, despite its rare prevalence [1]. The term describes an aberrant branching, where the innominate artery is not present and four main arteries branch separately off the aortic arch. In such case the right subclavian artery passes behind the esophagus to continue its path into the subclavian region. The incidence of the lusorian artery is reported to vary between 0.5-2% and is highly common among patients with trisomy 21 (up to 100%) [2,3].

An aberrant origin with an atypical course of the right subclavian artery could result in many pathological conditions, which have been classified by Kieffer et al. into four major groups [4]:

- Group 1: Patients with dysphagia caused by non-aneurysmal aberrant RSA (dysphagia lusoria)
- Group 2: Patients with symptomatic occlusive disease of non-aneurysmal aberrant RSA
- Group 3: Patients with aneurysmal aberrant RSA without aortic lesions, with or without oesophageal compression or arterial thromboembolism
- Group 4: Patients with an aortic (usually aneurysmal) lesion involving the origin of the aberrant RSA, with or without aberrant RSA aneurysm

Owing to the general trend in modern vascular surgery, open treatment modalities of the above conditions are being increasingly replaced by novel endovascular solutions [5], unless other aortic arch deformations necessitate an open approach [6]. Surgical therapies themselves have been improved with hybrid-techniques [6], especially in case of dysphagia and aneurysms, resulting in low postoperative morbidity [7,8]. Nevertheless, no guidelines or systematic reviews addressing treatment regimens have been published so far.

#### **Case Presentation**

#### History

We present the case of a 47 year old female who was primarily referred to our inpatient neurology department due to unclear persisting dizziness and vertigo, which have been deteriorating for a year. While the complaints were continuous, they tended to worsen upon turning the head in both directions and while turning around in bed. Swallowing difficulties, nausea and vomiting were denied.

Other than Obesity (BMI of 34 kg/m²), smoking (45 Pack years) and a conservatively treated cervical disc protrusion (C5-C6), her previous medical history was unremarkable.

#### **Examination and diagnostics**

To the time of presentation, Magnetic Resonance Imaging (MRI) and Electroencephalography (EEG) had already been ordered by an outpatient practicing neurologist and have shown no abnormal results. The patient was hence initially diagnosed with vestibular paroxysmia and treated with Carbamazepine. This treatment didn't result in any improvement and had to be stopped two weeks after that due to intolerance. The patient later complained of increasing weakness in her right leg upon strenuous exertion.

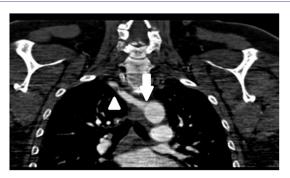
Clinically, the patient was in a good general condition. Brachial blood pressure was considerably lower on the right side and the radial pulse was relatively weaker ( $\Delta$  60 mmHg). The clinical neurological exam was normal and showed no focal or general pathologies (including normal reflexes and no nystagmus).

Blood tests revealed elevated cholesterine and LDL values. Taking the blood pressure difference and the existing cardiovascular risk factors into consideration, a duplex ultrasound of the supraaortic vessels was carried out. While both carotid arteries exhibited a regular flow, post-stenotic disturbances (peak systolic velocity of up to 270 m/s) were found in the right subclavian artery. In addition, the right vertebral artery showed a retrograde flow which decreased upon compression of the arm, suggesting a subclavian steal syndrome.

Further investigation with CT angiography showed an 8 mm long high-grade stenosis of the right subclavian artery, directly proximal to the origin of the vertebral artery (Figure 1), together with an aberrant origin directly from the descending a rta (Figure 2).



**Figure 1:** Computer assisted 3D reconstruction of the aortic arch showing the aberrant origin of the right subclavian artery directly from the descending aorta (arrow) and the stenosis (triangle).



**Figure 2:** CT angiogram showing the aberrant origin of the right subclavian artery directly from the descending aorta (arrow) and the stenosis (triangle).

After an inter-disciplinary discussion of the case by neurologists, interventional radiologists and vascular surgeons, an interventional treatment of the subclavian stenosis was recommended.

