

SUPERIOR VENA CAVA SYNDROME: STILL A MEDICAL DILEMMA

- A Case Report -

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Implication Statement

Many options are available for treating superior vena cava syndrome. However, the decision of the appropriate plan to be taken can be challenging in critically sick patients with multiple coexisting illnesses, particularly, if it is associated with coagulopathy.

Abstract

Purpose

The purpose of this report is to highlight the dilemma and the associated clinical implications in treating a patient with superior vena cava syndrome (SVCS) with a coexisting coagulopathy.

Clinical Features

This case report describes a post-bone marrow transplant patient who was admitted to our ICU because of bronchiectasis complicated with nosocomial pneumonia. Following the recovery from pneumonia and long ventilatory support, he developed superior vena cava syndrome (SVCS) due to mediastinal lymphadenopathy. The diagnosis was delayed due to associated confounding clinical factors. Due to the rapid deterioration in patient's condition, the immediate tissue diagnosis of mediastinal lymph nodes and re-canalization of superior vena cava by stenting was not done though it was, our priority. He had many other medical problems as well such as thrombocytopenia, deranged coagulation profile, old cerebral infarction with hemiplegia, seizure disorder and cardiac arrhythmias that complicated the treatment plan.

Ultrasonography (USG) guided biopsy followed by stenting of the SVC was done after discussing the risks and benefits with patient's relatives. But, he had bleeding from biopsy site due to deranged coagulation profile. He was not given any anticoagulants.

Within 24 hours, the stent was blocked by clot that was diagnosed by the deteriorating clinical features and a repeat CT scan. Then he was given Enoxaparin in therapeutic dose and the clot cleared within a day, possibly partly due to Enoxaparin and partly coagulopathy.

Conclusion

Meticulous care should be practiced in deciding the appropriate treatment of SVCS especially when it is associated with other complicating medical problems particularly coagulopathy.

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Introduction

Superior vena cava syndrome (SVCS) in an uncommon complication of many disease conditions, which include malignancies¹, hematological disorders and patients with transvenous pacemakers² and central venous catheters³.

Most cases are caused by malignancies, and SVC obstruction due to malignancies usually progress rapidly leading to complete obstruction which can cause a patient's condition to deteriorate rapidly⁴. Also benign conditions are increasing due in large part to iatrogenic injuries from central venous catheters (CVC) and transvenous pacemakers.

Obstruction due to benign disease may be indolent. SVC obstruction usually presents with swelling of face and upper extremities, conjunctival suffusion, periorbital swelling, proptosis, dyspnea, respiratory distress and pleural effusion and rarely life-threatening complications, one of which is pulmonary embolism (PE)^{1,5,6}.

As symptoms, such as respiratory distress, systemic hypotension, and intracranial hypertension are life threatening, this condition has to be treated on an emergency basis with definitive strategies, ranging from anticoagulation^{7,8} medical management⁹ to radiological stenting^{10,11}, chemotherapy¹², radiotherapy¹³ or surgical bypass¹⁴. We present an interesting case of SVCS in a patient admitted to our ICU.

Case Report

A 20 yr old patient, who had undergone allogenic bone-marrow transplant for acute myeloid leukemia one year earlier, presented to our Emergency Department with breathing difficulty. He was a known case of bronchiectasis with history of exacerbations for which he was admitted many times. He had multiple other complications of bone-marrow transplant such as, liver graft versus host disease (GVHD), hemosiderosis on monthly venesections and Desferol, 6 month old watershed infarct involving right middle cerebral artery and anterior cerebral artery with hemiparesis, 4 month old chronic subdural hematoma with regular neurosurgical follow ups and past history of seizures on Valproic acid.

Patient was tachypneic, tachycardic and restless.

Chest had rhonchi and crepitations, the oxygen saturation (pulse oximetry) was around 75% on 10 L/min O₂ flow by facemask with reservoir bag. Arterial blood gas showed respiratory acidosis due to CO₂ retention (pCO₂ = 16 kPa).

He was admitted to the ICU and was given a trial of noninvasive ventilation without improvement. His trachea was intubated and started on pressure controlled mechanical ventilation. Patient had a turbulent course in ICU; he developed multiple complications such as nosocomial infection with septic shock, right-sided heart failure, uncontrolled seizures and arrhythmias such as intermittent paroxysmal supraventricular tachycardia (PSVT).

