In this report we describe nongranulomatous uveitis followed by bilateral retinal vasculitis and much later by the loss of accommodation as initial presentations of demyelinating disease in a 42-year-old female with no other neurologic manifestations. The absence of demyelinating plaques in the initial magneric resonance imaging (MRI) (orbit and cranium) and its occurrence 2 years later, have been described as “lesions appearing with time”. Extensive laboratory investigations ruled out infections, systemic vasculitis, and connective tissue disorders. Due to the presence of oligoclonal bands in both cerebrospinal fluid (CSF) and serum, absence of anti-aquaporin-4, antemyelin-oligodendrocyte glycprotein immunoglobulin G (IgG) antibodies, and negative vasculitis profile, the exact cause of demyelination (multiple sclerosis/vasculitis related) could not be ascertained. She has currently received 2 cycles of rituximab and at the last follow-up did not show any recurrences.

**Key words:** Accommodation, demyelinating plaques, oligoclonal bands, retinal vasculitis, rituximab

We report a rare case of nongranulomatous uveitis followed by retinal vasculitis and much later by the loss of accommodation and red-green desaturation as sole manifestations of a later onset demyelinating disease in a middle-aged lady.

**Case Report**

A 42-year-old female presented to us first in July 2015 with nongranulomatous anterior uveitis in the left eye (OS) and increase in intraocular pressure (IOP) due to primary angle closure in both eyes (OU). She had received topical prednisolone acetate eye drops along with timolol 0.5% eye drops (OU). Nd-Yag Peripheral iridotomy was done (OU) after the resolution of uveitis. Her BCVA was 20/20(OU) and the fundus examination showed normal optic nerve head (OU) and a small scar near the fovea (OD). Her baseline visual fields were normal. She presented to us again 2 years later with blurring in vision (OS) of 2 days duration. Slit lamp and IOP examinations were normal. The fundus examination now showed cuffs of retinal vasculitis in the midperipheral retina (OU). [Fig. 1a] Occasional vitreous cells were noted. Fluorescein angiography showed patchy areas of fuzzy hyper fluorescence corresponding to the cuffs of perivasculitis. [Fig. 1b] There was no optic nerve head leakage. Optical coherence tomography (OCT) showed a hyperreflective scar near fovea (OD). Laboratory investigations included a normal complete hemogram, a negative Mantoux test and QuantiFERON TB gold test, non-reactive Venereal disease research laboratory (VDRL) tests, and treponema pallidum hemagglutination test (TPHA) tests, normal level of serum angiotensin-converting enzyme, negative antinuclear antibody profile, antineutrophilic cytoplasmic antibody, and lupus anticoagulant. Weil Felix test, ELISA for toxoplasma, Lyme and HIV were negative. There was a slight elevation of C-Reactive protein (7.9 mg/L; normal: <6) and serum homocysteine (16.54 µmol/L; normal: 4.4-13.6). Computed tomography (CT) of the thorax was normal. Visual fields now showed some paracentral scotomas and depressed areas. [Fig. 2] Color vision, pupillary examinations, and magnetic resonance imaging (MRI) of orbits and cranium did not reveal any abnormality. [Fig. 3] She was started on oral steroids (1 mg/kg body weight), tapered slowly over 6 months based on the ocular inflammation. Repeat visual fields showed improvement.

One year later, she complained of sudden blurring of vision (OS) of 4 days duration. Her BCVA was 20/20, N/6 (OD), and 20/20p, N/18 (OS). Slit lamp, IOP, pupillary, fundus, and color vision examinations did not reveal any significant abnormality. Loss of accommodation was noted (OS). Visual fields now showed paracentral areas of depressed sensitivities (OU). The left eye showed slight red-green desaturation. Visual evoked potentials showed a normal amplitude and slight prolonged P100 latency (111.6 ms in OD and 113.1 ms in OS). Her MRI cranium now showed few nonenhancing T2 and FLAIR hyperintensities in periventricular white matter, centrum semiovale and subcortical white matter of left frontal lobe arranged perpendicular to the long axis of the ventricular system as well as in callosseptal interface likely suggestive of demyelinating plaques. Oligoclonal bands were seen both in the CSF and serum. Antiaquaporin-4 and...
antimyelin oligodendrocyte glycoprotein IgG antibodies were negative. Cervical spine MRI showed 2 small T2 hyperintense foci in the upper medulla and at the cervicomedullary junction. She received intravenous methyl prednisolone 1 gm/day for 3 days followed by a tapering schedule of oral steroids over 2 weeks. Detailed neurological examination showed no involvement of sensory, motor pathways, or brainstem. Repeat visual fields post steroids showed improvement and BCVA was 20/20, N/6 (OU). On the advice of the neurologist, intravenous rituximab was initiated. She received 2 cycles (each cycle comprised of an initial dose of 2 gm followed by 1 gm a month later. The second cycle was repeated after 6 months) of intravenous rituximab. At last follow-up (1 year after rituximab), her BCVA was 20/20, N/6 (OU) with no new ocular symptoms. Repeat MRI cranium and spine did not show any new lesions.

**Discussion**

Multiple sclerosis (MS) is a neuroinflammatory condition characterized by repeated, focal demyelination in the central nervous system. It can be the more common relapsing, remitting type or the chronic primary progressive type. Ocular manifestations include optic neuritis, oculomotor abnormalities, and uveitis. Retrospective studies show approximately 1% of patients with uveitis carry diagnosis of MS and approximately 1% of MS develop uveitis. Uveitis is a rare presentation in MS and includes anterior uveitis, intermediate uveitis, and retinal vasculitis.

Our case is unique as she presented initially with uveitis, followed by retinal vasculitis and much later by the loss of accommodation with the development of demyelinating plaques. She had no systemic signs of neurological disease. This is rare and has not been described in Indian literature (Medline search). The absence of anti-aquaporin-4, antimyelin oligodendrocyte glycoprotein IgG antibodies, and presence of oligoclonal bands in both CSF and serum in our case raises doubts regarding the nature of the demyelinating disease – Is it an atypical form of MS or is there a vasculitis component?

There was a significant improvement of ocular symptoms and signs with systemic steroids on both occasions. As rituximab is a useful drug in both MS and vasculitis, it was considered by the treating neurologist. After 2 cycles of rituximab, 6 months apart, visual fields and MRI orbits and cranium were stable, with no onset of new lesions.

**Conclusion**

To conclude, this case highlights the initial presentation of ocular involvement as uveitis, retinal vasculitis, and loss of accommodation at different time frames, preceding the onset of demyelinating disease in a middle-aged lady with no other neurologic involvement.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Olsen TG, Frederiksen J. The association between multiple sclerosis and uveitis. Surv Ophthalmol 2017;62:89-95.
2. Cunningham ET Jr, Pavesio CE, Goldstein DA, Forooghian F, Zierhut M. Multiple sclerosis-associated uveitis. Ocul Immunol Inflamm 2017;25:299-301.

3. Pedraza-Concha A, Brandauer K, Tello A, Rangel CM, Scheib C. Bilateral anterior and intermediate uveitis with occlusive vasculitis as sole manifestation of relapse in multiple sclerosis. Case Rep Ophthalmol Med 2019:8239205.

4. Shields MK, Simon S, Chan W, Gilhotra JS. Acute hypertensive uveitis as the first presentation of multiple sclerosis. Indian J Ophthalmol 2019;67:163-4.