

## A Veritable Vascular Quandary

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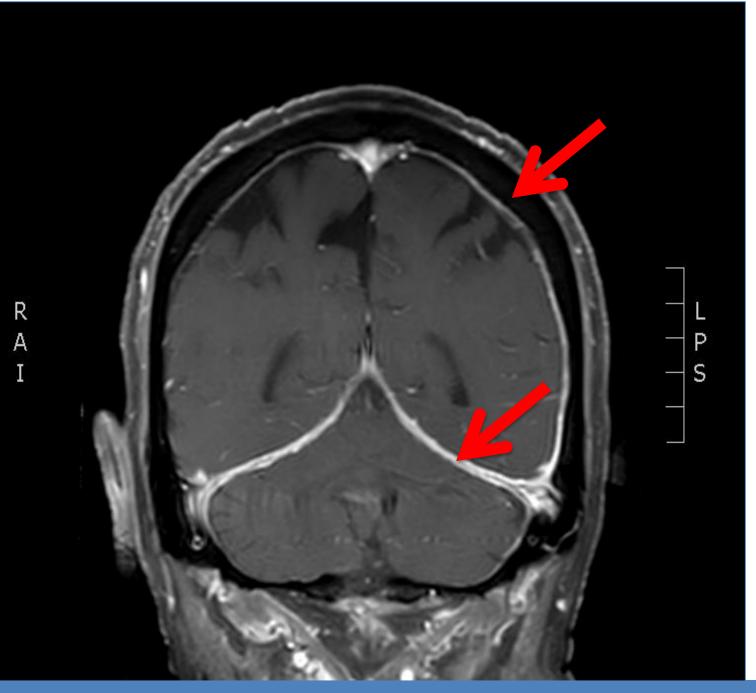


## **Chief Complaint**

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.





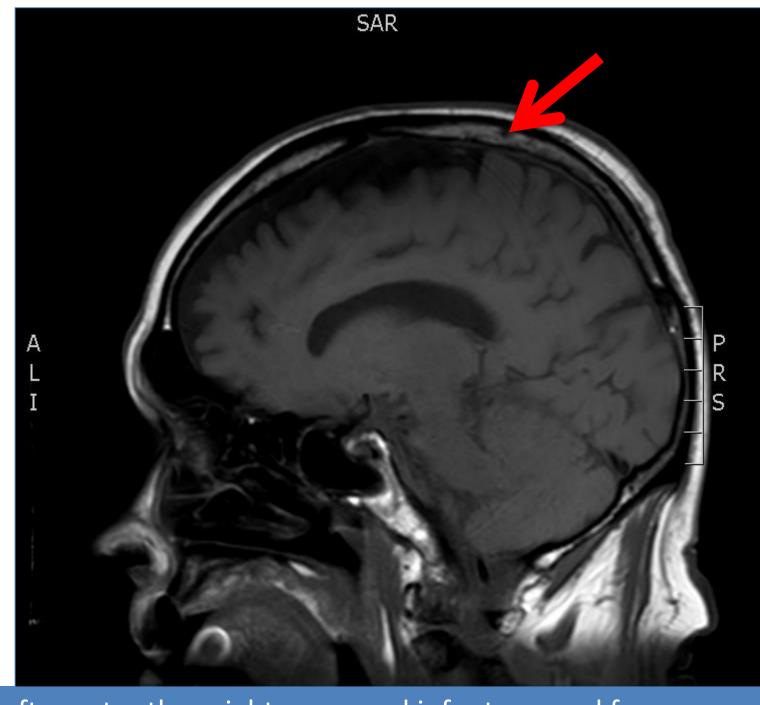


Figure 1: MRI Brain with and without contrast showing extensive pachymeningeal contrast-enhancement involving the left greater than right supra and infra temporal fossa.

## **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).</li>

#### VI. Brain biopsy

Chronic inflammation and small vessel vasculitis.

#### VII. Treatment

Azathioprine and prednisone.

#### VIII. Monitoring

Regularly attends rheumatology clinic.

### **Learning Points**

- 1) Hypertrophic Pachymengitis 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**

1. Horino T, Takao T, Taniguchi Y, Terada Y. Hypertrophic pachymeningitis with MPO-ANCA-positive vasculitis. *Clin Rheumatol*. 2010;29(1):111-113.

2. Kazem IA, Robinette NL, Roosen N, Schaldenbrand MF, Kim JK. Best cases from the AFIP: Idiopathic tumefactive hypertrophic pachymeningitis. *Radiographics*. 2005;25(4):1075-1080.

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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.