A CVC was inserted in his right internal jugular vein (IJV) to monitor central venous pressure and for giving various intra venous medications. A CT scan of chest done during ICU stay showed significant bronchiectatic changes with bilateral upper lobe fibrosis and his arterial CO₂ was always between 10 to 12 kPa. Patient was tracheostomised anticipating prolonged ventilatory support.

Once his respiratory and hemodynamic parameters stabilized, weaning was gradually attempted. His ventilatory requirements came down from a pressure controlled mode to a pressure support mode (PSV), but he could not be weaned further. He intermittently complained of headache and pain in the upper chest, which we attributed to hypercarbia and tracheotomy wound. The tracheostomy tube tie was loosened and some analgesics prescribed.

On 26th day of ICU stay, patient developed significant puffiness of face, neck and both upper extremities, along with tachypnea, irritability, drowsiness and oliguria. Initially, the renal impairment was attributed to recurrent sepsis and drug toxicity, as he was on (Amikacin, Amphotericin B, and Cyclosporine). Over a period of two days the swelling became worse in the same region without involving the lower extremities. From the day of admission his coagulation profile was deranged (INR varying from 1.5 to 1.7) and platelet count was low (40 × 10⁹/L to 80 × 10⁹/L) due to GVHD. He received fresh frozen plasma and platelet concentrates before invasive procedures such as CVC cannulation, tracheostomy, rectal biopsy etc. were done.

Although patient had coagulopathy, SVCS was suspected due to external compression of superior vena cava by mediastinal lymphadenopathy from a relapsed primary malignancy (AML). Initial Doppler ultrasonography (USG) was inconclusive. But, subsequent CT scan confirmed the diagnosis of a superior venacaval obstruction by enlarged mediastinal nodes. A femoral venous catheter was immediately inserted for administration of intravenous medication, but the CVC in the right IJV was retained as advised by the radiologist for potential radiological interventions. A decision was made to perform an USG guided mediastinal lymph node biopsy for tissue diagnosis and SVC stenting in the same sitting. The radiologist performed the mediastinal biopsy and stenting of SVC. There was local bleeding from the biopsy site due to a deranged coagulogram (INR = 1.73, APTT = 73 seconds and platelet count = $79 \times 10^9/L$) and high venous pressure in the collateral circulation. Bleeding stopped with local pressure and wound sutured. It was decided not to transfuse any blood products because of fear of thrombosis of the stent. CVC in IJV was withdrawn and fixed just above the stent by the radiologist for possible future use. He was not given any post-stenting anticoagulant due to the deranged coagulogram and possibility of bleeding from biopsy site.

On the next day (day 27 of ICU stay) there was a gross increase in facial, upper limb swelling and diminished air entry at both lung bases. There was no decrease in CVP measured through the CVC in right IJV. Urgent CT scan showed thrombosis of SVC and right subclavian vein along with bilateral pleural effusion. He was started on Enoxaparin in therapeutic dose (40 mg twice daily). Thrombolytic therapy or intravenous Heparin was withheld for fear of possible rebleed from mediastinal biopsy site and also from chronic subdural hematoma and an old cerebral infarct site. Vascular surgeon ruled out the possibility of surgical intervention due to poor general condition of the patient.

He was now complaining of more chest pain. There was a gradual decrease in urine output and increase in blood urea and creatinine, possibly due to contrast induced nephropathy. His lung compliance was deteriorating steadily with a parallel increase in

pCO₂ in spite of full ventilatory support. He developed frequent episodes of PSVT and atrial fibrillation. Cardiologist started him on Amiodarone in spite of fibrotic lungs as he was already on Sotalol for PSVT with no response.

There was no improvement in his symptoms after 24 hours of treatment with Enoxaparin. The biopsy of the mediastinal lymph node was reported as crushed tissue. Hence no definitive treatment for the enlarged nodes was possible. Instead it was decided to repeat the CT scan and give local thrombolytic therapy (with Alteplase) through the CVC in the right IJV if found to have persistent thrombosis. Fortunately, the radiologist found the SVC to be patent. So he was continued on Enoxaparin without any further intervention.

