Case Report

A case of superior vena cava syndrome

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Abstract

Superior Vena Cava Syndrome is the clinical manifestation of the Superior Vena Cava obstruction, with severe reduction in venous return from the head, neck and upper extremities. More than 80% cases of SVCS is nearly attributable to advanced malignancy, most commonly lung cancer. SVC syndrome is characterized by congestion and swelling of the face and upper thorax, with distended superficial chest veins. The most severe complications of SVC syndrome include Glottic edema and venous thrombosis in the central nervous system. We represent a case SVC syndrome due to Small Cell Cancer of the lung.

Key words

Superior vena cava syndrome, Small cell cancer of lung, Radiotherapy, Chemotherapy.

Introduction

Superior vena cava (SVC) syndrome is a collection of clinical signs and symptoms resulting from either partial or complete obstruction of blood flow through the SVC. This obstruction is most commonly a result of thrombus formation or tumor infiltration of the vessel wall.

The superior vena cava is formed by the junction of the left and right innominate (brachiocephalic) veins and is tasked with returning blood from the head, neck, upper extremities, and torso back to the heart.

Today, this syndrome is most commonly seen secondary to malignancy, although there has been a more recent rise in benign etiologies.

The resulting venous congestion produces a clinical scenario relating to increased upper body venous pressures.

The most common signs and symptoms include face or neck swelling, upper extremity swelling, dyspnea, cough, and dilated chest vein collaterals [1, 2, 3].

Etiology

Superior vena cava syndrome (SVCS) is the clinical manifestation of superior vena cava (SVC) obstruction, with severe reduction in venous return from the head, neck, and upper extremities.

Malignant tumors, such as lung cancer, lymphoma, and metastatic tumors, are responsible for the majority of SVCS cases [4].

With the expanding use of intravascular devices (e.g., permanent central venous access catheters, pacemaker/ defibrillator leads), the prevalence of benign causes of SVCS is now increasing, accounting for at least 40% of cases [4].

Lung cancer, particularly of small-cell and squamous cell histologies, accounts for ~85% of all cases of malignant origin.

In young adults, malignant lymphoma is a leading cause of SVCS. Hodgkin's lymphoma involves the mediastinum more commonly than other lymphomas but rarely causes SVCS.

Other causes include benign tumors, aortic aneurysm, thyromegaly, thrombosis, and fibrosing mediastinitis from prior irradiation, histoplasmosis, or Behçet's syndrome. SVCS as the initial manifestation of Behçet's syndrome may be due to inflammation of the SVC associated with thrombosis [4].

Pathophysiology

Obstruction of blood flow through the SVC can be caused by extrinsic compression of the vessel, or by thrombus within the SVC. If this blockage is subacute or chronic, then venous collaterals will develop, allowing for alternative pathways back to the right atrium. Collateral veins may form off the azygous, internal mammary, lateral thoracic and esophageal venous systems. But even with development of these collaterals central venous pressures are typically elevated, producing the signs and symptoms of SVC syndrome. But if complete obstruction happens acutely (rapidly growing tumor) there may not be enough time for collaterals to develop and signs and symptoms will rapidly progress [9].

Clinical presentation and grading system

Clinical presentation varies depending on the severity, location, and rapidity of onset of obstruction and establishment of collateral veins [5]. The most common presenting symptoms include facial and neck edema, distended neck and chest veins, watering eyes, and dizziness particularly when leaning forward. Patients may also present with symptoms that are neurological (headache, blurry vision, decreased level of consciousness), laryngopharyngeal (tongue swelling, dyspnea), upper extremities (edema), and facial (conjunctival/ periorbital edema) [6]. Patients also typically describe worsening of their symptoms in the supine position. Rarely, proximal esophageal varices may be seen [7].

Case report

A 63 year old male patient admitted to the hospital with the complaints of puffiness of face since one month, breathlessness since 5 days, swelling of upper limbs since 2 days, hoarseness of voice since 2 days. No history of chest pain/ paroxysmal nocturnal dyspnea/ palpitations/ hemoptysis/ oliguria/ abdominal distension/ pedal edema/ syncope/ choking/ fever.

Past history

No history of similar complaints in the past. Not a known case of Diabetes Mellitus/ Hypertension/ Coronary Artery Disease/ Bronchial Asthma/ Tuberculosis/ Thyroid disorder. No History of any drug or food Allergies. No history of previous blood

transfusions. No History of any surgery in the past.

Addictions: Smoker since 20 years, he smokes more than 20 cigarettes per day (20 pack years).

