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Case Report

Emergency Department Diagnosis and Management of Superior Vena Cava Syndrome Secondary to Hodgkin's Lymphoma: Case Report and Limited Literature Review of Emergency Department Management

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ABSTRACT

Superior vena cava (SVC) syndrome refers to the obstruction of blood through the SVC with limited case reports and literature reviews of its presentation and management. Here we discuss a 26-year-old male who presented with progressive chest pain and shortness of breath for 5-weeks. The exam showed facial and neck plethora and distension of the veins in the neck and chest wall. This case discusses SVC syndrome due to external compression by Hodgkin's Lymphoma. The diagnostic approach and the clinical management of superior vena syndrome are reviewed.

Keywords
Superior vena cava syndrome; Superior vena cava obstruction; Jugular venous distention; Hodgkin's disease.

INTRODUCTION

Superior vena cava (SVC) syndrome is an uncommon condition seen in the emergency department (ED). There are limited case reports and literature reviews. Because of more sophisticated imaging technologies and medical care, emergency physicians now rarely see advanced cases of SVC and may be less familiar with the presentation and initial management.

Classic SVC syndrome presents with chest pain, shortness of breath, facial and neck plethora, prominent neck veins, collateralized chest wall veins and a cyanotic appearance of skin with an area of demarcation at the lower chest. Up to 85% of SVC syndromes are due to a malignant process with the most common being small cell lung cancer (SCLC).

Superior vena cava syndrome requires rapid assessment and management. Untreated SVC syndrome may lead to life threatening cerebral and laryngeal edema.

CASE REPORT

A 26-year-old male construction worker presented to the ED with a complaint of chest pain and shortness of breath that was present and worsening for approximately 5-weeks. The pain localized over the right upper chest region was described as intermittent and pressure-like. Any time the patient walked more than forty yards he reported feeling presyncopal. He also reported that he would become lightheaded and pale when working out in the heat. Additionally, a work colleague noticed that with prolonged exposure to heat spider veins over his chest became prominent with a demarcated rash at the level of the diaphragm. In addition to symptoms of generalized fatigue and malaise, the patient reported a fifteen-pound weight loss occurring over one month. The weight loss was attributed to a reduced appetite.

One-week prior to the ED visit the patient sought care at a community urgent care center where a presumptive diagnosis of asthma was tendered, and an inhaler was prescribed. No diagnostic testing or imaging was performed by those healthcare providers.

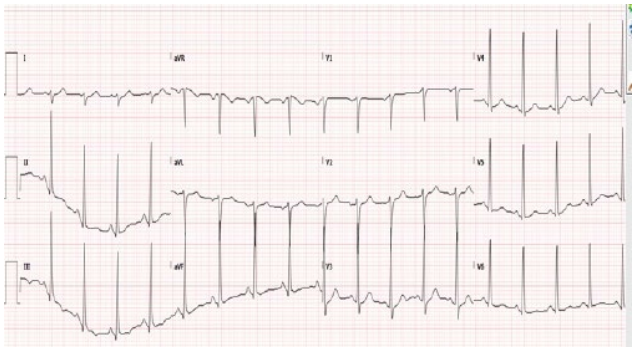
The past medical history was negative for any health problems. He denied using tobacco products or any recreational drugs. Alcohol was consumed socially and in moderation. No allergies were reported and there were no previous hospitalizations. The family history was remarkable for a younger sibling having

recently died from a lung cancer.

The patient's physical exam revealed a well-nourished male in no acute distress. He ambulated to the patient care area without difficulty. Vital signs were as follows: T: 36.7 C (Oral) HR: 106 bpm (monitored) RR: 18 breaths/minute, BP: 127/76, SpO_2: 100%. Weight was 63.5 kg.

