A 71-year-old woman presented to the general medicine floor with a two-week history of profound fatigue and anorexia in the setting of chronic bilateral vision loss. Ten months earlier, she experienced sudden painless unilateral vision loss in her right eye. This was followed by progressive loss of visual acuity in her left eye culminating in complete vision loss three months before admission. Once hospitalized, she was started on corticosteroid therapy and subsequently regained partial vision in her left eye. Review of systems revealed a history of chronic sinus infections and progressive unilateral right-sided hearing loss.

Upon admission, she was afebrile and her vital signs were normal. Physical examination demonstrated a right afferent pupillary defect with marked loss of visual acuity in the right eye and decreased visual acuity in the left eye. The remainder of the physical examination was unremarkable. Laboratory testing demonstrated markedly elevated inflammatory markers, including erythrocyte sedimentation rate of 67 mm/hr and C-reactive protein of 195.3 mg/L. Antinuclear antibody, anti-double-stranded DNA IgG, proteinase 3 (PR3), Lyme titers, Epstein-Barr virus, CMV, toxoplasmosis, HIV, syphilis, West Nile virus and angiotensin-converting enzyme tests were unremarkable. Cerebrospinal fluid studies showed IgG index of 0.58, 3 oligoclonal bands and normal myelin basic protein. Anti-myeloperoxidase antibody was positive at 1.9 U and p-ANCA was positive. The c-ANCA was negative at the time of discharge.

Magnetic resonance imaging revealed diffuse dural enhancement over the cerebral hemispheres (arrow). Diffuse dural enhancement is shown over the cerebral hemispheres (arrow).

Discussion

Idiopathic pachymeningitis is a rare clinical entity that has only been described in a few cases. Its presentation is variable but it often has features of a limited-type vasculitis with myeloperoxidase and ANCA positivity. It appears to have a predilection for the cranial nerves and has been particularly implicated in optic neuropathies, visual field losses and blindness. The disease responds to corticosteroids, which are often continued long-term because recurrence is common with treatment tapers. If untreated, it results in progressive neurological dysfunction.

Since MPO-ANCA vasculitis pachymeningitis is an uncommon problem, it was ultimately a diagnosis of exclusion for this patient. However, it was also the diagnosis most consistent with the clinical scenario and evidence. A lesson learned during the care of this patient was to persist when patients present with definitive unexplained symptoms.

REFERENCES