Teaching NeuroImage: Pachymeningitis and Aortitis as the Initial Presentation of Granulomatosis With Polyangiitis

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A 66-year-old man presented with a 1-year history of progressive cognitive decline, gait instability, and hearing loss. MRI of the brain showed extensive pachymeningeal thickening and enhancement with marked occlusion of intracranial dural venous sinuses (Figure 1). Lumbar puncture revealed pleocytosis at 140 cells/mm³ (96% lymphocytes) and elevated protein at 560 mg/dL. Serum syphilis screen, immunoglobulin G4, and proteinase 3 antibody were negative. Myeloperoxidase antibody was borderline positive. Systemic evaluation revealed evidence of aortitis and severe aortic regurgitation (Figure 2A). Dural biopsy was consistent with granulomatosis with polyangiitis (GPA).1 Special stains for microorganisms were negative (Figure 2, B–D). The patient was treated with intravenous methylprednisolone, followed by an oral prednisone taper. Rituximab was initiated.2 He returned in 2 months with near resolution of neurologic symptoms. Pachymeningitis can rarely be the initial manifestation of GPA, leading to cranial nerve impingement and venous outflow obstruction. Tissue biopsy and systemic evaluation are important in making the diagnosis.

Author Contributions
X. Li: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; analysis or interpretation of data. D. Stitt: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; analysis or interpretation of data. G. Lanzino: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; authorship contribution.

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Figure 1 Imaging at Initial Evaluation

(A, B) T1-contrasted weighted MRI showed diffuse pachymeningeal thickening and enhancement. (B) Enlarged diploic veins as accessory drainage pathways (arrow). Incidental fibrous dysplasia (arrowhead). (C) Diffuse stenosis of the venous sinuses on MR venogram (arrow). (D) Prominent ectasia of the optic nerve sheath on the left, suggesting intracranial hypertension.

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Figure 2 Systemic Imaging and Biopsy Results

(A) PET-CT showed increased fluorodeoxyglucose uptake at the root of the aorta (arrow).
(B) Dural biopsy showed extensive necrosis, macrophages infiltration with vague granulomatous features, and microabscesses (asterisks). (C) Area marked with asterisk under 200× magnification. (D) Area marked with double asterisks under 400× magnification.

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