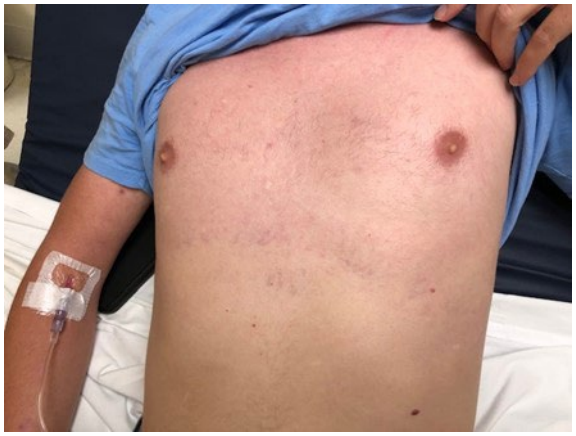
His electrocardiogram demonstrated a sinus tachycardia with right axis deviation (Figure 1).

Figure 1. *Sinus Tachycardia with a Right Axis Deviation*



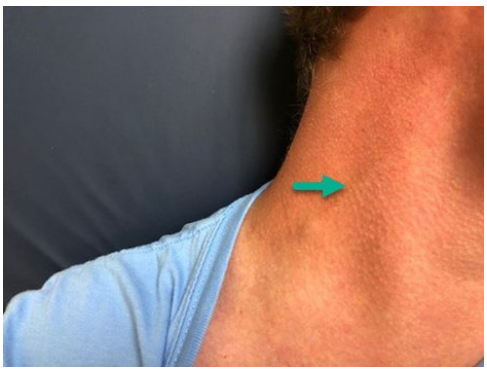
Collateralized chest wall veins with cyanotic appearance of skin (Figure 2), and facial and neck plethora with associated prominent neck veins (Figure 3) were noted. Pemberton's sign manifested by facial congestion and cyanosis when both arms were elevated was positive.

Figure 2. *Collateralized Chest Wall Veins with Cyanotic Appearance of Skin*



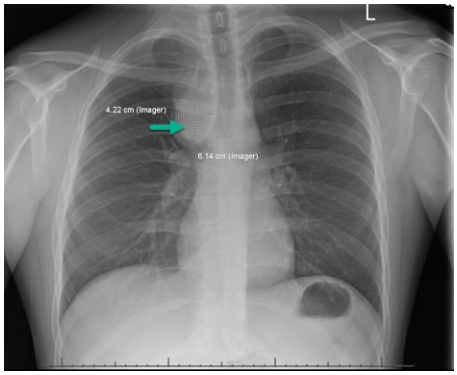
Labs showed: Hemoglobin 10 g/dL, Hematocrit 34 g/dL, white blood cells (WBC's) 5.9×10/9 L, D-Dimer 0.86 mg/L, Total Protein 8.7 g/dL, and Albumin 2.8 g/dL. Cardiac profile including complete blood count (CBC), Prothrombin Time/International Normalized Ratio (PT/INR), Pro-brain natriuretic peptide (BNP) and chemistries were all within reference range.

Figure 3. *Prominent Jugular Venous Sistension, (Green Arrow) and Neck Plethora*



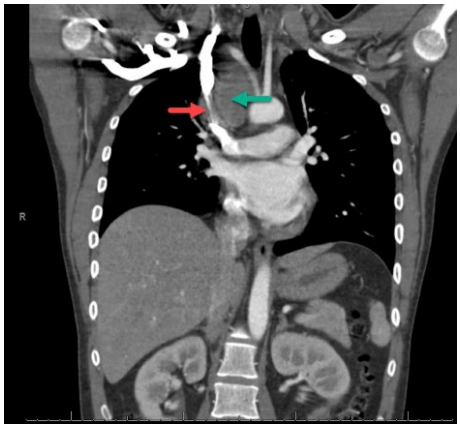
Imaging studies were performed, and his chest radiograph (Figure 4) showed a right paratracheal superior mediastinal mass measuring 4.2×6.1 cm.