General Examination

Patient was conscious, coherent, oriented to time, place and person. Moderately built, moderately nourished. Puffiness of face was present. No Pallor/ Icterus/ Cyanosis. Posterior Cervical Lymphadenopathy present. No Thyromegaly. No prominent Jugular venous pulsations. Diffuse edema of both upper limbs was present. There were multiple dilated veins seen over the anterior part of the chest wall on both sides in the infraclavicular regions. Grade 3 clubbing is present. No thickened nerves. No abdominal distension or pedal edema.

Vitals: Temperature - 98.2 F, Pulse- 96/min, regular, Blood pressure - 140/90 mm of Hg in both limbs supine position, Respiratory rate- 32/min, Abdomino thoracic type, Saturation - 90% on Room air, 97% with 2 litres of oxygen.

Respiratory system examination

Inspection: Use of accessory muscles of respiration is seen, Barrel shaped chest is seen. There were multiple dilated veins seen over the anterior part of the chest wall.

Palpation: Apical impulse felt in the left 5th intercostal space in mid-clavicular line. Decreased movements on the right side of the chest.

Percussion: Stony dull note heard on right side in 4-7th intercostal spaces and lower interscapular and infra scapular areas. Resonant heard in all other areas.

Auscultation: Absent breath sounds in infra axillary and lower interscapular and infra scapular areas. Normal vesicular breath sounds heard in other areas.

Other systems examination

Cardiovascular system: S1, S2 heard.

Gastrointestinal system:

Oral cavity: Poor oral hygiene with nicotine staining present on the teeth.

Per Abdomen: Soft, non tender, no organomegaly.

Central nervous system: No focal neurological deficit.

Investigations

Complete blood picture

Hb: 12.1 gms/dl, WBC: 8000 cells/cu.mm, Platelet count: 3.3 lakh/cu.mm.

ESR: 60 mm - 1st hour,

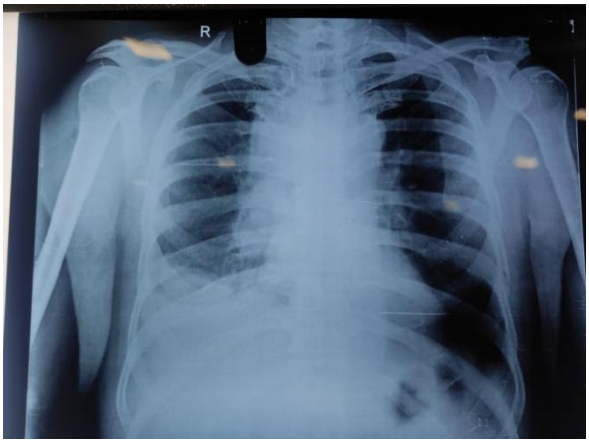
RFT: Blood urea – 14 mg/dl, Serum creatinine: 0.9 mg/dl,

LFT: Total Bilirubin: 0.3 mg/dl, Direct Bilirubin: 0.1 mg/dl, Indirect Bilirubin: 0.2

SGOT/AST: 30 U/L, SGPT/ALT: 26U/L, ALP: 105 U/L, Total Protein: 7.1 g/dl, Albumin: 3.8 g/dl, Globulin: 3.3 g/dl, A/G ratio: 1.2.

Chest X ray PA view: Showing evidence of large heterogenous lymph node. Enlargement noted in right paratracheal region, which appears to encase and compress the supraazygous part of superior vena cava, with right mild pleural effusion (**Figure – 1**).

Figure – 1: Chest X-ray PA view.



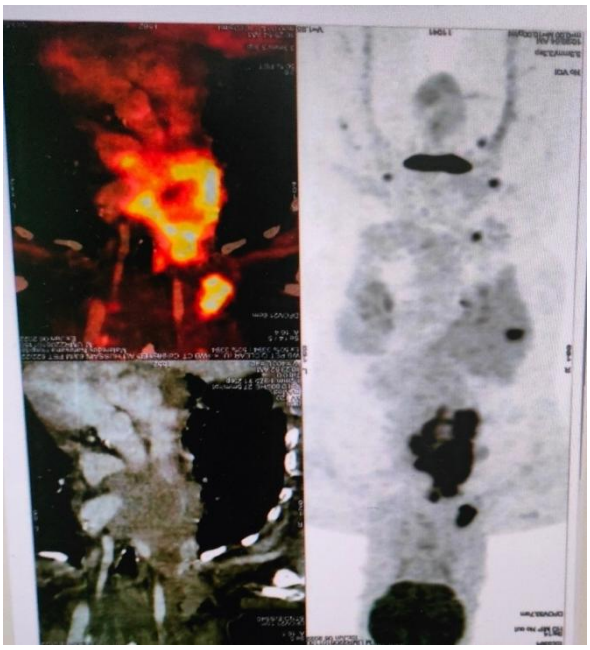
2D ECHO: EF: 65%, Normal sized cardiac chambers. No LV RWMA. Normal LV function. Grade 1 LV diastolic dysfunction present. Mild MR, Trivial MR. No Pericardial effusion /clots.

