Superior Vena Cava Syndrome

Kristen Bridges M.D.
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Case

• 64F
  – ESRD on HD since 2006 s/p R DDRT 2009
    • RUE AVF 1/2006
    • Also with LUE AV access
    • Stents x 2 of R subclavian vein
  – DM
  – HTN
  – GERD
  – HLD
More PMH

- Januvia
- Calcitriol
- Lasix
- Famotidine
- Prednisone 5
- Donepezil
- Tacrolimus
- ASA 81
- Nateglinide
- Sitagliptin
- Metoprolol
- Mycophenolate

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Presentation

• Presented to ED on 6/8/15 with 1 day facial swelling & dyspnea
HD #1: CT neck with contrast
CT Neck Interpretation

• Multiple vascular collaterals throughout the anterior neck and chest wall
  – raising suspicion of **SVC occlusion/in-stent subclavian vein thrombosis**
• Questionable filling defect within the **L IJ**
  – Known hx of thrombus
• Bovine aortic arch
Venogram Interpretation

- Tapering ➔ **abrupt occlusion of R cephalic vein** at proximal humerus
  - reflux into multiple collaterals
- **No flow through R subclavian/brachiocephalic stents**
- Nonvisualization of R basilic, axillary, or subclavian veins
Hospital Course

- **HD#8**: Underwent L innonimate vein → R atrial bypass with bovine pericardium
- **POD #1-2**: kept intubated for difficult airway requiring fiberoptic intubation
Hospital Course

• POD #1-2 continued:
  – Air leak test negative x 2 days, swollen, congested airway

• POD #3:
  – Extubated
  – A fib- chemical cardioversion with amiodarone
  – 1U PRBC transfused
Hospital Course

• POD #4
  – Weaned off amiodarone gtt successfully

• POD #5-14
  – Resolving AKI with preserved urine output
  – PO amiodarone

• POD #15
  – Discharged to rehab
SVC Syndrome
History

• **1757**: William Hunter described 1\textsuperscript{st} case of SVCS
  – Cause: saccular syphilitic aneurysm of ascending aorta

• **1837**: William Stokes described first case secondary to malignancy

• **1949**: McIntire/Sykes reported first series 502 cases
  – Primary benign, syphilitic aortic aneurysm/chronic fibrous mediastinitis from TB
  – Only 1/3 were due to primary thoracic tumors
History

• **Klassen 1951, Bricker & McAfee 1952:**
  – First successful bypasses in humans using autologous femoral vein grafts

• **1954:** Schechter reported study of 274 cases
  – 40% due to syphilitic aneurysms or tuberculous mediastinitis
Anatomy

- SVC surrounded by:
  - Trachea
  - Right bronchus
  - Sternum
  - Aorta
  - Pulmonary artery
  - Paratracheal/perihilar LN

- Easily compressed because of low pressure/thin wall
Introduction

• Compression of **SVC** by various causes leading to:
  – facial/upper body edema
  – formation of collaterals
  – cyanosis/dyspnea

• **15,000** new cases each year
  – **60%** associated with malignancy
  – 25-40% from benign sources
    • Syphilis, tuberculosis, central venous catheters
Etiology

• Prior to 1949, SVCS largely secondary to infectious etiology (tuberculous/syphilitic mediastinitis) now largely secondary to thoracic malignancies
  – Until mid-1900s, aortic aneurysms were 2\textsuperscript{nd} most common cause

• <40% today caused by benign sources
  – Mostly central venous devices (pacemakers, AICDs, central venous infusion ports, long term HD catheters)
Etiology

• **Malignant : 60%**
  – NSCLC 50%
  – SCLC 22%
  – Lymphoma 12%
  – Metastatic cancer 9%
  – Germ cell cancer 3%
  – Thymoma 2%

• **Benign : 40%**
  – Mediastinal fibrosis, granulomatous disease (ie histoplasmosis)
  – Indwelling CVC/ cardiac pacemakers
  – Radiotherapy to mediastinum
  – retrosternal goiter
  – aortic dissection
CVC/Pacemakers

