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

Author(s):
Xiaoyang Li¹; Derek Stitt, M.D.¹; Giuseppe Lanzino, MD²; Caterina Giannini, MD, PhD³; Divyanshu Dubey¹,³; Ivan D Carabenciov, MD¹

Corresponding Author:
Ivan D Carabenciov, carabenciov.ivan@mayo.edu

Affiliation Information for All Authors: 1. Department of Neurology, Mayo Clinic, Rochester; 2. Department of Neurosurgery, Mayo Clinic, Rochester; 3. Department of Pathology and Laboratory Medicine, Mayo Clinic, Rochester

Neurology® Published Ahead of Print articles have been peer reviewed and accepted for publication. This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes.
Equal Author Contribution:

Contributions:
Ivan D Carabenciov: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data; Additional contributions (in addition to one or more of the above criteria)
Xiaoyang 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; Additional contributions (in addition to one or more of the above criteria)
Derek 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; Additional contributions (in addition to one or more of the above criteria)
Giuseppe Lanzino: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data; Additional contributions (in addition to one or more of the above criteria)
Caterina Giannini: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data; Additional contributions (in addition to one or more of the above criteria)
Divyanshu Dubey: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data; Additional contributions (in addition to one or more of the above criteria)

Figure Count:
2

Table Count:
0

Search Terms:
[ 132 ] Autoimmune diseases

Acknowledgment:

Study Funding:
The authors report no targeted funding.
A 66-year-old man presented with a 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). 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. 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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Neurology published online August 31, 2023
DOI 10.1212/WNL.0000000000207762

This information is current as of August 31, 2023

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