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.

1. José Hernández-Rodríguez, et al; Diagnosis and Classification of Polyarteritis Nodosa, J Autoimmun. 2014 Feb-Mar;48-49:84-9

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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

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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

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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

Constitutional Symptoms

Foot Drop

- 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.

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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

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A **kidney biopsy** was done and showed

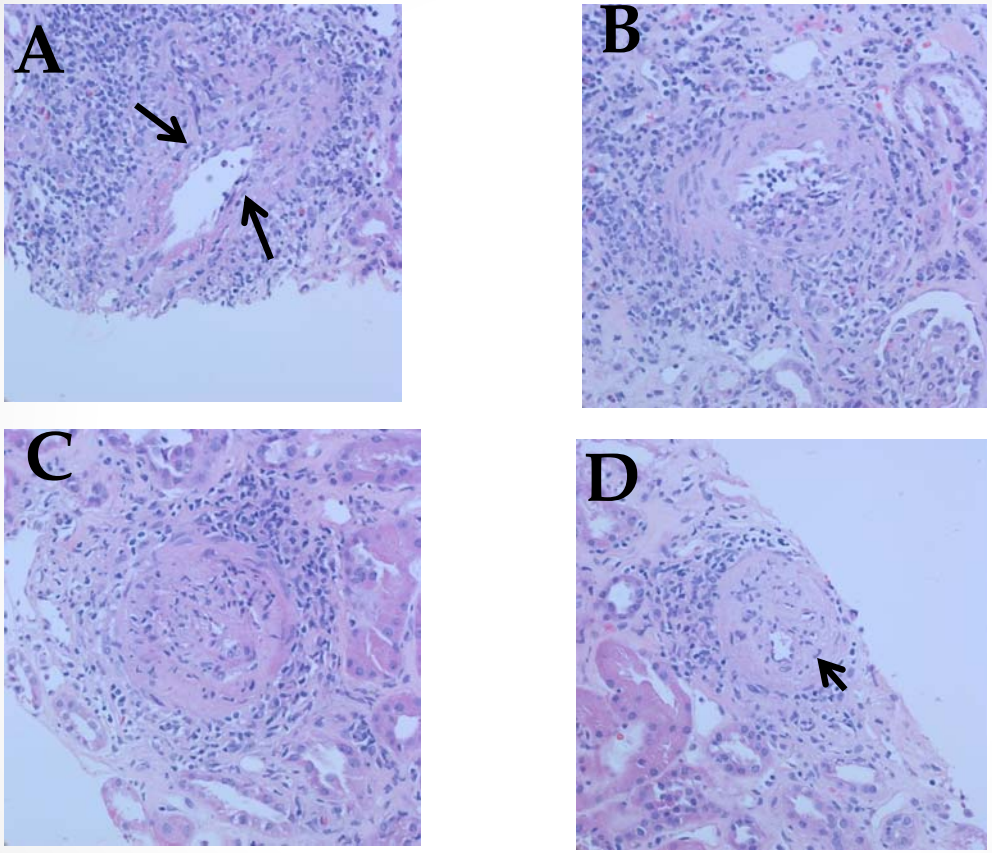

Figure 1. Vasculitis of four different medium-sized arteries. A, Transmural inflammation with necrotizing intimal arteritis (arrows). B, Florid intimal arteritis. C, Transmural arteritis with luminal near-occlusion. D, Recanalization with several small lumina (arrowheads).

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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.

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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]

3. Pagnoux C, et al; French Vasculitis Study Group. Clinical features and outcomes in 348 patients with polyarteritis nodosa: a systematic retrospective study of patients diagnosed between 1963 and 2005, Arthritis Rheum 2010;62:616e26.

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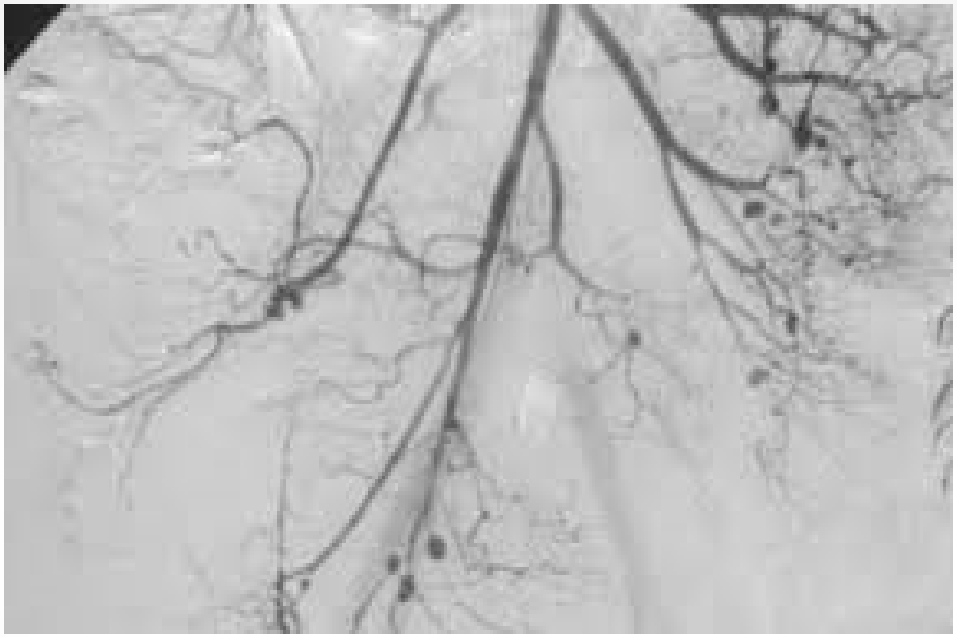

Figure 2: Mesenteric angiogram showing involvement of arteries supplying small and large bowel [3]

3. Stone et al Polyarteritis Nodosa,JAMA, October 2002, Vol 288

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]

4. Karadag and Jayne. Clin Exp Rheum.2018;36(suppl 111): s135-142
5. Ozen et al. Nature Reviews Rheumatology 2017

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Prognosis

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

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

6. Hernandez-Rodriguez J, Alba MA, Prieto-Gonzalez S, Cid MC. Diagnosis and classification of polyarteritis nodosa. J Autoimmun. 2014;48-49:84-9.

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.