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Review Article

Superior vena cava syndrome: a comprehensive review

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ABSTRACT

Superior vena cava syndrome (SVCS) is a condition resulting from obstruction or compression of the superior vena cava, leading to venous congestion of the head, neck, and upper extremities. This review provides a comprehensive discussion on the etiology, pathophysiology, clinical presentation, diagnostic approach, and management strategies for SVCS. Advances in imaging and endovascular interventions have improved the diagnosis and treatment of SVCS. Although malignancies remain the leading cause, benign etiologies have gained significance in the modern era. Despite these advancements, the treatment of SVCS remains challenging, requiring a multidisciplinary approach.

Keywords: Superior vena cava syndrome, Malignancy, Endovascular stenting, Radiotherapy, Oncology

INTRODUCTION

The superior vena cava syndrome (SVCS) is a rare clinical condition caused by compression of the blood vessels. Its occurrence is uncommon globally, and only a few case reports have been published. In 1757, William Hunter was the first to describe it as a complication of a syphilitic saccular aneurysm.[1] The diagnosis and treatment of SVCS have advanced, but there is still a lack of clinical trials to provide guidance for diagnosis and treatment protocols.

ANATOMY AND PATHOPHYSIOLOGY

SVC is a thin-walled, low-pressure vein formed by the right and left brachiocephalic veins providing venous drainage to the head, neck, upper limbs and upper thorax. The azygous vein is the major tributary of SVC, which passes along the thoracic vertebrae's anterior border to the tracheal carina's level and drains posteriorly into the SVC. Located in the Right mediastinum, it is prone to compression via abnormalities of the trachea, Right bronchus, aorta, pulmonary artery or perihilar and paratracheal lymph nodes. When there is increased resistance to the blood flow of SVC, venal enlargement occurs proximal to the site of obstruction and collateral circulation through the azygous vein or inferior vena cava occurs with the dilatation of collaterals over time. SVCS can be more severe if the obstruction is below the azygous vein as a primary bypass circulation is obstructed. The rapidity of onset in obstruction also influences the severity as the collaterals take weeks to dilate and provide good diversion and blood flow.

ETIOLOGY

Before the antibiotic era, many cases of SVCS were attributed to the syphilitic aortic aneurysm and tubercular mediastinal lymphadenitis. In the present era, malignancy has been associated with 70% of the cases, and benign causes account for the rest.[2] In malignancy, lung malignancy (75%) is the most common, non-Hodgkin lymphoma (15%), others like breast cancer, oesophageal cancer, germ cell tumours, and thymoma make up the rest.

SVCS incidence is higher in adult males as lung carcinoma is more common in males compared to females. Benign causes of superior vena cava obstruction include intravascular devices (30%), cardiac devices, mediastinal fibrosis, benign mediastinal tumours, vascular diseases and infections. In 30% of patients who have pacemaker

leads, there can be clinically silent thrombosis. However only in 0.1 to 3.3 of these patients does SVC syndrome occur, and this has been attributed to the fibrin deposition on the surface of the leads and incorporation to intima followed by vessel wall inflammation, fibrosis, thrombus formation and stenosis.[2]

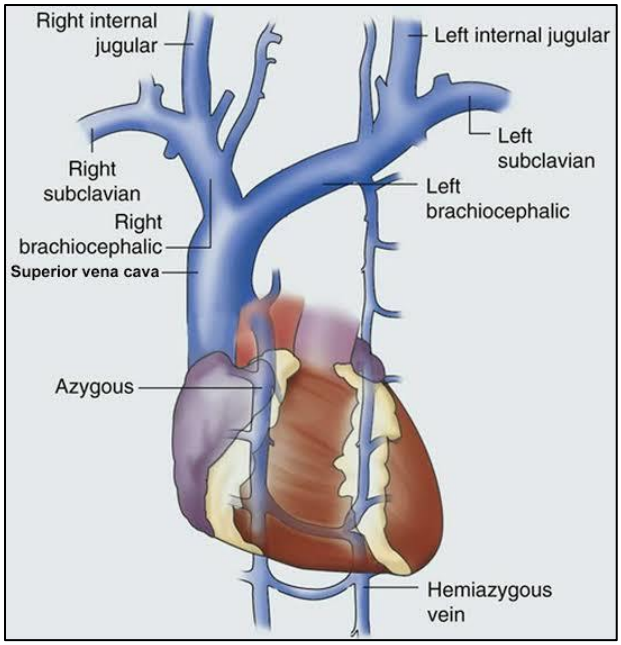

Figure 1: Venous anatomy of the superior vena cava system.

This schematic diagram illustrates the anatomy of the superior vena cava (SVC) and its major tributaries. The SVC is formed by the convergence of the right and left brachiocephalic veins and drains venous blood from the upper extremities, head, and neck into the right atrium. The azygos and hemiazygos veins provide alternative drainage pathways, particularly important in superior vena cava syndrome (SVCS), where obstruction of the SVC can lead to venous congestion and collateral circulation formation. This diagram highlights key venous structures involved in systemic venous return and potential compensatory pathways in SVC obstruction.

