A Mis-LEADing Presentation of Angioedema

KATIE BATLEY, M.D. AND ALLISON NITSCH, M.D.

COLORADO STATE ACP CONFERENCE

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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

Audience response

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

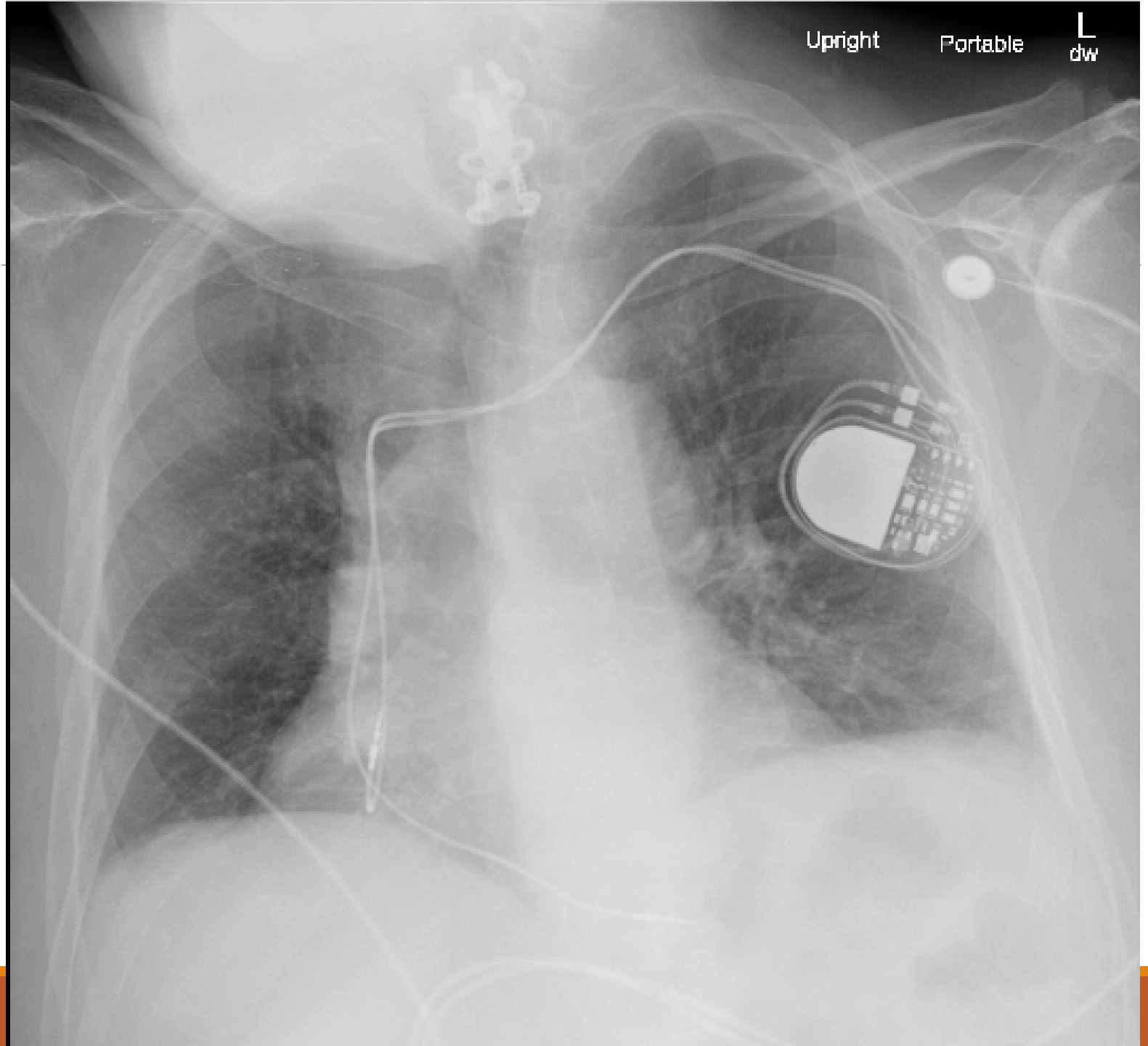
- Non-smoker, denied EtOH or illicit substances
- Lives in his home with his son and daughter-in-law, limited mobility due to BKA with multiple falls

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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Initial clinical course