Figure 4. *Chest Radiography Showing a Right Paratracheal Superior Mediastinal Mass. (Green Arrow) Measuring 4.2×6.1 cm*



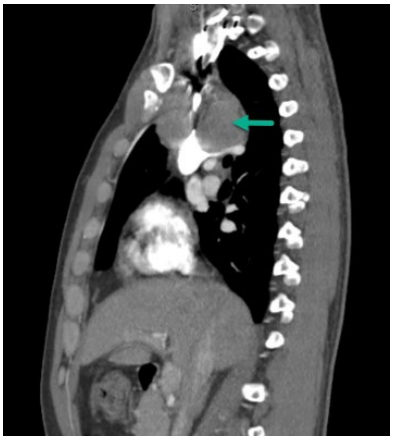
Computed tomography angiography (CTA) of the thorax (Figure 5A) demonstrated a 6.7×4.2×5.9 cm superior mediastinal mass on the right with near complete encasement of the right SVC

Figure 5A. *Chest CT Demonstrating a Superior Mediastinal Mass (Green Arrow) almost Completely Compressing the Superior Vena Cava (Red Arrow) with a Maximum Diameter of 0.7 cm*



with mass effect and luminal narrowing but no evidence of invasion. The maximum diameter of the SVC measured 0.7 cm (Figures 5A and 5B). Narrowing of the distal inferior vena cava (IVC) with mediastinal adenopathy was also present. Findings were consistent with malignancy. Cardiovascular and thoracic surgery was subsequently consulted for these findings.

Figure 5B. *Chest CT Sagittal View Demonstrating Superior Mediastinal Mass (Green Arrow) Compressing the Superior Vena Cava*



Emergency department management consisted of sitting the patient in an upright position. There was no indication for supplemental oxygen as breathing was non-labored and oxygen saturations were optimal. A single dose of dexamethasone, 6 mg, was administered intravenously.

The patient's symptoms were consistent with a SVC syndrome due to external compression by the mediastinal mass. The differential diagnosis of the mass was broad and tissue sampling was required for diagnosis. After admission to the cardiovascular thoracic surgery service, the patient underwent a mediastinoscopy with biopsy and lymph node resection. His post-operative diagnosis was classic Hodgkin's lymphoma favoring nodular sclerosis. After undergoing a staging positron emission tomography-computed tomography (PET-CT) scan, counseling regarding long-term chemotherapy and radiation therapy options was provided to the patient.

DISCUSSION

Superior Vena Cava Obstruction

Superior vena cava syndrome refers to the obstruction of blood through the SVC. SVC syndrome is often regarded as an oncologic emergency that requires rapid assessment and management.

Epidemiology

An estimated 15,000 cases of SVC syndrome occur each year in the United States with studies pointing to increasing frequency due to concomitant rise in the use of semi-permanent intravascular catheters.[1-4]

Pathophysiology

The pathophysiology of SVC syndrome involves thrombosis, invasion of the vessel or extrinsic compression. When the SVC becomes obstructed collateral veins start to form. When these collateral veins can no longer compensate for the degree of SVC obstruction, the clinical features of SVC syndrome begin to manifest. If the obstruction happens quickly and before collateral flow has been established, the clinical features can present sooner. More commonly, it is an insidious process and patients can be asymptomatic for years. Collateral veins may come from the azygous, internal mammary, lateral thoracic, paraspinous, and esophageal venous systems.[5]

The SVC is a thin walled, valveless low-pressure system that drains deoxygenated blood from the upper half of the body including the head, arms, and thoracic wall into the right atrium. It also drains blood from below the diaphragm *via* the azygous vein. There are three main tributaries contributing to the SVC. The first is the right and left brachiocephalic veins which form the SVC which in turn drains into the right atrium. There are no valves in the SVC or brachiocephalic veins. Before the SVC enters the pericardium, it receives the azygous arch, the second main tributary. The third tributary of the SVC is the mediastinal and pericardial veins. The brachiocephalic veins are formed at the confluence of the subclavian and internal jugular veins behind the sternoclavicular joint.[6]

Superior Vena Cava Syndrome Etiologies

Causes of SVC syndrome can be benign or malignant. Up to 85% of SVC syndromes are due to malignant processes with the top 3 causes being SCLC, non-small cell lung cancer (NSCLC) and non-Hodgkin's lymphoma.[5,7] Other causes include germ cell tumors, thyroid malignancies and metastatic disease. Approximately 2-4% of lung cancer patients will develop SVC obstruction. SCLC presents the highest risk for SVC obstruction due to rapid growth of the disease.

