SVC Syndrome due to Mediastinal Malignancy

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Introduction

The Superior Vena Cava (SVC) is a 7 cm long, 2 cm wide, thin-walled, and low-pressure valveless vein responsible for the venous return from the head, neck, upper extremities, and torso to the heart. It is formed by the union of the internal jugular and subclavian veins coursing down to through the mediastinum into the right atrium. Pathologies of the surrounding structures leads to extrinsic compression and obstruction. Compression of the SVC may lead to facial and upper extremity edema, shortness of breath, facial plethora, and distended neck and chest veins. Compilation of these symptoms is known as SVC syndrome.

This case depicts a systemic approach used with from patient presentation, diagnostic tests used and treatment.

Case Presentation

A 65yo African American man presented to ED with bilateral upper extremity edema since last 3 days and acute chronic left knee pain.

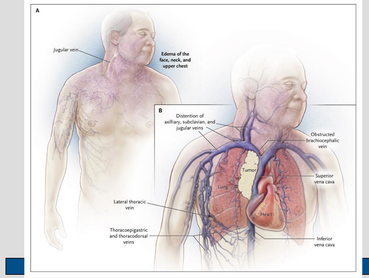
Past medical history significant for type 2 diabetes mellitus, tobacco smoker, hyperlipidemia, chronic pain syndrome, lumbar spinal stenosis, and Parkinson's disease, which was diagnosed 6 months prior.

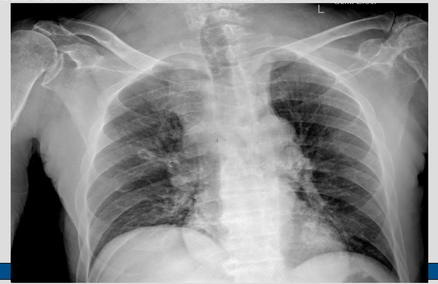
Home medications include aspirin, atorvastatin, baclofen, benzonatate, gabapentin, Levaquin, and Percocet: taking since last few months for generalized pain.

Physical exam in ED noted 2+ edema of the upper extremities extending from hands to the axilla, 2+ edema of bilateral lower extremities, and distended neck veins with veins overlying the anterior chest wall.

CTA of the chest with contrast showed pulmonary nodules and marked upper mediastinal lymphadenopathy with severe narrowing of the SVC and opacification of extensive venous collaterals in the right shoulder and right hemithorax.

CTC spine without contrast and CT Head without contrast were unremarkable.





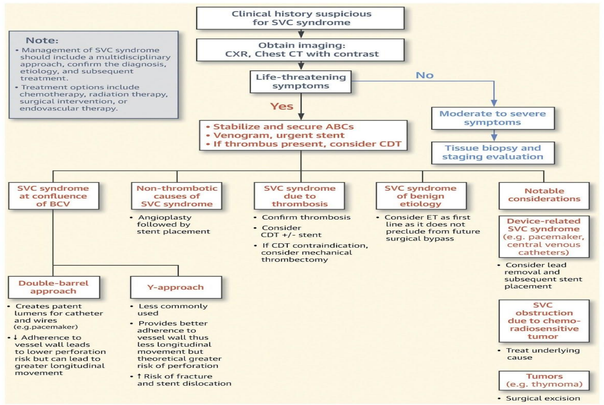
Discussion

Approximately 60% of SVCS cases are the result of mediastinal malignancies. The remaining 40% cases are caused by thrombus formation associated with intravascular devices such as catheters and pacemakers. Malignancies cause SVCS via obstruction from thrombosis, direct invasion of tumor cells inside the vessel, or mass effect causing external compression.

SVCS is diagnosed based on several clinical manifestations, including cough, dyspnea, dysphagia, and edema or discoloration of the neck, face, and upper extremities. When these vessels dilate, it manifests as distension of the superficial veins in the chest wall. These patients frequently complain of a sense of head "fullness" and dizziness aggravated by bending forward. Though SVC syndrome is a clinical diagnosis, plain chest radiography, contrast-enhanced computed tomography (CECT), and venography are used for confirmation. Treatment is then tailored to the findings.

Yu et al. proposed a classification system in 2008 to provide an objective and universal language to determine the severity of the syndrome and therefore help determine the role of intervention. The system grades the severity on a scale of 0-5. A score of 0 indicates asymptomatic or incidental finding. With the severity of symptoms, the score increases to such a score of 4 or 5 indicates life-threatening symptoms with significant cerebral edema, significant laryngeal edema, or significant hemodynamic compromise or death. Azizi et al. have taken these criteria and proposed a management algorithm that is based on the severity of the symptoms and identification of the underlying malignancy (Central Illustration). SVCS treatment targets symptom relief by attempting to alleviate the compression.

CENTRAL ILLUSTRATION: Proposed Management Algorithm of SVC Syndrome Based on Severity of Symptoms and Common Etiologies



Azizi, A.H. et al. J Am Coll Cardiol Intv. 2020;13(24):2896–910.

Treatment

There are no gold standard guidelines for treatment. The mainstay of treatment is treat the cause of obstruction. Malignancies and thrombotic events are the most common causes of SVC Syndrome. As done in the case, the standard of diagnosis is CT imaging to differentiate thrombosis vs compression. Treatment centers around treating the cause of the compression: malignancy, infectious causes, or aneurysm. Patient was started on dexamethasone 4 days. Patient then underwent an EBUS-guided FNA of the right mediastinal mass which was positive for malignancy; poorly differentiated squamous cell carcinoma also present in the right bronchus. The patient then started palliative radiation therapy (RT) the following day. Stenting held in setting of initiation of RT.

Different approaches: By shrinking the mass it should relieve the compression. Stenting, another viable option to relieve venous congestion and edema from compression. As all problems in medicine a systematic approach is utilized: clinical presentation, diagnostic imaging and labs following treatment based on those findings. As 60% are a result of malignancies chemotherapy with or without radiation therapy is common; surgical bypass, or endovascular techniques such as angioplasty, stenting, and catheter-based thrombus removal are other treatment options depending on the case. Additional general measures include diuretics, and corticosteroids. The use of both glucocorticoids and loop diuretics are found to demonstrate no benefit with lack of supporting data.

References

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