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A forgotten vascular disease with important clinical implications. Subclavian steal syndrome

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Summary

Background: Subclavian Steal Syndrome (SSS) is a fascinating vascular phenomenon in which a steno-occlusive lesion of the proximal subclavian artery causes retrograde flow in the vertebral artery away from the brain stem subsequently causing vertebrobasilar insufficiency. SSS can present with a myriad of neurological and vascular signs and symptoms, but most commonly this phenomenon presents as an incidental finding in an asymptomatic patient.

Case Report: Our patient is a 73-year-old female sent to the cardiology clinic for surgical clearance in preparation for an elective cholecystectomy. Shortness of breath was her only complaint. Review of systems was remarkable for left arm pain and blurry vision with repetitive movement. Physical examination noticeable for absence of left radial pulse. Percutaneous angiography demonstrated a totally occulted left subclavian artery with collateral circulation form the vertebrobasilar apparatus.

Conclusions: Atypical presentation of this unique entity represents a challenge for physicians who require a high index of suspicion to make the diagnosis. We present an atypical case with radiographical evidence of the steal syndrome, followed by an extensive literature review of the most current diagnostic methods as well as latest recommendations for treatment options and secondary prevention.

key words: **subclavian steal syndrome • subclavian-steal-syndrome**

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BACKGROUND

Subclavian Steal Syndrome (SSS) is a fascinating vascular phenomenon in which a steno-occlusive lesion of the proximal subclavian artery causes retrograde flow in the vertebral artery away from the brain stem, subsequently causing vertebrobasilar insufficiency [1,2]. Most commonly this phenomenon presents as an incidental finding. The majority of patients are asymptomatic, however, symptomatic patients typically present with neurological symptoms, such as lightheadedness, hearing loss, or pre-syncope. Atypical presentation of this unique entity represent a challenge for physicians who require a high index of suspicion. We present an atypical presentation case with a review of the most current literature on subclavian steal syndrome diagnostic and treatment options.

CASE REPORT

Our patient is a 73-year-old female sent to the cardiology clinic for surgical clearance in preparation for an elective cholecystectomy. Her past medical history includes hypertension, dyslipidemia, and coronary artery disease with bypass surgery 11 years prior to presentation. Her current complaint is shortness of breath with minimal exertion. Review of systems was remarkable for left arm pain with repetitive movement. Occasionally, she would also experience blurry vision with the same activity. She denied dizziness, lightheadedness, and syncopal episodes.

Physical examination revealed a BP of 140/80 in the right arm, no blood pressure in left arm, and a heart rate of 70 bpm. The left radial pulse was absent. Cardiac ausculatation revealed a 2/6 systolic ejection murmur over the right base. Bilateral carotid bruits where noted but no subclavian bruits. Lungs where clear to auscultation. Pulses in lower extremities were intact.

Other diagnostic studies included an ECG that demonstrated sinus rhythm with no ST segment or T wave abnormalities and CXR revealed no acute cardiopulmonary process. The patient had recently undergone a nuclear stress test which revealed ECG abnormalities suggestive of inferior wall ischemia however the nuclear portion was unremarkable.

The patient underwent coronary angiography that revealed the following anatomy; Left main artery was free of disease. Circumflex artery was represented by an obtuse marginal branch with a 50 percent narrowing. Left anterior descending artery was closed in the proximal segment. Right coronary artery had a high grade stenosis in the range of 80 percent. The vein graft to the LAD and the vein graft to the diagonal branch where widely patent and angiographically free of disease.

Because of the suspected subclavian occlusion a selective left subclavian arteriogram and a nonselective abdominal aortogram where performed during this procedure. The subclavian artery was found to be totally occluded (Figure 1). Injection of the left carotid artery demonstrated good collateral circulation by forward flow into the internal carotid artery, Circle of Willis, basilar artery, and then a retrograde flow through the ipsilateral vertebral artery and filling the left subclavian artery (Figure 2). The aortogram revealed a high grade stenosis of the left renal artery and a patent right renal artery.

The subclavian steal syndrome review

The Subclavian Steal Syndrome is a rare yet well-known phenomenon that presents when a steno-occlusive lesion of the proximal subclavian artery results in the flow reversal of the vertebral artery, giving rise to vertebrobasilar insufficiency [1,2].

It was first described on 1961 by Reivich in the New England Journal of Medicine [1]. He presented a patient with neurological symptomatology and directly attributed to this reversal phenomenon. However, the term "Subclavian Steal" was introduced by Dr. Miller Fisher on an editorial a few months later [2].

The true prevalence of this syndrome is still unknown. It has been estimated by multiple authors from small or single center studies to be between 0.6% and 6% [3,4]. The controversy is generated because up to 80% of patients with flow reversal phenomenon are asymptomatic. Most patients with symptoms will have concurrent cerebrovascular lesions [5].