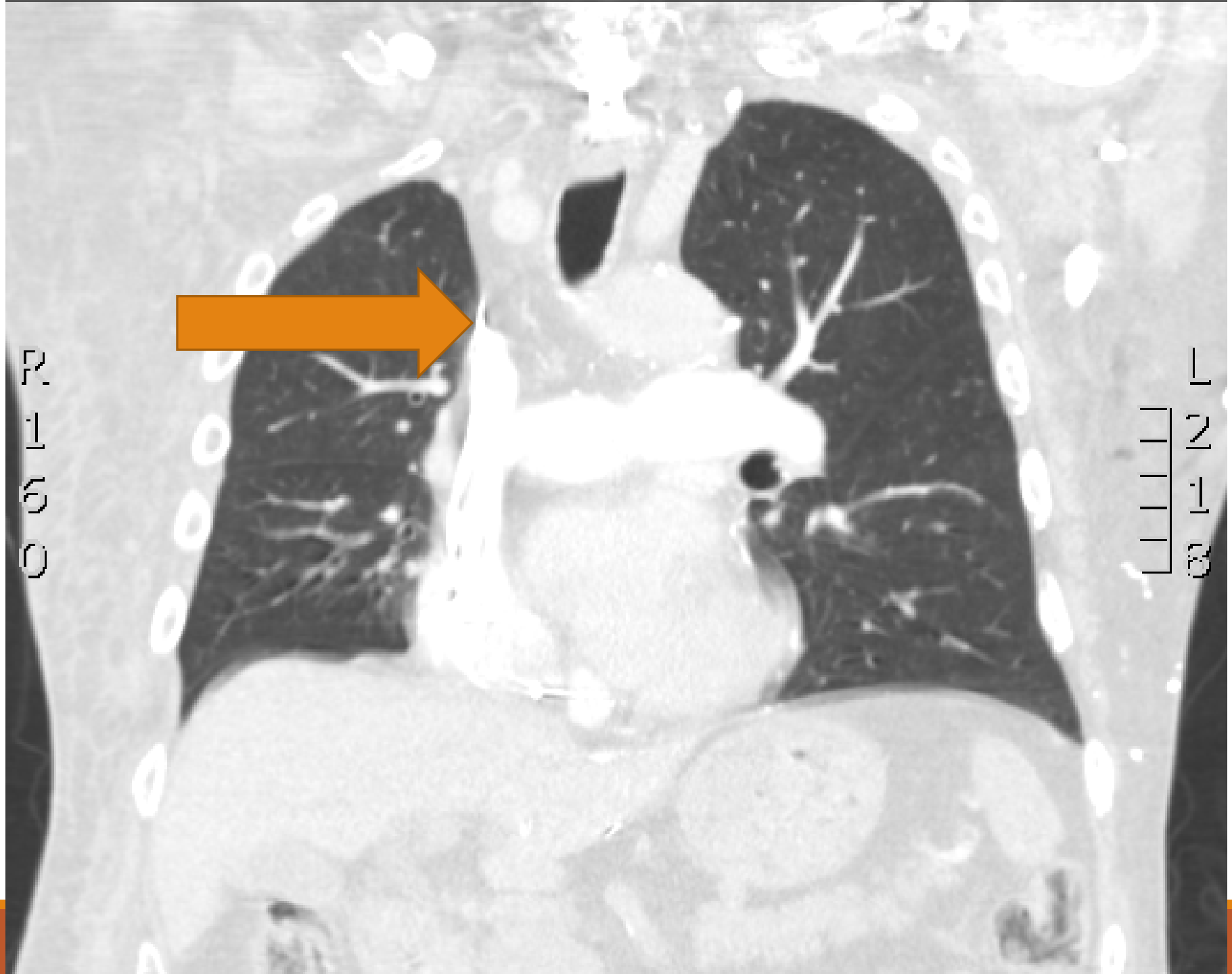
- Admitted to the MICU for airway monitoring
- Epinephrine, solumedrol, famotidine, and diphenhydramine administered
- Bedside fiber-optic exam
  - Mild epiglottic and supraglottic edema
  - No vocal cord abnormalities

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**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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**