CLINICAL PRESENTATION

The clinical presentation varies depending on the location, rapidity of onset, obstruction severity, and collateral circulation. A patient presenting to the emergency room will most likely have breathlessness. Dyspnea is the presenting feature in about 63% of cases of SVCS.[3]

When the underlying aetiology is malignancy compared to benign, the patient is likelier to have an associated cough with dyspnea. Other common presentations include facial and neck swelling (80%), distended neck and chest veins (38%), watering of the eyes, and dizziness when leaning forward.[4] There can be a classical history of worsening of symptoms in the supine position. A malignant SVC obstruction due to an aggressive tumour can result in acute and life-threatening symptoms of cerebral, laryngeal and pharyngeal edema like stridor, confusion and obtundation due to lack of time for developing and enlarging collateral channels. There can also be symptoms of associated malignancies like stridor, haemoptysis, B symptoms in Hodgkins's lymphoma. Physical examination findings include facial and neck edema (most common), distended neck and chest veins, extremity swelling, cyanosis, plethora, altered mental status, and papilledema. Hemodynamic compromise is rare and is seen when SVC obstruction is associated with an underlying malignancy causing compression of cardiac chambers. Acute mortality owing to SVCS is rare (0.3%), while median life expectancy in SVCS secondary to malignancy is only six months.[5]

There is no standardised grading for the evaluation of SVCS. A study group from Yale University has proposed a recent classification for the severity of SVCS, which does help in the approach and triage of these patients.[6] It includes five grades: asymptomatic (grade 0), mild symptoms (grade 1), moderate symptoms (grade 2), life-threatening symptoms (grade 3,4), and fatal/death (grade 5). Urgent treatment is indicated in grades 3,4. Kishi score is a rating system which can guide decisions of stent therapy, grade 4 or more is an indication of endovascular intervention.[7]

DIAGNOSTIC EVALUATION

Superior vena cava syndrome is a clinical diagnosis and confirmed with the imaging studies. Common imaging includes a chest radiograph, which can be abnormal in 84% of cases, with common findings like mediastinal widening and pleural effusions.[7] Contrast-enhanced computed tomography can provide the level and extent of the blockage and the evaluation of collaterals. The presence of collaterals on CECT is an accurate predictor of clinically relevant and symptomatic SVC syndrome.[8] It is highly sensitive and specific in diagnosing SVCS and can also help in identifying the cause of obstruction in most cases.

A duplex scan of the upper limb can evaluate the extent of thrombosis in the subclavian and axillary veins and identify access sites for endovascular interventions. Digital subtraction venography is considered a gold standard for the evaluation of SVCS.[9]

It can help in identifying the collaterals, and severity of obstruction and also helps the interventionalist develop a strategy for definitive revascularisation. A significant drawback of duplex scan is the inability to evaluate the cause of extrinsic SVC obstruction.MR Venography can be equally sensitive in patients allergic to IV Contrast or renal failure. Along with the above investigations, a complete diagnostic workup has to be done to delineate the suspected underlying malignancy.

MANAGEMENT

The standard of care in the treatment of SVC syndrome has evolved. Once considered a medical emergency, it now rarely requires emergent treatment modalities in the ED. The approach has become multidisciplinary with physicians, oncologists, pulmonologists, radiologists, vascular surgeons and endovascular specialists. In the ED, initial management includes head end elevation, supplemental oxygen, loop and osmotic diuretics, and potent glucocorticoids. There is lacking efficacy in the use of steroids and diuretics in initial management and steroid use can hinder tissue diagnosis, especially in lymphomas.[10]

Anticoagulation is only indicated currently in cases with demonstrated thrombosis to prevent embolic consequences.[11] Life-threatening situations should be dealt with immediately, with the stabilisation of the patient quickly followed by endovascular recanalization procedures to provide immediate relief of symptoms.[12] In SVCS, the evidence guiding decisions are derived from case series as the data from randomised clinical trials are scarce. The choice of definitive as opposed to palliative care is to be made depending on the underlying etiology, type and stage of malignancy. Treatment modalities include radiotherapy, chemotherapy, and endovascular interventions.

RADIATION AND CHEMOTHERAPY

Radiotherapy is an effective treatment of SVCS due to malignancy. Rowell and Gleeson documented that radiotherapy provided overall relief of symptoms in one-third of SVCS caused by small-cell lung cancer and two-thirds of cases of non-small cell lung cancer.[13] RT may provide a rapid relief of symptoms as early as 72 hours of initial therapy.[14] The use of initial RTs has decreased in recent times as it may hinder the further histological diagnosis of underlying malignancy; often, the benefits are temporary, with a significant fraction reexperiencing the symptoms. Chemotherapy is often used in the treatment of SVCS due to lymphomas, small-cell lung cancer, and germ-cell tumours, which are chemo-sensitive and can lead to a high rate of response and quick tumour shrinkage.[15]

ENDOVASCULAR INTERVENTIONS

A rapid relief of symptoms by restoring the venous flow is now possible with the advent of endovascular therapies in SVCS. The relief of symptoms can be observed as early as 24 hours following the procedure and contributes to overall improvement in morbidity and mortality.[16,17] The procedures commonly used are stenting, luminal angioplasty and intravascular thrombolysis. The effectiveness and outcome of endovascular procedures are better in benign causes of SVCS compared to malignant causes.[18] No randomised trials exist, but extensive data strongly supports stents as the most effective treatment for SVCS due to malignancy.

CONCLUSION

Even though SVCS affects a small fraction of patients, the effects can be devastating, causing significant morbidity. Although there is a larger incidence of benign causes of SVCS in the current era, malignancy remains the most common cause. Endovascular interventions have replaced traditional radiotherapy as the first line of management. Currently, there are no validated clinical trials for the management of SVCS, and there is a need for standardisation of the treatment strategies to guide the subsequent management.

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