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A Comprehensive Review of Subclavian Steal Syndrome: Pathophysiology, Diagnosis and Management

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Abstract

Subclavian Steal Syndrome (SSS), or subclavian-vertebral artery steal syndrome, is a hemodynamic disorder caused by significant stenosis or occlusion of the subclavian artery, leading to the reversal of blood flow in the vertebral artery. This condition results in neurological and upper extremity symptoms such as dizziness, blurry vision, arm pain, and fainting, often triggered by physical activity. The left side is more commonly affected due to anatomical predispositions. SSS is notably prevalent in patients with coronary artery bypass grafting (CABG), where it can manifest as coronary-subclavian steal syndrome (CSSS), highlighting the need for heightened awareness among clinicians. The pathophysiology of SSS involves compensatory blood flow dynamics due to subclavian artery stenosis, primarily caused by atherosclerosis. Diagnostic approaches include Doppler Ultrasound, CT Angiography (CTA), Magnetic Resonance Angiography (MRA), and Conventional Angiography. Each modality provides critical insights into blood flow abnormalities and vessel structure, aiding in accurate diagnosis. Management of SSS varies based on symptom severity, ranging from conservative medical therapy for asymptomatic patients to endovascular interventions like angioplasty and stenting for symptomatic individuals. Surgical options such as carotid-subclavian bypass are considered for complex cases or failed endovascular treatments. Despite generally favorable outcomes, SSS is a marker of increased cardiovascular risk, necessitating aggressive secondary prevention strategies. This review underscores the importance of early detection and appropriate management of SSS to prevent severe complications and improve patient outcomes. Advances in diagnostic imaging and treatment techniques have significantly enhanced the prognosis for individuals with SSS.

Keywords: Subclavian Steal Syndrome (SSS); Diagnostic Approaches in SSS; Management Strategies in SSS; Coronary-Subclavian Steal Syndrome (CSSS)

Abbreviations: SSS: Subclavian Steal Syndrome; CSSS: Coronary-Subclavian Steal Syndrome; CABG: Coronary Artery Bypass Grafting; IMA: Internal Mammary Artery; PTFE: Polytetrafluoroethylene; MFA: Master of Fine Arts; MPH: Master of Public Health; CTA: CT Angiography; MRA: Magnetic Resonance Angiography; BES: Balloon Expandable Stent; PAD: Peripheral Arterial Disease

Introduction

Subclavian steal syndrome, now termed as subclavian-vertebral artery steal syndrome (SSS) is a hemodynamic condition characterized by blood flow reversal in the vertebral artery caused by significant stenosis or occlusion of the proximal subclavian artery or innominate artery [1]. The blood flow is redirected from the brain to the arm leading to neurological manifestations

including but not limited to, blurry vision, dizziness, pain/numbness/tingling in the arm during active motion, hearing loss and fainting [2]. SSS is generally seen on the left side of the body due to more acute proximity to the left subclavian artery causing accelerated turbulence and atherosclerosis. A coronary variant of subclavian-vertebral artery steal syndrome can occur as a complication following coronary artery bypass surgery, particularly when the arm on the same side as the internal mammary artery graft, used to bypass the narrowed or blocked coronary vessel, is involved [3].

The important of the CSSS is an underestimated and potentially easily missed cause of angina. The number of cases is increasing given the increased number of patients with CABG and greater life expectancy after CABG. A high degree of suspicion in patients with differential blood pressure readings in both arms and angina symptoms after CABG can lead to appropriate diagnosis and timely intervention [4].

Coronary-subclavian steal syndrome is an underestimated and easily overlooked complication of IMA grafting with potential disastrous outcome. The importance of an accurate diagnosis is often challenged by a variable clinical presentation and a low level of suspicion. Since endovascular treatment is easily accessible with excellent outcome, the importance of increasing awareness among (interventional) cardiologists needs to be emphasized [5].

