UNUSUAL PRESENTATION OF SUPERIOR VENA CAVA SYNDROME

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· 26 year old African American female with PMH significant for SLE and ESRD who was transferred from outside facility for management of acute hypoxic respiratory failure associated with subepiglottic narrowing.

· On arrival, she was emergently nasally intubated by ENT due to impending airway compromise from what appeared to be acute bacterial epiglottitis.
▪ She was also noted to have diffuse edema of the face, lips, gingiva, and neck which was then presumed due to angioedema. The patient was treated with solumedrol, plasmapheresis x 7 days, 2 doses of a C1 inhibitor, and Ecallantide® (Kalbitor), a Kallikrein inhibitor. The patient failed to respond to antibiotic and steroid therapy.

▪ The patient also experienced severe epistaxis which required transfusion of 7 units of pRBCs. An epistaxis embolization procedure also failed to achieve hemostasis.

▪ When symptoms of angioedema failed to respond adequately to these measures, rheumatology and hematology/oncology were consulted for further investigation.

▪ It had been previously noted during an attempted tunneled right IJ PICC placement, that the patient’s SVC and other named veins within the thorax were occluded with collateral formation.
RELEVENT HISTORY

▪ Past Medical History
  ▪ Systemic Lupus Erythematosus and End Stage Renal Disease secondary to lupus nephritis

▪ Past Surgical History
  ▪ Left upper extremity dialysis fistula-chronically thrombosed and not in use
  ▪ Right subclavian dialysis catheter

▪ Family & Social History
  ▪ Recent sick contact (young child with presumed viral URI)

▪ Review of Systems
  ▪ Shortness of breath, cough, subjective fevers

▪ Medications
  ▪ Plaquenil and prednisone

▪ Allergies
  ▪ Iodinated Contrast
DIAGNOSTIC WORKUP

▪ Physical Exam on admission
  ▪ Temp: 98.1, BMI 21.7
  ▪ Inspiratory and expiratory stridor with bilateral wheezing
  ▪ Extensive soft tissue swelling of the head, face, neck, and upper chest

▪ Laboratory Data
  ▪ WBC 4.7, Hemoglobin 8.6 g/dL, Hematocrit 27.6%, C3 level of 39 (low), and C4 level of 8 (low), double stranded DNA antibodies were positive
DIAGNOSTIC WORKUP

- Non-Invasive Imaging
- Hospital Day 1: Soft Tissue Neck Radiograph performed at outside facility shows marked narrowing of the trachea at the glottis (arrow). Lateral image hints at the diffuse soft tissue swelling.
Hospital Day 9: CT Soft Tissue Neck for neck swelling shows diffuse, extensive edema and swelling of all imaged subcutaneous soft tissues of the head, face, neck, and upper chest. Edema extends into the deep soft tissues of the neck.
During attempted placement of a tunneled right IJ PICC, it was noted that the SVC and all named veins in the chest were occluded. Opacification of collateral veins was demonstrated.

During epistaxis embolization, it was noted that the right IJ was patent to the level of brachiocephalic with minimal residual lumen (arrow).
DIAGNOSIS

- Superior Vena Cava Syndrome

- As mentioned in the HPI, this was initially diagnosed as bacterial epiglottitis and then diagnosed as angioedema. All treatments failed to achieve significant response.
Venogram and Treatment of SVC Syndrome

- The patient was referred for recannalization treatment. Catheter venogram of the right IJ demonstrated chronic occlusion extending from the lower neck to the clavicle with opacification of numerous venous collaterals in the lower neck and chest wall.

- A fibrin sheath surrounding the right subclavian hemodialysis catheter left a small residual patent lumen in the SVC. From this approach, multiple attempts were made to reconnect the occluded portion of the right IJ to the residual patent lumen of the right brachiocephalic via a sheath and snare placed in the right subclavian.

- Ultimately, using sharp recannalization of a 22 gauge 20cm needle, the chronically occluded portions of the right IJ and brachiocephalic veins were snared and bridged to the right subclavian.

- Balloon angioplasty of the right IJ, brachiocephalic, and SVC was performed.
• Following balloon angioplasty, a 20mm Gianturco tracheobronchial Z stent was placed in the SVC.
• Simultaneous placement of 12 mm Wallstents in the right IJ and brachiocephalic/subclavian veins was performed. Stents were brought to profile by balloon angioplasty. The central aspect of both stents were within the Gianturco SVC stent.
• The right IJ stent successfully extended across the previously occluded segment.
• Subsequent venography demonstrated wide patency of all 3 stents with no further opacification of the previously noted collaterals. A right subclavian hemodialysis catheter was replaced.
1) Which of the following is NOT a reason to perform therapeutic intervention on someone with SVC syndrome?

A: Acute respiratory failure

B: Mental status changes

C: Symptomatic improvement after antineoplastic therapy

D: Persistent head, neck, and upper extremity soft tissue swelling
1) Which of the following is NOT a reason to perform therapeutic intervention on someone with SVC syndrome?

A: Acute respiratory failure. This is a reason for urgent intervention. Venous hypertension is causing soft tissue edema which may be life-threatening.

B: Mental status changes. This is another reason for urgent intervention. Venous hypertension is causing cerebral edema which may be life-threatening.

C: Symptomatic improvement after antineoplastic therapy. Current antineoplastic therapy can be continued if the patient’s symptoms have significantly improved or resolved.

D: Persistent head, neck, and upper extremity soft tissue swelling. Persistent symptoms despite maximal medical therapy is an indication for intervention and improves the patient’s quality of life.

CONTINUE WITH CASE
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C: Symptomatic improvement after antineoplastic therapy. Current antineoplastic therapy can be continued if the patient’s symptoms have significantly improved or resolved.

D: Persistent head, neck, and upper extremity soft tissue swelling. Persistent symptoms despite maximal medical therapy is an indication for intervention and improves the patient’s quality of life.
Because of the patient’s contrast allergy and resolution of symptomatology, no direct imaging follow up has been performed. The patient remains anticoagulated with Coumadin.

Pre- and post-procedural chest radiographs demonstrate much improved soft tissue swelling.
SUMMARY & TEACHING POINTS

▪ SVC syndrome can have a confusing presentation with many overlapping symptoms with other diseases\(^1\)\(^4\).

▪ The patient’s clinical presentation was probably caused by acute on chronic venous obstruction secondary to hypercoagulability from active lupus (positive ds-DNA antibodies) and the long standing presence of a dialysis catheter\(^5\), \(^6\).

▪ Diagnosis was confounded by the recent sick contact in the setting of acute hypoxic respiratory failure. Diagnosis was further delayed because her clinical presentation was then presumed to represent complement mediated angioedema secondary to active lupus\(^7\).

▪ The unsuccessful treatment of her epistaxis by embolization also likely reflected the ongoing venous hypertension\(^6\), \(^8\).

▪ Today, SVC syndrome is usually (~70%) seen in the setting of an obstructing neoplasm, though benign causes (~30%) are occasionally noted\(^8\)\(^-\)\(^10\).

▪ Before antibiotics were widely available, this ratio was essentially reversed. Many cases were caused by syphilitic aortic aneurysms or an infectious mediastinitis.

▪ SVC syndrome was initially described by William Hunter in 1757. Hunter was an Scottish physician and anatomist who described a case of a 39 yo man with SVC syndrome secondary to a syphilitic aortic aneurysm\(^11\).

▪ More recent cases seem to suggest that an increasing incidence of SVC syndrome is caused by indwelling catheters\(^1\), \(^5\), \(^6\), \(^8\).
REFERENCES


