An unusual case of rapidly progressive multiorgan system failure

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Disclosures

None
Your pager goes off at 2AM on ICU night float
Mr. Smith is decompensating

Increasing pressor requirements
Increasing oxygen requirements
Worsening altered mental status
Previously healthy 76-year-old man
PMH: hypertension, Legionnaire’s disease in 2019
  • No outside records, from FL
No home meds
Presenting with **progressive dyspnea, fevers, fatigue x 5 days**
Initial Data

• VS: BP 110/73, HR 101, RR 24, T 36.8C, SpO2 95% on 2L NC

• Exam:
  • General: robust elderly man
  • Pulm: tachypnea, **mild respiratory distress**, clear lungs
  • CV: **tachycardic, irregularly irregular** rhythm, no murmurs
  • Extremities: **cool**, no edema
Labs

BNP 54,447
Troponin T 0.07
Lactate 5.6
CRP 282
Sed rate 7

Pending:
- COVID
- Flu
- Blood cultures
Initial Imaging

CT Abdomen + Pelvis without contrast: bilateral perinephric fat stranding

Chest CT: ground glass opacities in left lower lobe
Initial Cardiac Studies

EKG: atrial fibrillation, right bundle branch block
Transthoracic echocardiogram

1. Dilated left ventricle with segmental wall motion abnormalities as described. Overall LVEF is moderately reduced
2. Dilated hypokinetic right ventricle
3. Severely dilated ascending aorta (5.2 cm)
4. Moderate tricuspid regurgitation
5. Mild pulmonary hypertension
6. No transthoracic echocardiographic evidence of endocarditis
Initial differential diagnosis?

- COVID
- Acute coronary syndrome
- Myocarditis
- Severe sepsis in the setting of pneumonia, bilateral pyelonephritis
- Pulmonary embolism
Initial Management

- Empiric broad spectrum antibiotics and fluids for pneumonia
- Cardiology consult with plan for diagnostic catheterization given concern for ACS
Over the next several hours...

- Hypotension, rising lactate
- Chest pain, new lateral ST depressions, rising troponin
- Medical management of NSTEMI
- ICU admission for empiric dobutamine due to concern for cardiogenic shock
You arrive at bedside...

- **General:** ill-appearing, in distress, writhing in bed with severe neck and back pain
- **HEENT:** necrosis of tongue tip
- **Cardiac:** tachycardia, asymmetric blood pressures in left and right arms
- **Respiratory:** tachypnea, respiratory distress, now on max high flow settings
• Abdomen: diffuse tenderness
• Extremities: cool, no edema, pulses not palpable
• Skin: mottling of skin overlying abdomen, legs. Duskeness of bilateral pinnae, glans of penis
• Neuro: no longer oriented, more somnolent, new left facial droop, left-sided weakness
What are you worried about?
What would you do next?
“Extensive large vessel vasculitis with multiple sites of ischemia”
Ascending aortic aneurysm

Aortic arch aneurysm with intimal thickening
Concentric wall thickening around celiac axis
Rheumatology consultation

• Likely rapidly progressive large vessel Giant Cell Arteritis
• High dose methylprednisolone initiated
• Possible risk factor = flu vaccine 1 week prior
Emergent CRRT for acute renal failure, severe lactic acidosis & hyperkalemia

Escalating requirements on 3 pressors

Intubation for progressive respiratory failure
Development of shock liver
Family withdrew care
Patient died less than 72 hours after admission
Giant Cell Arteritis

- Inflammation of medium and large arteries
- Subacute tempo, associated with PMR
- Usually involving branch(es) of carotid (esp. temporal artery)
- Can involve arteries in multiple locations, esp. aorta and its branches
- **Up to 30% present with extracranial large vessel disease**
Large Vessel GCA

- Involves aorta and proximal branches
  - Aneurysms, dissections, stenoses, occlusions
  - Thoracic aorta, brachiocephalic trunk, subclavian, femoral arteries
  - Uncommonly involves CNS and mesentery
- Temporal artery biopsy positive in ~50%
  - Imaging required for dx
- Constitutional sx present, cranial sx absent
- Up to 2/3 of patients with cranial GCA have subclinical LV arteritis
Giant cell arteritis and polymyalgia rheumatica after influenza vaccination: report of 10 cases and review of the literature

Giant cell arteritis or polymyalgia rheumatica after influenza vaccination: A study of 12 patients and a literature review
Teaching Points

- GCA can present without temporal artery involvement and involve multiple large vessels especially aorta
- Suspect it in elderly patients with fever, elevated ESR/CRP, constitutional symptoms even in absence of cranial symptoms
- Usually subacute but can be hyperacute, progressive, and fatal
- Flu vaccine may be a risk factor due to immune activation
References