Pathophysiology

The subclavian arteries, located just below the clavicles, are crucial for supplying blood to the upper extremities as well as contributing to the blood flow to the head and neck. The right subclavian artery originates from the brachiocephalic trunk, whereas the left subclavian artery stems directly from the aortic arch. These arteries extend laterally between the anterior and middle scalene muscles, and their distal limit is the lateral border of the first rib, where they transition into the axillary arteries. Key branches of the subclavian arteries include the internal thoracic artery, vertebral artery, costocervical trunk, thyrocervical trunk, and dorsal scapular artery. Despite their similarities, the left and right subclavian arteries have distinct origins; the left arises from the aorta, making it more prone to issues, while the right branches from the brachiocephalic artery. Both arteries give rise to the vertebral arteries, which are essential for supplying blood to the brain (Figure 1) [6,7].

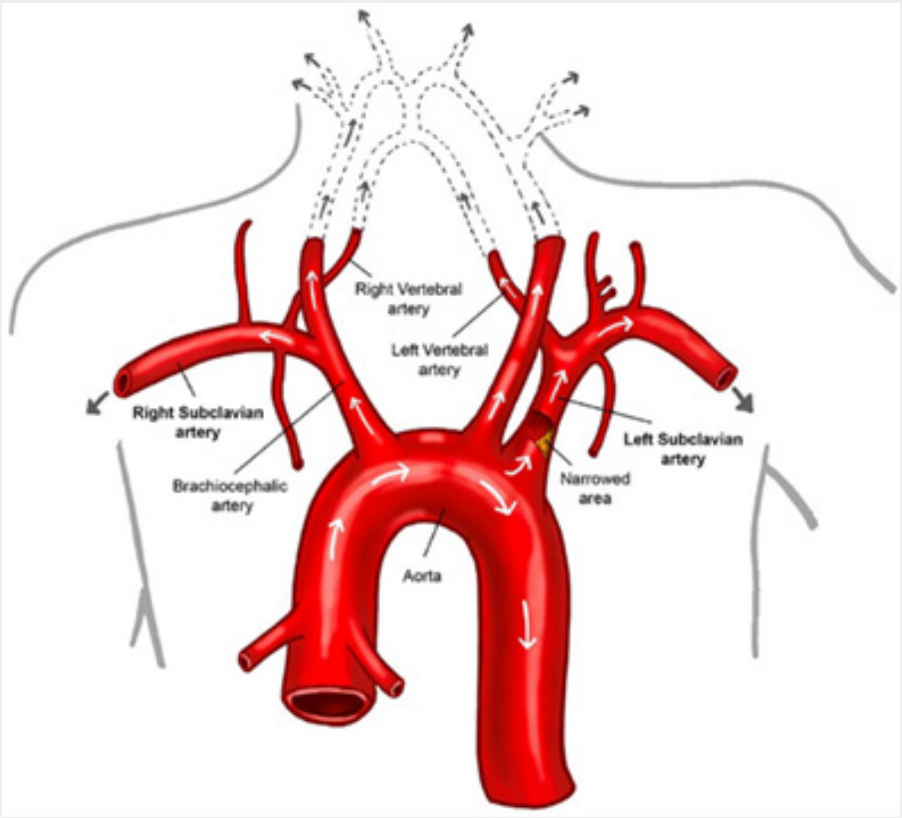

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Figure 1: Courtesy of Jessica Johnson, MFA, MPH (Icahn School of Medicine at Mt Sinai, New York, NY).

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Subclavian steal syndrome occurs when there is a narrowing or blockage in the subclavian artery near the origin of the vertebral artery, causing blood flow to reverse in the affected vertebral artery. This reversal happens because the pressure downstream of the blockage drops, prompting blood from the opposite vertebral artery to redirect through the basilar artery towards the affected subclavian artery. This altered flow pattern aims to compensate for reduced blood supply to the arm on the affected side. Consequently, blood intended for the brainstem and cerebellum may instead be diverted towards the arm, potentially leading to vertebrobasilar ischemia. This condition manifests with symptoms like dizziness, vertigo, or syncope, particularly during physical activity or sudden head movements [8].

Atherosclerosis represents the primary cause of subclavian artery stenosis, which triggers subclavian steal syndrome. Risk factors include smoking, hyperlipidemia, hypertension, diabetes mellitus, family history, and advancing age. While a stenosis greater than 50% commonly results in vertebral artery flow reversal, not all individuals exhibit symptoms. Symptoms typically occur when collateral blood supply fails to meet increased demands, such as during exercise or in the presence of arteriovenous fistula [8].