Subclavian steal symptoms presents secondary to arterial insufficiency, created by a retrograde flow that "steals" blood from the brain circulation, more specifically from the basilar artery via the vertebral artery. Classically it presents with neurological symptoms from the posterior brain and cerebellum [4,6]. Decreased flow over the basilar artery gives rise to symptoms like lightheadedness, ataxia, vertigo, dizziness, confusion, headache, nystagmus, hearing loss, presyncope and syncope, visual disturbances, focal seizures, and in extremely rare cases, death [6–10]. However the vast majority of patients are asymptomatic and rarely require any intervention [3,5,11].

The pathophysiology of SSS involves a proximal subclavian stenosis, which results in a lower pressure in the distal subclavian artery. Because the vertebrobasilar circulation is a closed hydraulic system, this pressure difference creates a retrograde flow, pulling blood from the contralateral vertebral artery to the basilar and then down the ipsilateral vertebral artery, "stealing" from the cerebral circulation [12–14]. This flow reversal has been classified as complete or incomplete meaning the flow reversal is continuous *vs.* intermittent [14,15]. The latter will reproduce the subclavian steal phenomena, but it is the continuous flow reversal that will be responsible for the majority of the true syndromes [14]. Because the vertebral artery serves as an important collateral circulation to the upper extremity, this same mechanism that may lead to arterial insufficiency and symptomatology from the arm. In such cases, patients may have atypical presentation with weakness of the arm and hand, claudication, cool temperature, and paresthesis [6]. Coronary-Subclavian syndrome is a similar entity that has been described in patients who have undergone CABG with LIMA and a subclavian stenosis reproduce the "steal" syndrome from the coronary arteries via the graft, giving rise to acute coronary symptoms [16–21].

The most common cause of this syndrome is atherosclerosis, which gives rise to stenosis and occlusion in more then 90% of the cases [17,22]. Because the left subclavian artery has a more prominent angle at its origin, turbulent flow will accelerate atherosclerosis and account for more

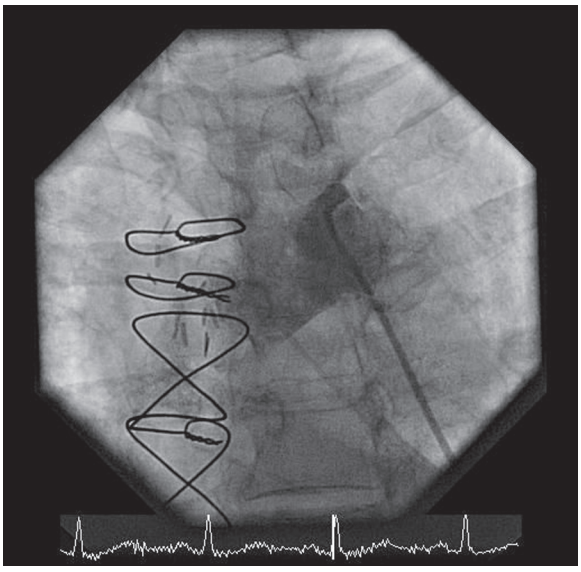

Figure 1. Selective angiogram of the left subclavian artery reveals a total occlusion just after its origin.

than 80% of the cases. In contrast, there is only a small number of patients that present with right sided symptoms and even fewer with bilateral [3]. Several other etiologies have been described as secondary causes of stenosis, most of them post surgical intervention, post radiation, congenital abnormalities, trauma, arteritis, and even thoracic outlet syndrome [23,24].

Disscusion

Due to the rarity of the subclavian steal syndrome, a high index of suspicion is necessary for diagnosis. The clinician must perform a detailed history and a careful physical examination looking for common subclavian steal syndrome findings to distinguish from the differential diagnosis that include intracraneal vascular disease, carotid artery disease, vertebral artery disease, brain tumors, and subdural hematomas [3,5,13,15,25]. The subclavian steal syndrome can present with absence or diminished ipsilateral radial pulse, cervical or supra clavicular bruits or thrills, and a presence of a systolic blood pressure difference greater than 20 mmHg between upper extremities [26]. Labropoulos et al. suggest that the later highly correlates with the presence of SSS. They also demonstrated a positive correlation of increasing arm BP differential with occurrence of symptoms [3,27]. Symptoms can be provoked with physical maneuvers that compress the vertebral artery, like rotation of the head towards the opposite side. Raising the patients arms can reproduce the syndrome and lead to a syncopal episode. Exercise can be used to induce neurological symptoms or it can also be performed to detect the disappearance of the radial pulse in the affected side [4].

