A Mis-LEADing Presentation of Angioedema

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Our Case

80 year-old male veteran presenting to the ED with complaints of facial swelling and dyspnea after completion of his scheduled hemodialysis session

- Increasing facial and tongue swelling after each HD
- First time he’s had dyspnea
What further questions do you have?

A) New medications?
B) Any changes in dialysis?
C) ACE/ARB use
D) Rash or itching?
E) All of the above
Medical History

Past Medical History
- ESRD due to diabetic nephropathy on HD M/W/F
- Type 2 Diabetes Mellitus
- Complete Heart block s/p pacemaker placement in 2010
- Bilateral BKA
- Coronary Artery Disease with MI in 2010
- GERD

Social History
- Non-smoker, denied EtOH or illicit substances
- Lives in his home with his son and daughter-in-law, limited mobility due to BKA with multiple falls

Medications
- Glargine 10 units qPM
- Plavix 75mg daily
- Metoprolol 12.5mg BID
- Simvastatin 40mg QHS
- Sevelamer 800mg TID
- Midodrine 12.5mg TID with HD
Physical Exam

- General appearance: elderly male, dishelved appearing, NAD, speaking in full sentences with garbled speech
- Vital Signs unremarkable
- Eyes: PERRLA, EOMI, no scleral icterus and normal conjunctiva with periorbital edema
- HEENT: right greater than left neck and face swelling without stridor, enlarged tongue
- CV: RRR, no m/r/g, device palpable in left upper chest
- Resp: Lungs clear to auscultation without respiratory distress
- Ext: RUE A-V fistula in place with palpable thrill, bilateral BKA
- Skin: without rash or hives or other lesions
- Neuro: AAOx3, garbled speech due to enlarged tongue without focal deficit
Initial clinical course

- Admitted to the MICU for airway monitoring
- Epinephrine, solumedrol, famotidine, and diphenhydramine administered
- Bedside fiber-optic exam
  - Mild epiglottic and supraglottic edema
  - No vocal cord abnormalities
Hospital Course

- Transitioned to oral corticosteroids
- Intermittent episodes of mild to moderate facial swelling without dyspnea
- C4, C1 esterase inhibitor and C1q level all within normal limits
- Discharged to subacute rehab
Ongoing clinical course

- Completes oral corticosteroids in SAR
- Discharge day 2, re-presents with same symptoms
- Readmitted and restarted on high dose IV corticosteroids, famotidine, and fexofenadine
- Symptoms again improved quickly on steroids
- Continue to have intermittent facial swelling
- Observation reveals symptoms beginning after being placed in recumbent position
Case CT chest
- chronic superior vena cava narrowing surrounding the pacemaker leads with extensive collaterization
- (put what is in report)
Hospital course

- Vascular surgery was consulted
  - stenting recommended

- Balloon angioplasty with placement of a 14mm x 60mm stent from the right subclavian vein to the superior vena cava

- Complete resolution of his symptoms
Differential diagnosis

- Angioedema
  - Hereditary
  - Acquired
  - Medication Induced
- Allergic reaction/anaphylaxis
- Mass effect
- Trauma
- Superior vena cava syndrome
- Malignancy
- Late manifestation of pacemaker placement
Superior Vena Cava Syndrome

- Most commonly associated with malignancy (60-85% cases)¹
  - Lung cancer (both NSCLC and SCLC)
  - Lymphoma
  - Germ Cell Tumors
- Benign causes
  - Fibrosing mediastinitis
  - Infections
  - First described with syphilitic aortic aneurysm
  - Complication following aortic aneurysm repair or other surgeries
  - Related to intravascular device
  - Port-a-cath
  - Dialysis catheter
  - Pacemaker wire

¹ Rice et al, Medicine. 2006;85(1):37
Pacemaker induced SVC Syndrome

- First described patient with a permanent cardiac pacemaker in JAMA in 1973\(^2\)

- Incidence of symptomatic SVC obstruction from pacemaker/ICD insertion in the literature ranges from 1 in 650 to 1 in 3100\(^3\)

- Onset after pacemaker insertion ranges days to years
  - Average of 48 months

- Can involve SVC obstruction via thrombosis, stenosis, or a combination of both

2. Wertheimer et al. JAMA. 1973
Diagnosis of pacemaker induced SVC Syndrome

- Gold standard: Contrast venography
- MRA use is limited due presence of pacemakers/ICDs
Treatment of pacemaker induced SVC Syndrome

- No clear guidelines at this time
- Surgical management
- Transvenous stenting with angioplasty

4. Riley et al. Pacing Clin Electrophysiol. 2010
Fast forward to follow up

- No recurrence of symptom at 8 months status post procedure
- Plan with IR/Vascular surgery for stent exchange in future
- No anticoagulation
  - Bilateral BKA
  - Frequent falls at home
References


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