Various vascular conditions can also contribute to subclavian steal syndrome. Takayasu arteritis, more prevalent in younger patients, particularly in Asia, and giant cell arteritis are notable examples. Additionally, arterial thoracic outlet syndrome, involving compression of the subclavian artery due to anatomical variations, and congenital anomalies like isolated left subclavian artery in the setting of a right aortic arch are implicated. Furthermore, corrective surgeries such as coarctation of the aorta repair or tetralogy of Fallot correction, and the creation of hemodialysis arteriovenous access distal to a prevertebral stenosis, can precipitate or exacerbate symptoms due to altered flow dynamics [9]. The symptoms of subclavian steal syndrome can intensify with vigorous arm exercise or sudden head movements, exacerbating the retrograde flow in the vertebral artery by further decreasing pressure distal to the stenosis. The severity of resulting symptoms, ranging from mild dizziness to significant neurological deficits, depends on the adequacy of collateral circulation and the overall cardiovascular health of the patient.

Clinical Manifestations

Subclavian artery stenosis is asymptomatic in most patients. It is sometimes incidentally found when there is a blood pressure difference between the arms or on ultrasound testing of patients with coronary or carotid artery disease. It can be symptomatic in some patients and can present as arm pain, fatigue, numbness, or paresthesias. These symptoms are secondary to upper extremity ischemia during vigorous exercise. It can also present as a variety of neurological symptoms secondary to vertebrobasilar insufficiency, including dizziness, visual changes, syncope, vertigo, dysarthria, ataxia, tinnitus, or hearing loss. The neurological symptoms can be triggered by upper extremity exercise and also with head movements, with the rotation of face toward the opposite side. A detailed history can be useful in diagnosing subclavian steal [10-12].

Subclavian Steal Syndrome (SSS) occurs due to retrograde blood flow in the subclavian artery or the brachiocephalic trunk. This is often caused by stenosis or occlusion, resulting in various symptoms [10]. Studies have shown that SSS affects approximately 0.6% to 6.4% of the population, with 18% of those diagnosed with peripheral artery disease being affected; males are predominantly affected, and the age range is typically around 50 years [10]. Common manifestations include dizziness, which occurs because of impaired cerebral perfusion due to decreased blood flow in the vertebral arteries, which can compromise brain oxygenation [10,11]. Syncope, or fainting, can result from sudden drops in cerebral perfusion pressure, causing transient loss of consciousness [10,12]. Fatigue, reduced blood flow to the brain can lead to generalized fatigue as the body struggles to maintain adequate oxygenation throughout the body [2]. Claudication, pain or cramping in the arm during exertion is due to compromised blood flow to the upper extremities [10,11].

In a study by Song et al. (2021) and the Cleveland Clinic (2022), Subclavian Steal Syndrome (SSS) was defined as a condition resulting from the occlusion or severe stenosis of the subclavian artery, leading to reversed blood flow in the vertebral artery to compensate for reduced blood supply. This compensatory mechanism can redirect blood flow away from the brain and upper extremities, causing symptoms due to compromised cerebral perfusion and upper limb ischemia [10]. Symptoms related to brain involvement include dizziness, vertigo, and syncope, resulting from reduced blood flow to the posterior circulation of the brain [10,11]. Symptoms in the upper extremities, such as arm discomfort or weakness, claudication, pain, and fainting, occur because the subclavian artery is unable to adequately supply blood to the arm [10,11].

Diagnostic Approaches

Doppler Ultrasound

Doppler Ultrasound is a non-invasive and commonly used method for the initial assessment of Subclavian Steal Syndrome (SSS). This technique evaluates blood flow in the subclavian and vertebral arteries, helping detect abnormalities indicative of SSS. Doppler Ultrasound can identify differences in blood flow velocity and direction, which are critical for diagnosing retrograde flow in the vertebral artery, a hallmark of SSS [6].

CT Angiography (CTA)

CT Angiography (CTA) provides detailed images of blood vessels and is particularly valuable in visualizing the anatomy of the subclavian artery. It helps in detecting stenosis or occlusion by offering a comprehensive view of the blood vessel's structure. Radiologists use CTA to identify the precise location and severity

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of the blockage, as well as any signs of collateral circulation that might have developed as a compensatory mechanism. This detailed imaging aids in accurately diagnosing SSS and planning appropriate interventions [7].