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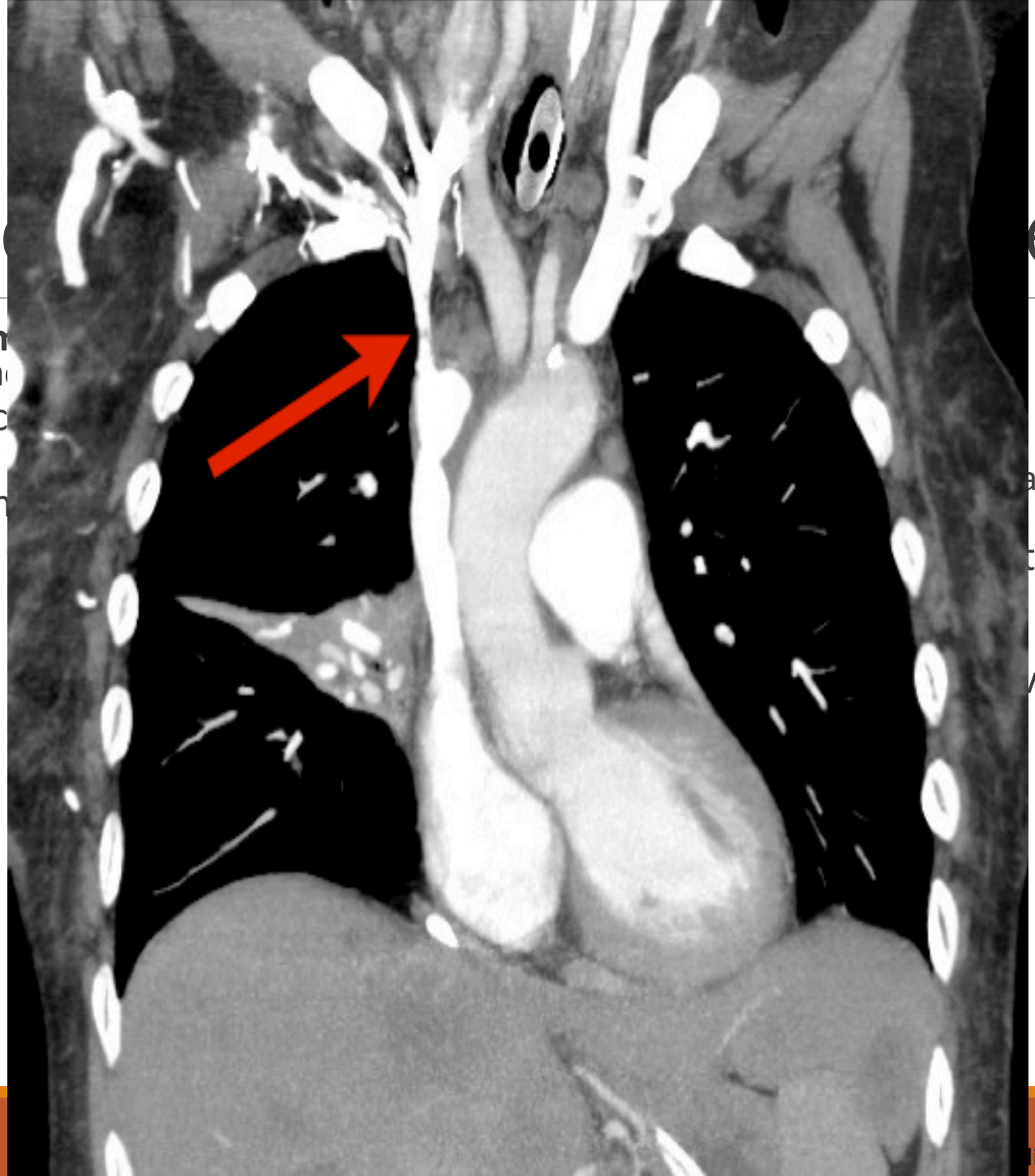