Benign causes of SVC syndrome include thrombosis secondary to intravascular devices[2-4] goiter, fibrosing mediastinitis, fungal infection, aortic aneurysm, and retrosternal thyroid.

Signs and Symptoms

Signs and symptoms of SVC syndrome vary depending on the degree of obstruction. Regardless of etiology, dyspnea is the most common symptom. The most common physical findings are facial edema and distension of the veins in the neck and chest wall. Arm edema, cyanosis and facial plethora occur less frequently.

Patients who have an SVC obstruction are unable to drain blood efficiently from the upper limbs, neck, and face into the heart. As a result, collateral flow occurs which results in several specific and common signs and symptoms (Table 1).

Diagnosis

A thorough history and clinical examination and recognizing the

Table 1. *Signs, Symptoms and Percentage of Patients with Superior Vena Cava Obstruction*

Facial Symptoms	Pharyngeal/Laryngeal Symptoms	Chest Wall and Upper Extremities	Neurologic Symptoms	Cardiovascular
Distension: Facial edema and plethora (20%)[1,12]	Dyspnea (54%) and cough (54%)[12]	Distension: Upper extremity edema and plethora (46%)[12,21]	Altered mental status due to cerebral edema (4%)[22]	Chest pain
Distension: Conjunctival and periorbital edema[1]	Facial Angioedema (83%)[12,23]	Dilated chest wall (53%) and neck veins (63%)[12,20]	Headaches (9%) and syncope (10%)[12]	Hemodynamic symptoms due to poor cardiac output
Proptosis	Laryngeal edema stridor (4%) and hoarseness (17%)[12]		Papilledema and visual disturbances (2%)	

Table 2. *Treatment Classification System of SVC Syndrome*

Grade	Symptoms	Incidence	Management	Clinical Features
Grade 0	Asymptomatic	10%	Imaging	Radiographic findings
Grade 1	Mild	25%	Diagnostic and staging procedures if malignancy.	Edema in the head or neck, plethora, cyanosis
Grade 2	Moderate	50%	Diagnostic and staging procedures if malignancy.	Edema in the head or neck with functional impairment. (Visual disturbances caused by ocular edema, cough, mild dysphagia and impairment of head, jaw neck or eyelid movements)
Grade 3	Severe	10%	Diagnostic and staging procedures if malignancy.	Mild or moderate laryngeal edema or diminished cardiac reserve (syncope particularly after bending). mild or moderate cerebral edema such as headache or dizziness
Grade 4	Life-threatening	5%	Sit in upright position, oxygenation for hypoxemia, intravenous fluids if tachycardic or hypotensive, intubation for severe respiratory distress, diuretics, steroids (Decadron 4 mg Q 6-hours)	Laryngeal edema (stridor), cerebral edema (confusion, obtundation), or hemodynamic compromise (syncope without precipitating factors, hypotension, or renal insufficiency).
Grade 5	Fatal	< 1%	Same as Grade 4	Extension of Grade 4 with life-threatening hemodynamics.

**Grade 3, 4 or 5 symptoms are regarded as rare*

classic features are key to diagnosing SVC syndrome (Table 1). Pemberton's Sign, a lesser-known finding, is demonstrated when both arms are elevated resulting in facial congestion and cyanosis.[8]

The electrocardiogram (EKG) may have several characteristic findings. Increased central venous pressures, resulting from SVC obstruction can cause a right axis deviation, a finding noted in our patient (Figure 1). Central venous pressures (CVP) can be increased in pulmonary artery hypertension (PAH).