Magnetic Resonance Angiography (MRA)

Magnetic Resonance Angiography (MRA) is useful for visualizing blood flow without using ionizing radiation, making it a safer alternative for certain patients. MRA provides high-resolution images that are effective in visualizing blood flow dynamics and the extent of vessel abnormalities. It is particularly beneficial for assessing the degree of stenosis or occlusion and the impact on blood flow to the brain and upper extremities. Radiologists look for similar diagnostic criteria in MRA as in CTA, such as the presence of retrograde flow in the vertebral artery and collateral circulation [8].

Conventional Angiography

Conventional Angiography is considered the gold standard for diagnosing SSS, despite being more invasive compared to other techniques. This method involves injecting a contrast dye into the blood vessels and taking X-ray images. Conventional Angiography offers the most detailed and accurate visualization of blood vessels, allowing for precise assessment of stenosis or occlusion and the extent of collateral circulation. Radiologists use this technique to confirm the diagnosis of SSS by identifying the retrograde flow in the vertebral artery and evaluating the overall blood supply to the brain and upper extremities [9].

Epidemiology

Subclavian-vertebral artery steal syndrome (SSS) is an uncommon vascular condition and the prevalence of this disorder varies across various populations. Research indicates the precise incidence and prevalence is unknown. However, most literature reports estimate SSS prevalence between 0.6% to 6.4% [2]. Approximately 18% of people diagnosed with peripheral arterial disease (PAD) are also considered to have SSS [13]. This condition is more common in males than females and predominantly affects individuals over the age of 55 [2,3]. Risk factors include hypertension, hyperlipidemia, diabetes, and smoking, all of which contribute to the development of atherosclerosis, the leading cause of the condition.

Management Strategies

The management of subclavian steal syndrome or subclavian artery steal syndrome (SSS) depends on whether the condition is symptomatic or asymptomatic [3]. Only a tiny percentage of patients with SSS require intervention. The severity of the blood pressure differences between the arms predicts the need for intervention [9].

In asymptomatic SSS patients with isolated symptoms, conservative management is recommended irrespective of the severity of vertebral artery steal [9]. There is no decisive medical therapy for the management of SSS. The current recommended medical therapy is directed toward controlling atherosclerotic and peripheral vascular disease risk factors, which include aspirin and high-intensity statin therapy [14]. Additionally, anti-anginal therapy, beta-blockers, calcium channel blockers, and nitrates are used as temporizing agents in patients with symptomatic coronary-subclavian steal syndrome (CSSS) [14]. In patients with subclavian artery stenosis due to subclavian artery atherosclerosis, the tight control of hypertension, diabetes, and dyslipidemia and lifestyle modification, including smoking cessation and periodic monitoring with ultrasound, is beneficial in these patient populations [3,9].

In symptomatic SSS patients, open surgical bypass, especially extra-anatomic revascularization such as carotid-subclavian bypass or carotid transposition, which has an excellent safety profile and has been shown to improve symptom severity and mortality, can be an option [3,9]. Minimal-risk endovascular therapy with angioplasty and stenting has changed SSS management with similar outcomes as open surgical methods [9,15]. Endovascular revascularization involving angioplasty and stenting is the first-line therapy due to its long-term efficacy profile, cost-effectiveness, and improvement in morbidity and mortality. However, open revascularization can be considered after failed endovascular therapy [14].

Surgical treatments in Subclavian steal syndrome are reserved for patients with severe symptoms or failed endovascular attempts like angioplasty and stenting. Major surgery may involve carotid-subclavian bypass and subclavian-carotid transposition as the options. In a carotid-subclavian bypass, an end-to-side anastomosis between the common carotid artery and the subclavian artery distal to the stenosis using PTFE (Polytetrafluoroethylene) or Dacron grafts. This procedure is performed through a supraclavicular incision rather than major thoracic incisions, reducing complications associated with more invasive approaches. Subclavian-carotid transposition, however, involves changing the direction of blood flow in the subclavian artery towards the carotid artery thus necessitating proximal dissection of this vessel [11,16,17].

