Teaching NeuroImages: Papilledema and Pachymeningitis: An Atypical Presentation of Granulomatosis With Polyangiitis

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A 53-year-old man presented with worsening headaches and progressive vision loss. His medical history was notable for biopsy-proven granulomatosis with polyangiitis (GPA) which was diagnosed after an episode of fever and hemoptysis years prior to the current presentation.

Ophthalmologic examination was remarkable for bilateral papilledema (Figure 1). Brain MRI with and without contrast demonstrated pachymeningitis and ventriculomegaly. Following ventriculoperitoneal shunt placement, there was resolution of headaches, papilledema and ventriculomegaly (Figure 2). He was subsequently treated with rituximab. Pachymeningitis - pathological thickening of the dura mater - stands as a rare manifestation of GPA. The most common manifestation of pachymeningitis is cranial nerve paralysis, but it can also hinder venous sinus drainage and cause a communicating hydrocephalus.
## Appendix 1. Authors

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[Teaching Slides -- http://links.lww.com/WNL/B409]
References


Figure 1 Title: Pachymeningitis, ventriculomegaly, and papilledema in a patient with granulomatosis with polyangiitis

Figure 1 Legend: (A) Post-contrast T1-sequence MRI axial view demonstrating meningeal enhancement and ventriculomegaly. (B) Post-contrast T1-sequence MRI coronal view. (C) Elevated optic disc with blurred margins and surrounding flame hemorrhages consistent with papilledema.
Figure 2 Title: Resolution of ventriculomegaly and papilledema after ventriculoperitoneal shunt placement

Figure 2 Legend: (A) Post-contrast T1-sequence MRI axial view demonstrating persistent meningeal enhancement and normal-sized ventricles. (B) Post-contrast T1-sequence MRI coronal view. (C) Resolution of papilledema.
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