Whole body F-18 FDG PET-CT report

Metabolically active soft tissue density lesion in the right hilar region extending into right lower lobe (**Figure – 2**).

Intense FDG concentration noted in heterogeneously enhancing conglomerate nodal mass in the anterosuperior mediastinum with areas of photopenia extending into the subcarinal region comprising superior mediastinal (1), Right upper and lower paratracheal (2R, 4R), Prevascular (3a), Subcarinal (7), measuring 9 cm (AP)*7cm(ML)*9.4cm(CC) with max SUV of 11. The mass is encasing the SVC and left brachiocephalic vein abutting aorta.

Figure – 2: Whole body F-18 FDG PET-CT.



Metabolically active enlarged right supraclavicular lymph nodes.
Metabolically quiescent right sided pleural effusion with passive collapse of the right lower lobe.
Metabolically active hypodense lesion in the right lower lobe.
Metabolically active lytic skeletal lesions in the pelvis.

Histopathology report
Immunohistochemistry:
CK (Epithelial marker) - Positive
Synaptophysin - Positive
TTF-1 - Positive
INSM-1 - Patchy positive
Diagnosis: Small cell cancer of lung.

Treatment given
Inj. Dexa 8 mg IV TID
Inj. Lasix 40 mg IV BD
Palliative radiotherapy
Palliative chemotherapy with Carboplatin and Etoposide.

Discussion

Signs and symptoms are presented in order of most common to least common. Adapted from Yu JB, et al. [8] as per Table – 1.

Table – 1: Signs and Symptoms.

Signs and Symptoms	Incidence (%)
Facial edema	60–100
Non pulsatile distended neck veins	27–86
Distended chest veins	38–67
Dyspnea and cough	23–70
Arm edema	14–75
Hoarseness and/or stridor	0–20
Syncope and/or headache	6–13
Confusion, obtundation	0–5

The severity of SVC syndrome based on the clinical presentation has been described by Yu JB, et al. [8]. The scoring system proposed by these authors, with ranges from grade 0 to grade 5, can be helpful in the diagnostic approach and determination of treatment as per **Table – 2**.

Table – 2: Grading of SVC Syndrome per Yu JB, et al. [8].

Grade	Finding(s)
0	Asymptomatic: SVC on imaging without symptoms
1	Mild: edema of head or neck
2	Moderate: edema in head or neck with functional impairment
3	Severe: mild or moderate cerebral edema/laryngeal edema, or diminished cardiac reserve
4	Life-threatening: significant cerebral edema, laryngeal edema, hemodynamic compromise
5	Fatal: death

SVC = superior vena cava.

Patterns of obstruction and collateral pathways

In SVC obstruction, the flow of blood is diverted to the right atrium through a collateral venous network, which can take several weeks to accommodate the usual blood flow of the SVC. The severity of presentation of SVC syndrome is inversely related to the development of these collateral veins and the rapidity with which SVC obstruction develops [9]. SVC obstruction typically causes the venous pressure to rise as high as 20 to 40 mm Hg proximal to the obstruction. This increased venous pressure produces symptoms of facial, neck, and chest wall edema [9]. There are 4 main collateral pathways: 1) the azygous venous system, which is the largest and consists of azygous, hemiazygos, intercostal, and lumbar veins; 2) the internal mammary pathway; 3) the lateral thoracic pathway; and 4) the vertebral venous pathway.

The Azygous system consists of the azygos, hemiazygos, intercostal, and lumbar veins. Of note, superior vena cava (SVC) obstruction from infra-azygous occlusion utilizes this pathway, leading to milder symptoms. The internal mammary pathway consists of internal thoracic, superior and inferior epigastric, and superficial thoracic veins (not labelled). The lateral thoracic pathway (not labelled) consists of small veins mainly including the lateral thoracic, thoracoepigastric, superficial circumflex, and femoral veins. The vertebral pathway collateralizes to the azygos and internal mammary pathways through the vertebral, intercostal, and lumbar veins.