Useful laboratory values may include a D-Dimer which is elevated in most cases of SVC syndrome[9] although this is not sensitive or specific for SVC thrombus.

Patients with high clinical suspicion for SVC should undergo imaging of the vasculature and upper body. Ultrasound of the subclavian, innominate, and jugular veins can help to identify a thrombus within the vessel lumen. Radiographic imaging to include chest radiography and magnetic resonance imaging (MRI) also play a significant role providing additional information as to the severity, location, and etiology of the SVC obstruction.[10] CT of the chest is associated with a diagnostic specificity of 92% and sensitivity of 96%. Lastly, venography is widely accepted as the gold standard for diagnosing and visualizing a venous obstruction.

Magnetic resonance imaging has potential advantages over CT scanning in the diagnosis of SVC syndrome. MRI does not require iodinated contrast material, allows for direct visualization of blood flow and provides images in several planes of view. Disadvantages include increased cost, and increased scanning time.[11]

Management

The objective of treatment is to relieve the symptoms and treat the cause. Yu et al[12] proposes treatment based on a classification system which defines the role of interventions (Table 2).

This classification system is based on the National Institute of Health's Common Terminology Criteria for Adverse Events (CTCAE) v.3.0. Management of patients with Grades 1, 2, or 3 (Table 1) usually undergo diagnostic and staging procedures as the etiology is usually a malignancy. Grades 3, 4 and 5 are rare.[12]

Immediate intervention such as intubation may be required for life-threatening symptoms (Grade 4). Acute management in patients (Grades 4 or 5) include sitting the patient in an upright position, oxygenation and intravenous fluids if the patient is tachycardic or hypotensivenive. Use of diuretics may be indicated in patients with severe respiratory distress[11] as well as a course of parenteral steroids (Dexamethasone, 4 mg every 6-hours).[13] Review of the literature indicates steroids are routinely used in managing SVC obstruction where malignancy is suspected. However, there is no evidence to support or negate their use. Steroids have been used in conjunction with radiotherapy to reduce radiation edema.[14,15] Advanced airway management may be reserved for progressive glossal or laryngeal edema.

The literature is unclear regarding the benefits of anticoagulation in the acute setting. More than 85% of cases are due to malignant diseases that arise because of metastasis or primary tumors. Tumor masses can be necrotic and friable and may be sub-

ject to increased venous pressures. Anticoagulant therapy would therefore pose a risk of bleeding. Consequently, fibrinolysis should be avoided.[11] On the other hand, when patients do not improve after the first week of radiotherapy a thrombus may be responsible, and anticoagulants should be considered.[11] Thrombolytics are useful in caval thrombosis.[16]

Many patients require additional urgent intervention such as endovenous stent placement[17] to open up the SVC vessel, or radiotherapy. The therapy of choice is radiation therapy if the cause is malignancy.[18] In this case, high-dose short interval radiation therapy is the primary treatment.

Definitive Management

Once stabilized, more specific management usually requires a histologic diagnosis to determine the best treatment options. Chemosensitive malignancies can be treated with chemotherapy, radiation, or surgery. In general chemotherapy is best for small cell lung cancer and lymphomas. Chemo-insensitive malignancies can be treated with a stent or radiotherapy. Radiotherapy is best for NSCLCs. In rare circumstances surgical intervention may be considered.[19]

Prognosis and Outcomes

Prognosis is highly variable, depending on the cause and stage at diagnosis. For benign causes of SVC syndrome, the life expectancy is not changed. However, when malignancy is diagnosed, there is a significant decrease in survival. Individuals who present with life-threatening symptoms such as cerebral[7,20] or laryngeal edema due to malignancy usually have shortened life-expectancies of less than 24-months.

CONCLUSION

Superior vena cava syndrome, obstruction of blood through the SVC, is an uncommon life-threatening emergency condition. We present a case report and a limited review of the literature concerning the diagnostic approach and the clinical management of superior vena syndrome.

CONSENT

The authors have received written informed consent from the patient.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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