



A Veritable Vascular Quandary

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Chief Complaint

Left-sided Headache, Bilateral Leg Pain, and Unintentional Weight Loss

History of Present Illness

An 82 year-old man with a history of hypertension, type 2 diabetes, and stage 3 chronic kidney disease, presented with a subacute left-sided headache, unintentional weight loss, bilateral calf pain, and shoulder tenderness. Symptoms began with tender lower legs two months prior to presentation after he was outside on a cold night. The pain improved with both rest and ambulation, but over the next month, he began to notice bilateral shoulder pain with movement. Just prior to presentation, he developed left-sided headaches without visual changes, aura, nausea, or vomiting that improved with NSAIDs.



Figure 1: MRI Brain with and without contrast showing extensive pachymeningeal contrast-enhancement involving the left greater than right supra and infra temporal fossa.

Physical Exam

Vitals: BP 160/74, HR 58, RR 20, O2 97% on Room-Air

HEENT: no tenderness or cords along bilateral temples or scalp, vision intact

Cardiac: regular rate and rhythm, no murmurs

Pulmonary: clear breath sounds throughout

Abdomen: benign, soft, non-tender, bowel sounds present

Extremities: pain with active overhead shoulder movements, tenderness along both calves without palpable cords or edema

Neurologic: within normal limits

Initial Labs

CBC	WBC: 14,000/uL
Chemistry Panel	Creatinine: 2.0 mg/dL (1.5)
Urinalysis	Moderate Blood 11 RBC/HPF
ESR	95 mm/hr
CRP	155 mg/L

Differential Diagnosis for Pachymeningitis

Infection

- Lyme Disease
- Syphilis
- *Mycobacterium tuberculosis*
- Fungal Infection

Inflammatory Disorders

- Granulomatosis with Polyangiitis
- Rheumatoid Arthritis
- Sarcoidosis
- Sjogren syndrome
- Temporal arteritis
- IgG4-related Disease

Malignancy

- Dural carcinomatosis
- Metastatic disease in adjacent skull

Meningioma

Idiopathic

Workup

I. Temporal artery biopsy

- Small vessel vasculitis but no temporal artery involvement.

II. MRI Brain

- Extensive pachymeningeal enhancement.

III. Lumbar Puncture

- Elevated protein, negative infectious studies.

IV. Sarcoid workup

- Negative after conjunctival biopsy, normal CSF ACE, lack of hilar adenopathy.

V. Immune-complex vasculitis

- Negative rheumatoid factor, and normal C3/C4. IgG4 was normal.
- **pANCA resulted at 1:40 (reference <1:20) with MPO Abs 26 (ref <9).**

VI. Brain biopsy

- Chronic inflammation and small vessel vasculitis.

VII. Treatment

- Azathioprine and prednisone.

VIII. Monitoring

- Regularly attends rheumatology clinic.

Learning Points

- 1) **Hypertrophic Pachymeningitis** is a rare disorder that refers to inflammatory thickening of the dura.
- 2) There is a broad differential for this disease process that includes malignancy, infection, and inflammatory disorders.
- 3) If temporal arteritis is a serious consideration, empiric steroids are unlikely to affect biopsy results.
- 4) Treatment for pachymeningitis generally involves steroids tailored to the patient's response. Other agents such as azathioprine may limit need for chronic high-dose steroids.

Citations

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4. Li S, Tang H, Rong X, Huang X, Li Q. Pachymeningitis as a manifestation of ANCA-associated vasculitis: A care report and literature review. *Int J Clin Exp Med.* 2015;8(4):6352-6359.