Anatomic classification of SVC obstruction includes 3 levels of obstruction: obstruction of upper SVC proximal to the azygous vein, obstruction at the level of the azygos vein, and obstruction distal to the azygos vein. An obstruction cephalad to the azygos vein causes the blood to return to right atrium through the azygous system and intercostal veins into the SVC. In SVC obstruction at the level of azygos, blood cannot re-enter the SVC through the azygos system and is forced to utilize other collateral veins, leading into the inferior vena cava (IVC) and from there into the right atrium. In SVC obstruction below the level of azygos vein, blood will be redirected via a robust azygos and hemi-azygos system in a retrograde manner ultimately to the IVC, hence causing less severe symptoms [10]. However, in many cases, the brachiocephalic veins (or portions thereof) are also involved, such that the collateral veins formed are multiple and may include any of the previously listed pathways [11].

Diagnostic approach

The diagnosis of SVC syndrome is based on the clinical presentation and advanced imaging. Imaging modalities include chest radiography, contrast-enhanced computed tomography (CT) scanning, duplex ultrasound, conventional catheter-based digital subtraction venography, and magnetic resonance venography.

Contrast-enhanced CT scanning provides optimal visualization of the SVC and can localize the extent of venous blockage, differentiate thrombosis from extrinsic compression, and identify collateral pathways [12].

Digital subtraction venography is the gold standard for evaluation of SVC obstruction, including the presence of thrombus. Venography identifies collateral venous pathways and defines the severity of obstruction, and enables the interventionalist to develop a strategy for definitive revascularization. Intravenous access enables assessment of the hemodynamic significance of the blockage, as well as the presence of any congenital anomalies.

The limitation of invasive venography is the inability, even when combined with intravascular ultrasound, to evaluate the specific cause of extrinsic SVC compression [13].

Treatment approach

The treatment approach in patients with SVC syndrome should be multidisciplinary and may include oncology, pulmonology, radiology,

surgery, and vascular and endovascular specialists. Treatment options can include chemotherapy with or without RT, surgical bypass, or ET such as angioplasty, stenting, and catheter-based thrombus removal. For advantages and disadvantages of different treatment modalities.

Initial management for all patients with SVC syndrome includes elevation of the head of the bed to decrease the hydrostatic pressure in the head and neck. The management of SVC syndrome related to malignancy is centered on immediate relief of symptoms, as well as specific treatment of the underlying cancer. In life-threatening situations, initial stabilization with ABCs (airway, breathing, circulation) is quickly followed by endovascular recanalization with or without stenting to quickly address the obstruction and provide relief of symptoms [14].

Steroids are often used as prophylaxis against radiation-induced edema and are also used in patients with airway compromise [15].

Radiation therapy

Traditionally, SVC syndrome had been viewed as a relative emergency and RT was considered as the first-line treatment. RT reduces tumor burden, but the benefits are often temporary with 5% to 30% of patients experiencing recurrence of SVC syndrome [16, 17].

Surgical intervention

Open surgical intervention, such as bypass grafting and SVC reconstruction, is reserved for cases of extensive venous thrombosis or occlusion that are highly symptomatic and not amenable to endovascular intervention. The surgical bypass is usually performed from the innominate or the jugular vein to the right atrial appendage or the SVC using a spiral saphenous vein graft [18].

Endovascular therapy

Over the last 2 decades, endovascular intervention with stenting has become the standard of care for SVC obstruction, for both benign and malignant etiologies.ET does not affect the subsequent histologic diagnosis and can be combined with other treatment modalities including chemotherapy and radiation, if needed [19].

SVC syndrome with thrombosis

In some cases of SVC obstruction, there is superimposed thrombosis, likely related to stagnation of flow, a hypercoagulable state from underlying malignancy, or the presence of indwelling catheters. Thrombus removal with CDT or aspiration thrombectomy is recommended prior to revascularization, in order to prevent pulmonary embolism and reduce the length of lesion to be treated [20].

Prognosis

The average life expectancy for patients who present with malignancy-related SVC syndrome is 6 months, although the prognosis is quite variable depending on the type of malignancy. SVC obstruction in patients with NSCLC portends a particularly poor prognosis.

Conclusion

A diagnosis of SVC syndrome due to small cell cancer of lung was made. Radiation therapy with or without chemotherapy is the main stay of treatment in most patients. Now a days, endovascular therapy is considered the standard of care approach, providing rapid relief with high efficacy, without adversely affecting subsequent treatment with radiotherapy or chemotherapy when it is needed.

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