- 170,000 pacemakers implanted annually,
  >5 million CVC
  - 7-33% experience upper extremity/central venous DVT
  - See SVCS in 1→3% with CVC
  - SVCS in 0.2→3.3% with implanted pacemakers
Diagnosis

• Usually suggested from H&P
  – Physical findings
    – Feeling of fullness in head/neck
      – Bending forward/lying down worsens
    – Dyspnea on exertion/orthopnea
      – Grade severity by # pillows
    – Headache/mental confusion
    – Dizziness/Syncope
    – Visual problems
    – Cough
    – Head and neck swelling/dilated neck veins
    – Prominent chest wall collaterals
    – upper limb edema
Diagnosis

Pemberton’s sign:

– Have patient elevate both arms until they touch sides of face
– Wait one minute
– Positive with facial congestion, cyanosis, respiratory distress
Diagnosis

• Diagnostic confirmation:
  – Xray
  – Sonography
  – CT
  – Venography
  – MRI
• Mediastinal widening
• Right hilar/mediastinal mass
• Pleural effusion
• Bilateral infiltrates
• Upper lobe collapse
• Dilated collaterals
• >90% of patients: Dx can be made on basis of clinical findings and plain CXR!
• Aortic nipple- enlarged superior intercostal vein
  • (draining hemiazygous system)
Ultrasonography

- Good screening technique
- Can’t directly visualize SVC transthoracically
- Can see **subclavian/IJ veins**
  - In SVCS, normal respiratory variance lost
- Collaterals in chest wall/mediastinum visible
CTA

- Distinguishes benign from malignant sources
- Also shows:
  - Mediastinal mass
  - Pulmonary lesions
  - SVC obstruction
  - Hilar LAD
  - Pleural effusion
  - Collateral pathways
Magnetic Resonance Venography

- Anatomic excellence
- Good if contraindication to IV contrast
Venogram

• Most conclusive Dx tool
• Direction of collateral flow
• Identifies thrombus
• “Gold standard” for central venous obstruction
Collateral Venous Pathways

1. Azygos system
   - Includes azygos vein, hemiazygos vein, connecting intercostal veins

2. Internal mammary system, tributaries, and secondary communications to superior/inferior epigastric veins

3. Long thoracic venous system
   - Connected to femoral and vertebral veins
Manifestations of supra-azygos SVC obstruction
- Distended arm and neck veins
- Oedema of neck, face and arms
- Congested mucous membranes (mouth)
- Dilated, tortuous vessels on upper chest and back

Manifestations of infra-azygos SVC obstruction
- More severe symptoms but all of the features for obstruction distal to entrance of SVC
- Dilation of collateral vessels on anterior and posterior abdominal wall with downward blood flow into IVC, then back to heart
Stanford and Doty Class I

- Up to 90% SVC stenosis with **patency** and **antegrade** flow of azygos - R atrial pathway
- increased collateral circulation through the **hemiazygos** and **accessory hemiazygos veins**
Stanford and Doty Class II

- >90% SVC stenosis or occlusion with patency and antegrade flow in the azygos – R atrial pathway
• >90% stenosis or occlusion of the SVC with **retrograde** flow in both the **azygos** and **hemiazygos** veins
Extensive occlusion of the **SVC** and **innominate** and **azygos** veins with chest wall and epigastric venous collaterals
Treatment: Conservative

- Extra pillows
- Avoidance of bending over
- No constricting garments/tight collars
- Diuretics
- Malignant disease → anticoagulation
- Mediastinal malignancies → chemoradiation
Endovascular treatment of both acute and chronic SVC occlusion is first line of treatment
Treatment: Endovascular Stents

- Fast functional relief
- Femoral + upper extremity access
- Balloon angioplasty, stenting, thrombolysis, thrombectomy
- Patency rates good
  - 57-79% primary
  - 85-100% primary-assist or secondary
Endovascular Stents