Both procedures have been found to have excellent long-term patency and low rates of morbidity and mortality, rendering them suitable for patients with extensive occlusions or complex anatomical considerations. Successful outcomes require preoperative planning with detailed imaging and careful intraoperative management including systemic heparinization when considering shunting in contralateral cases of carotid disease. Symptom resolution monitoring postoperatively and provision of long-term anti-platelet therapy are some aspects of care that are needed after surgery [3,18]. Combining endovascular and open surgical techniques, hybrid procedures are fast gaining popularity as they allow for individualized revascularization

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strategies. Intraoperative imaging which includes angiography and ultrasound has become an indispensable component of such procedures that enables real-time assessment and modification of revascularization. Besides there is growing attention on preserving the internal mammary artery among patients who have undergone coronary artery bypass graft to prevent coronary-subclavian steal. These advances along with improved perioperative management and personalized treatment plans have greatly improved the surgical management of SSS, especially in instances where endovascular approaches have failed or are not possible [19].

Patient Outcomes

Subclavian Steal Syndrome (SSS) has in general a good prognosis as many patients are asymptomatic, or their symptoms are mild and non-disabling they don´t need any intervention [3]. The presence of subclavian stenosis is associated with increased total mortality and cardiovascular disease mortality and with an increased risk of cerebrovascular ischemic events related to progressive carotid stenosis and compromised collateral pathways. It is also a marker of atherosclerosis; it can indicate the risk for future events. Therefore, subclavian stenosis is a marker of cardiovascular risk and identifies a population that will benefit from aggressive secondary prevention [18].

The largest retrospective study comprised 167 consecutive patients undergoing coronary artery bypass graft (CABG) with left subclavian artery stenosis or occlusion demonstrated that endovascular revascularization with a balloon expandable stent (BES) before using the left internal mammary artery for CABG was safe and effective with a low incidence of peri-procedure complications and in-stent restenosis. The incidences from the time of stenting to 30 days afterward were 0.6% for death, 1.8% for stroke, and 1.8% for myocardial infarction. Patency rates were 95.7%, 93.8%, 86.5%, and 75.2% at 1, 2, 5, and 10 years, respectively. The event-free survival rates (all deaths, all strokes, transient ischemic attacks, non-fatal myocardial infarctions) were 97%, 95.1%, 88.2%, and 76.2% at 1, 2, 5, and 10 years, respectively [20]. Symptoms due to significant recurrent stenosis or obstruction occur in approximately 10% of the patients and are typically treated with repeat angioplasty and stent placement; however, surgery may be required in up to 5% of the patients [21].

Conclusion

Subclavian Steal Syndrome (SSS), now referred to as subclavian-vertebral artery steal syndrome, is a significant but often under-recognized condition characterized by retrograde blood flow in the vertebral artery due to proximal subclavian artery stenosis or occlusion. This hemodynamic alteration results in a range of neurological and upper extremity symptoms, which can be exacerbated by physical activity or sudden head movements. The prevalence of SSS varies, affecting up to 6.4% of the population, particularly in males over 55 years old with risk factors such as hypertension, hyperlipidemia, diabetes, and smoking. Diagnosing SSS involves various imaging modalities, each offering unique advantages. Doppler Ultrasound is a non-invasive, initial assessment tool, while CT Angiography (CTA) and Magnetic Resonance Angiography (MRA) provide detailed images of blood vessels and flow dynamics.

i. Conventional Angiography remains the gold standard, offering the most precise visualization for confirming SSS. Management of SSS depends on symptom severity and includes conservative medical therapy for asymptomatic cases and endovascular or surgical interventions for symptomatic patients. Endovascular treatments, such as angioplasty and stenting, are preferred for their efficacy and lower risk, while open surgical procedures, such as carotid-subclavian bypass and subclavian-carotid transposition, are reserved for complex cases or failed endovascular attempts. Patient outcomes for SSS are generally favorable, especially with timely diagnosis and appropriate intervention. While many patients remain asymptomatic or experience mild symptoms, the presence of subclavian stenosis is a marker for increased cardiovascular risk, necessitating aggressive secondary prevention strategies. Advances in both endovascular and surgical techniques have significantly improved the prognosis for patients with SSS, highlighting the importance of heightened awareness and early detection.

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