licine. 2006;85(1):37

Pacemaker induced SVC Syndrome

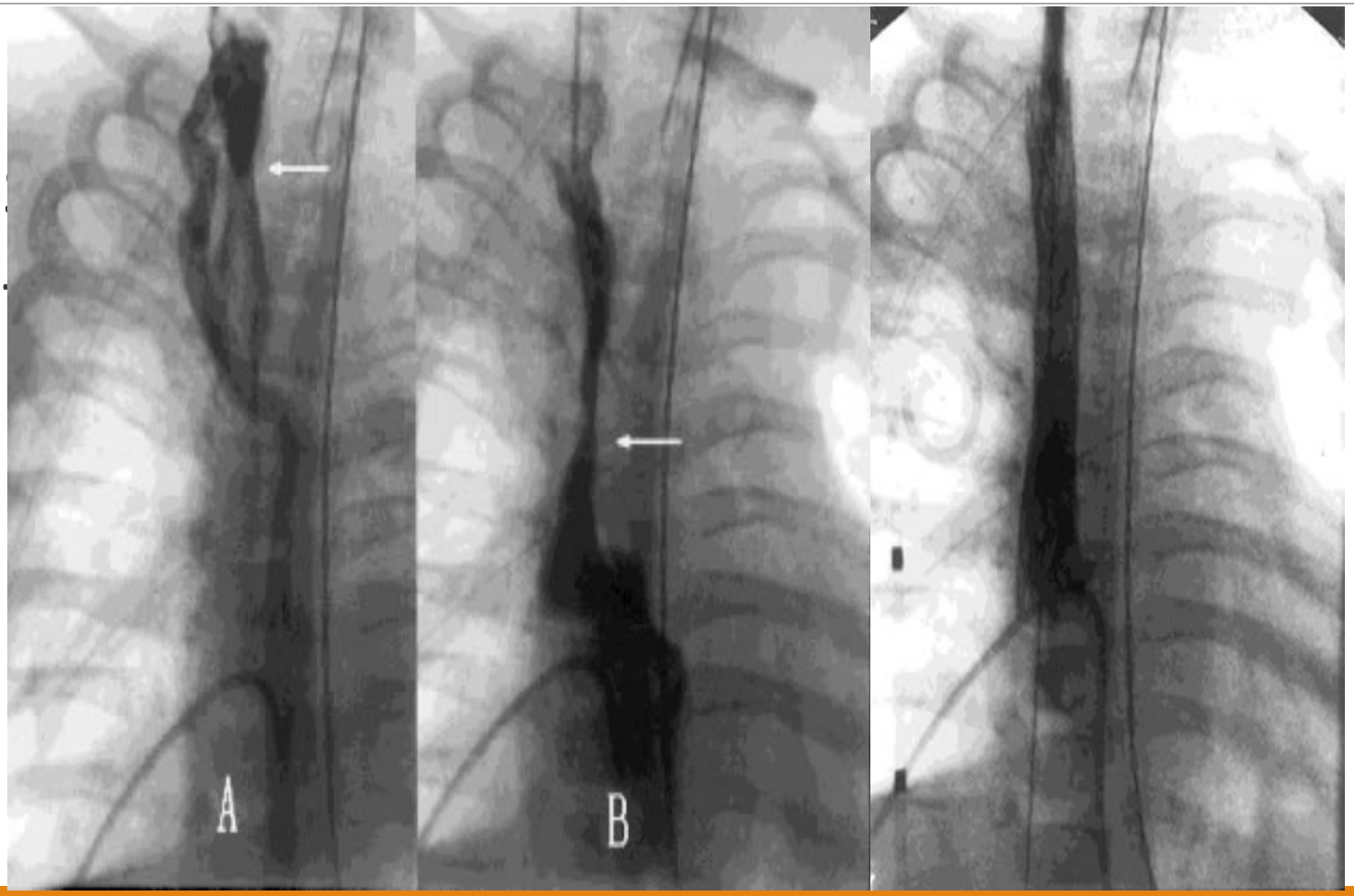
- First described patient with a permanent cardiac pacemaker in JAMA in 1973[2]
- Incidence of symptomatic SVC obstruction from pacemaker/ICD insertion in the literature ranges from 1 in 650 to 1 in 3100[3]
- Onset after pacemaker insertion ranges days to years
  - Average of 48 months
- Can involve SVC obstruction via thrombosis, stenosis, or a combination of both

2. Wertheimer et al. JAMA. 1973
3. Gebreyes et al. J Community Hosp Intern Med Perspect. 2012

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



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

1. **The superior vena cava syndrome: clinical characteristics and evolving etiology.** Rice TW et al. Medicine (Baltimore). 2006
2. **Superior vena cava syndrome. Complication of permanent transvenous endocardial cardiac pacing.** Wertheimer et al. JAMA. 1973 May 21;224(8):1172-3.
3. **Be aware of wires in the veins: a case of superior vena cava syndrome in a patient with permanent pacemaker.** Gebreyes et al. J Community Hosp Intern Med Perspect. 2012 Oct 15;2(3).
4. **Managing superior vena cava syndrome as a complication of pacemaker implantation: a pooled analysis of clinical practice.** Riley et al. Pacing Clin Electrophysiol. 2010 Apr;33(4):420-5.

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