Elusive Diagnosis: A Rare Case of Polyarteritis Nodosa

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Disclosures

• I have no disclosures
Introduction

- Polyarteritis nodosa (PAN) has become very rare since the widespread distribution of hepatitis B vaccine[^1]
- The recognition and diagnosis of PAN unlike ANCA associated vasculitides is difficult, requires a high level of clinical suspicion, and either angiographic or histologic confirmation.

Case Presentation

- A 78-year-old male with past medical history significant for HTN, HLD, CAD has a 6-7 month history of postprandial abdominal pain associated with subjective fevers, night sweats and 16lb weight loss
- Patient had been admitted multiple times in the past for similar complaints.
- He had been worked up for FUO in those admissions with no cause identified and had eventually been discharged on hospice care
Physical Examination

- **Vitals:** afebrile, PR- 70 bpm, BP- 114/74 mm of Hg, RR- 14 bpm
- **GEN:** frail looking Caucasian male, no apparent distress
- **HEENT:** dry mucous membranes.
- **HEART:** regular rate and rhythm, 2/6 systolic murmur appreciated at RUSB
- **LUNGS:** clear to auscultation bilaterally. no wheezes, rales, or rhonchi
- **ABD:** soft, non-tender, hypoactive bowel sounds.
- **LEGS:** 3+ pitting edema from knees to toes. dorsalis pedis pulses appreciated, warm extremities.
- **NEURO:**
  - Right ankle unable to plantar flex or dorsiflex. Unable to feel light touch on dorsum of right foot, DTR 1+ in right ankle
  - Otherwise unremarkable neurological exam
Course

- Initial workup showed an Acute Kidney Injury with a Creatinine of 1.5 and the cause was thought to be prerenal.
- Patient was given IV fluid resuscitation
- After further questioning, he reported right foot weakness for the past 2 months
Post Prandial Abdominal Pain

- **Laboratory Findings**
  - WBC - 18,000,
  - Platelet count - 660,000
  - ESR - 62
  - ANA, cryoglobulins, ANCA screen - negative
  - Urinalysis - 16 WBCs/HPF and 5 RBCs/HPF with no casts
  - Hepatitis panel was negative.

- **Radiology - Previous imaging**
  - CTA of the abdomen - mesenteric artery stenosis with vascular calcifications consistent with diffuse atherosclerosis without any aneurysms and normal renal arteries.

Constitutional Symptoms

Foot Drop
Further Course

• **UGI Endoscopy** was performed and showed a gastric ulcer
• Due to suspicion for vasculitis, **Rheumatology** was consulted but no further workup was recommended
• The patient’s **Serum Creatinine continued to rise to 2.6mg/dl** despite IV fluid resuscitation and repeat workup showed an intra-renal picture.
• **Nephrology** was consulted for a possible renal biopsy
A kidney biopsy was done and showed

**Figure 1.** Vasculitis of four different medium-sized arteries. A, Transmural inflammation with necrotizing intimal arteritis (arrows). B, Florid intimal arteritis. C, Transmural arteritis with luminal near-occlusion. D, Recanalization with several small lumina (arrowheads).
Hospital Course Contd.

- He was initiated on IV pulse steroids and oral cyclophosphamide.
- However, the patient developed acute hypoxic respiratory failure overnight due to pulmonary edema, declined intubation and further resuscitation and passed away.
Discussion

- PAN is a rare systemic vasculitis involving the medium sized vessels which is not ANCA driven and the pathophysiology is still not very clear.

- Acute kidney injury due to ischemia, renal infarction or intrarenal hemorrhage can occur.

- Renovascular hypertension may be present and arteriography may show vascular microaneurysms missed by CT angiography\[3\]

Figure 2: Mesenteric angiogram showing involvement of arteries supplying small and large bowel [3]

Proposed Phenotypes

- Classic PAN
- Viral associated PAN
- Monogenic disease associated PAN
- Cutaneous PAN
- Drug Induced PAN
- Pan associated with systemic diseases
- Single Organ PAN
- CNS/PNS PAN
- Primary Angitis limited to CNS\(^4\)
- New genetic associations- DADA2, ADA-2\(^5\)

5. Ozen et al. Nature Reviews Rheumatology 2017
Prognosis

The French Vasculitis Study Group (FVSG) proposed the Five Factor Score (FFS) which includes:

<table>
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<tr>
<th>FEATURES</th>
<th>5 YEAR MORTALITY</th>
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<td>Severe Gastrointestinal Tract disease (defined as bleeding, perforation,</td>
<td>• FFS 0 =12%, 1=26%</td>
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<td>infarction, or pancreatitis)</td>
<td>• When FFS was &gt;=2 mortality was 46%.</td>
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<td>Renal involvement consisting of Serum Creatinine &gt;1.58 mg/dL or proteinuria (&gt;1 g/day)</td>
<td>• The overall 7-year survival for PAN is 79%</td>
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<td>Cardiac disease (infarction or heart failure)</td>
<td>• FFS has been recently re-visited and age &gt;65 years has been also considered as a poor prognosis indicator [6]</td>
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<td>Central nervous system involvement</td>
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Conclusion

- Historically, PAN was often associated with hepatitis B. Most cases now are idiopathic.
- Due to delays in diagnosis similar to our case, there is worsening in the rates of mortality.
- PAN should be considered in patients with multisystem disease associated with organ ischemia.