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

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A 66-year-old man presented with one year history of progressive cognitive decline, gait instability and hearing loss. MRI brain showed extensive pachymeningeal thickening and enhancement with marked occlusion of intracranial dural venous sinuses (Figure 1). Lumbar puncture revealed pleocytosis at 140 cells/mm$^3$ (96% lymphocytes) and elevated protein at 560 mg/dL. Serum syphilis screen, IgG4, 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 2B-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 neurological 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.
Figure 1. (A-B) T1 contrasted weighted MRI showed diffuse pachymeningeal thickening and enhancement. 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.
Figure 2. (A) PET-CT showed increased FDG 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 200x magnification; (D) Area marked with double asterisks under 400x magnification.
Reference


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