Confirmation of a steal syndrome is usually made by imaging studies. All imaging tools can be used to detect subclavian stenosis and to observe reversal of flow from the vertebral artery. Carotid duplex ultrasound(US) is the most used and usually the first diagnostic test. US has been used as a screening tool because of accessibility and low cost. It can semi quantify subclavian stenosis and diagnoses other extracraneal carotid occlusive disease. Occasionally, transcraneal

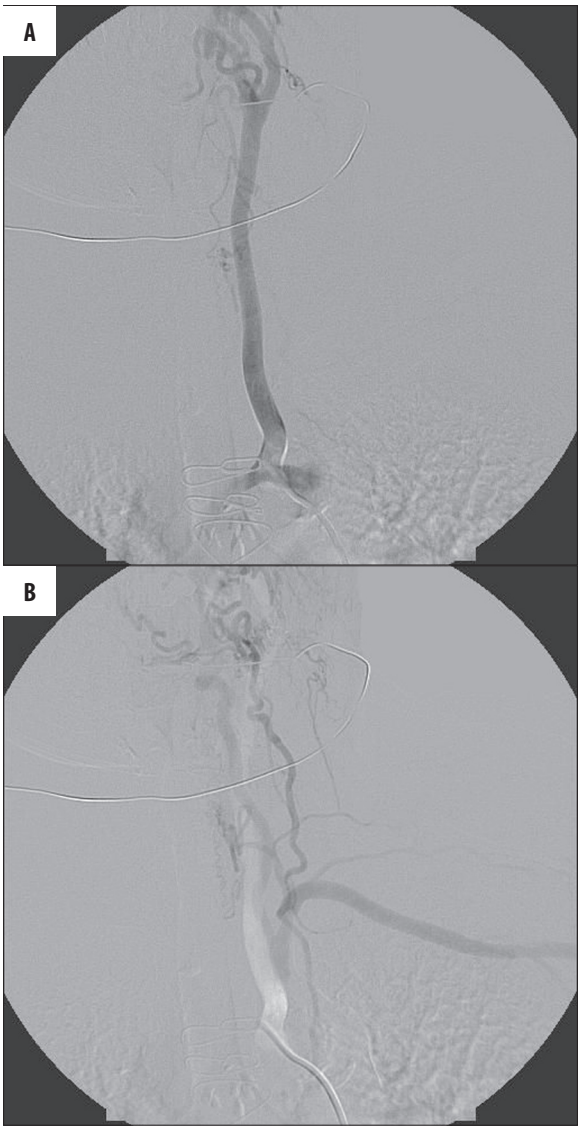

Figure 2A,B. Selective angiogram of the left carotid artery show retrograde filling of the left subclavian artery from the left vertebral artery.

ultrasound is utilized to evaluate the direction of the basilar artery blood flow [3,5]. Recently, higher quality images have been obtained through newer technology modalities such as MRA and have rapidly gained popularity [17,25]. When contrast MRA is not available, CT angiography is another excellent alternative [28]. Conventional cerebral angiography remains the gold standard, for it has the highest sensibility and specificity; However, this method caries the risks of an invasive procedure [10,23,27–31]. Additionally, in patients with CABG and LIMA, stress test with radiocontrast may be helpful to evaluate for coronary-subclavian syndrome [18–21,32].

Invasive treatments for SSS are usually reserved for highly symptomatic patients [11,15,33–35]. Current treatment options include subclavian artery angioplasty and stenting, and surgically bypassing the subclavian artery [10,30–31,36–38]. Stenting the subclavian artery is usually achieved through the femoral artery. It is the preferred method because of lower morbidity, shorter hospitalization, and faster recovery.

Retrospective studies suggest PTA with stent have grater than 93% success rate with sustained resolution of symptoms [30]. Inability to cross the occlusion is usually the only cause of failure of this method. DeVries et al concluded that major complications of PTA for SSS, such as stroke and death, can be has high as 3.6%, however surgical correction have a higher surgical risk and a variable success rate, estimated between 73–95% [36,38]. When surgically bypassing the subclavian stenosis, both prosthetic grafts and autogenous veins can be used. Surgically bypassing the subclavian artery restores normal blood flow in the subclavian artery after the point of bypass [34–36]. Axillo-axillary bypass is reserved as an alternative method for high risk surgical patients. Surgical treatment for those patients with coexisting sever carotid stenosis is controversial and carotid endarterectamy is usually perform prior to any intervention for the subclavian artery. Patients who fail intervention and who are not candidates for surgical correction can be treated with antiplatelet therapy and anticoagulation. It is also important to emphasize risk factor modification as part of the therapy for secondary prevention. Smoking cessation, life style modification, antiplatelet therapy, blood pressure control, management of hyperlipidema, and diabetes control, all play an essential role in the conservatory management of the subclavian steal syndrome [11].

Conclusions

Patients with subclavian steal syndrome who present with minimal symptomatology or with non life threatening symptoms are recommended to be managed with a conservative approach. Recommendations include antiplatelet therapy, aggressive lipid management, and life style modifications. Management of comorbid conditions as hyperlipidema, hypertension, and diabetes mellitus is also important. This approach targets atherosclerosis and secondary prevention. Our patient presented with advance atherosclerosis disease, multiple lesions including subclavian, renal, and coronary arteries. She was treated with balloon angioplasty and stenting of the right coronary achieving resolution of cardiac symptoms and in a latter time balloon angioplasty and stenting of the right renal artery for kidney preservation blood pressure control. In this case, conservative management required angiographic intervention of other sites.

Statement

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