• Downside:
  – Need for secondary interventions continues over mid (possibly long) term

• Upside:
  – Decreased procedure-related morbidity
  – Shorter recovery period
Treatment: Surgical Bypass

- **Candidates**: Stanford and Doty class III/IV and type I/II that failed to respond to endovascular treatment
- Rarely performed with malignant disease because of success rate of stenting
  - Malignant causes of SVCS should only undergo OSR if life expectancy >1 year
- More commonly used with benign causes of disease
Results of Open Bypass

• 87.5% graft patency at 11 years
• 86% secondary patency at 5 years
Graft Materials

• **Greater Saphenous Vein:**
  - Poor size match
  - Extra-anatomic bypass from jugular vein to femoral vein

• **Femoral Vein:**
  - Appropriate size match
  - **Risks:**
    - leg edema/pain,
    - chronic venous insufficiency
Graft Materials

• Spiral Saphenous Vein Graft
  – Increases diameter of saphenous vein
Graft Materials

- **ePTFE**
  - Used almost exclusively for large vein reconstruction

- **Iliocaval allograft**
  - Use when immunosuppressives already in use

- **Homografts, cryopreserved femoral vein, aortic arch grafts, autogenous or bovine pericardium**
Operation

• Median sternotomy
• Open pericardium to expose R atrial appendage
• Side-biting Satinsky clamp on R atrial appendage
• Open longitudinally
• End-to-side anastomosis with vein graft performed
• Peripheral anastomosis performed with internal jugular or innominate in end-to-end fashion
• Post op anticoagulation started 24hrs later with heparin, discharged on oral AC regimen
  – SSVG/femoral vein grafts: 3 months coumadin
  – ePTFE: lifelong AC
Follow up

• Surgical bypass:
  – CTA or MRV at 3-6 months and anytime afterward when symptoms reoccur
  – No need for continued reinterventions over time

• Endovascular repair:
  – Number of repeated interventions MUCH higher
Summary

• Four types of SVC syndrome Stanford and Doty classification
• Endovascular treatment first line
• Majority of SVCS caused by malignancy
Questions?
A 70yo female with stage IIIA lung cancer has an implanted venous access device placed via the right subclavian vein. She received 2 cycles of chemo and 70Gy radiotherapy. One year later, after the catheter was removed, she presents with SOB, headaches, edema of the head and neck. On physical exam, she has prominent distended veins in upper chest/arms. A venogram of the subclavian/innominate veins is performed.

Next step in management?

a) High dose steroids
b) Chemoradiotherapy
c) Fibrinolytic therapy
d) Stent placement to the innominate vein and SVC
e) Subclavian vein to right atrial bypass with PTFE graft
Question #2

A 38yo male has facial and upper extremity edema, venous distension in the neck and arms, and a cyanotic appearance. Which of the following is true?:

a) If a malignant lesion is identified, resection is indicated for palliation
b) Mediastinoscopy for diagnosis is contraindicated
c) If the cause is benign disease, gradual improvement without operation is expected
d) The most likely cause of the problem is mediastinal granulomatous disease
e) A venogram should be obtained to confirm the diagnosis
Resources

• **pre azygos**: in this conditions mainly the right superior intercostal veins serves as the collateral pathway to drain into the azygos vein.
  – Direct SVC bypass via superficial venous systems resulting in no clinical evidence of SVC syndrome

• **azygos**: when the azygos vein is also obstructed the collateral circulation establishes between SVC and IVC via minor communicating channels i.e. internal mammary veins, superior and inferior epigastric veins to iliac veins and finally into the IVC.

• **post azygos**: in this case the blood from the SVC is distributed into the azygos and hemiazygos and then into the IVC tubutaries i.e. ascending lumbar and lumbar veins.
  – Below/at azygos: SVC bypass via superficial venous systems: clinical evidence of SVC syndrome