Discussion

Superior vena cava syndrome generally occurs due to impairment of normal venous return through superior vena cava by either compression by adjacent tumor mass or lymph nodes, or thrombosis of SVC due to long term indwelling extraneous devices and hypercoagulable states^{1,2,3,4,5}. Even though the diagnosis depends on a high degree of alertness on the part of the treating physician, it can be quite challenging to diagnose SVC syndrome in a critically ill patient like ours.

Our patient had undergone allogenic bone marrow transplant one year earlier for acute myeloid leukemia (AML). There was neither any history of relapse of AML before this admission to ICU nor any signs, symptoms or laboratory reports, which would suggest the same during the current episode of acute illness. The main problem which led to ICU admission and respiratory failure was acute exacerbation of bronchiectasis which was later complicated by severe ventilator associated pneumonia and ARDS. He successfully recovered from all these problems.

On retrospective analysis, though our patient had many signs and symptoms of SVCS, the diagnosis was difficult at an earlier stage due to his various confounding medical factors. He had respiratory distress in the form of inability to wean from ventilatory support. This could have been due to his pathological conditions such as bronchiectasis, lung

fibrosis and poor muscle power. He complained of moderate chest pain in his upper chest, which was attributed to tracheostomy wound and was relieved by analgesics. His coagulation profile was deranged from first day of admission to ICU with low platelet count (40 to $70 \times 10^9/L$), high INR (1.5 to 2.0) and APTT (45 to 60 seconds) which made thrombosis related to a CVC a remote possibility. Patient was on multiple nephrotoxic drugs; hence the initial low-grade facial swelling was attributed to progressing renal failure. When the swelling progressed in upper limbs without involving lower limbs, patient was investigated as a SVC syndrome.

The treatment options for SVCS depend on the primary pathology and progression on the disease. Any malignant condition has to be treated by radiotherapy or chemotherapy after confirming the tissue diagnosis^{9,12,13}. If the symptoms progress rapidly leading to respiratory distress, neurological deterioration or hemodynamic instability, immediate intervention may be required in the form of balloon angioplasty, stenting or surgery^{10,11,14}.

Benign conditions leading to SVCS usually respond to medical treatment in the form of head elevation, anticoagulants, thrombolysis, diuretics and steroids^{1,7,8}. Nowadays it is increasingly being treated by angioplasty, local thrombolysis and stenting^{15,16}.

USG or CT guided biopsy are time tested methods of tissue diagnosis of mediastinal mass with few contraindications. The absolute contraindications include uncontrollable cough and suspicion of hydatid cyst, whereas relative contraindications include bleeding diatheses, vascular lesions, pulmonary

hypertension, uncooperative patient, and advanced emphysema¹⁷.

In this patient, CT scan revealed mediastinal lymph nodes compressing the SVC and patient's condition deteriorated rapidly. He developed frequent PSVT and paroxysmal atrial fibrillation (PAF) with hemodynamic instability and desaturation in spite of being treated with sotalol. SVC irritation by compressing lymph node could have been a focus of this atrial arrhythmias¹⁸. So, mediastinal biopsy was planned for tissue diagnosis and possible chemotherapy to treat the relapsed AML or secondary malignancies.

On retrospective analysis, the decision to do USG guided biopsy along with angioplasty and stenting of SVC was a therapeutic misadventure as it prevented us from giving anticoagulation for maintaining the patency of the stent. But, it was expected that the stent would not clot with a deranged coagulation profile. But next day when the symptoms increased, a repeat CT scan showed thrombosed stent. As this patient had a 6-month-old cerebral infarction and chronic subdural hematoma and was at high risk for intracranial bleed from thrombolysis and intravenous Heparin therapy, only Enoxaparin was started in therapeutic dose. Fortunately, the stent became patent radiologically within 24 hours.

To conclude, patients with rapidly progressing SVCS which requires urgent stenting, diagnostic mediastinal biopsy should not be attempted in the same sitting. In patients with deranged coagulation profile, some anticoagulants should be given after SVC stenting in order, to prevent stent thrombosis.

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