#### How we did it

After patient informed consent, the procedure was initiated by an ultrasound-guided retrograde cannulation of the weakly-palpable right common femoral artery and the insertion of a 4-F introducer over a starter Guide wire, which was replaced with a rounded soft-tip guide wire (0.035" outer diameter, 260 cm long, Radifocus®, Terumo Interventional Systems). The latter was placed into the thoracic aorta, over which a Bernstein-catheter was brought up to the calculated height of the aberrant origin of the subclavian artery (C4-C5). To facilitate the navigation, the application of different projections was necessary, among which the left anterior projection in 30° was most helpful.

The guide wire was then withdrawn, and a direct injection of contrast medium was used to identify and locate the target vessel, opposite to which the tip of the Bernstein-catheter was placed. Using slight axial rotation and supporting the tip of the catheter again the ridge of the subclavian branch, the guide wire was eventually successfully inserted in the right subclavian artery after several attempts. The catheter was then placed proximally, and the stenosis was identified through an antegrade angiography of the right subclavian artery, where the ipsilateral vertebral flow was absent (Figure 3). The Bernstein catheter was then exchanged with a 7F, 90 cm long guiding sheath (Destination®, Terumo Interventional System) over the guide wire in Seldinger-technique. A balloon expandable Stent (8 mm diameter, 27 mm length, Express LD®, Boston Scientific Corporation) was the placed into the stenotic segment and successfully implanted. The final angiographic examination showed a successful dilatation of the stenosis with a regular antegrade flow through the vertebral artery (Figure 4). An incidentally-diagnosed high grade stenosis of the right common iliac artery was then managed with the implantation of a 9 mm x 25 mm balloon expandable Stent (Express LD®, Boston Scientific Corporation).

# Results and Follow-Up

The patient's complaints resolved completely after the intervention and her blood pressure values were similar on both sides. She was then discharged with a lifelong single antiplatelet therapy (100 mg of Aspirin) and a Statin. She was also advised to cease

smoking. One year follow up examination of the right subclavian and iliac arteries was clinically and Duplex-Sonographically normal and the patient continued to lead a normal life.

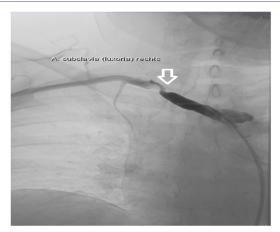


Figure 3: Antegrade angiography showing high grade stenosis of the lusorian artery (Arrow). Note the absence of the right subclavian artery.

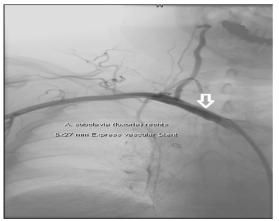


Figure 4: Result post-angioplasty showing successful dilatation of the subclavian stenosis with a regular antegrade flow in the right vertebral artery. The stent location is marked with an arrow.

#### **Discussion**

While unspecific indicators of atherosclerosis were evident, the vague complaints and symptoms of this patient have considerably contributed to the delayed diagnosis and treatment. Consequently, a targeted neurological exam would have only been complete with a simultaneous vascular evaluation. Computer assisted 3D reconstruction of CT findings was very helpful in the indication assessment and preprocedural planning and the wide range of available introducer sheaths, catheters and guide wires has inarguably added to the potential of endovascular interventions as in this case.

Through this case report we aim at drawing attention to the unique aspects of vascular anatomic variations such as the lusorian artery in the management, while praising the role of endovascular interventions in the treatment of occlusions and stenoses in such challenging anatomies. We also want to emphasize the importance of a global assessment of the cardiovascular patients and the key role played by interdisciplinary team work in contributing to correct diagnoses and effective treatment.

#### Conclusion

An aberrant right subclavian artery represents a rare anatomic vascular variation, which's pathologies have not been well addressed yet. Patients belonging to the second group of Kieffer's classification (Stenosis) are most likely to profit from the well-established endovascular interventional approach, which could be complicated by the challenging anatomies given.

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