Steroid Responsive Acute Isolated Ophthalmoplegia: A Rare Presentation of Anti-Gq1b Antibodies Syndrome

Sir,

A 24-year-old male presented with 10 days of bilateral ptosis with binocular diplopia which developed overnight. [Figure 1] There was no worsening or fluctuation of diplopia. He had no headache, confusion, seizures, or loss of consciousness. There was no prodromal fever, diarrhea, urinary tract infection, vaccination, dog bite, etc. He had been treated with cyclophosphamide, oncovin, procarbazine, and prednisolone regimen for Hodgkin's disease at the age of 5 years and was disease-free since. On examination, he had bilateral partial ptosis with mid-dilated, sluggishly reactive pupils with complete external ophthalmoplegia. Remaining neurological and systemic examination was normal. Clinical and repetitive nerve stimulation tests for fatigability were negative. His routine investigations were normal. He was positive for HbsAg and anti-HCV antibody. Serology for HIV, vasculitis profile (ANA, dsDNA, RA factor, ANCA), thyroid function test (including thyroid antibodies) was negative. Chest X-ray, abdominal ultrasonography, and whole body PET scan were normal. Cerebrospinal fluid (CSF) showed normal opening pressure, acellular with normal protein (29 mg%) and sugar (60 mg%, concomitant blood sugar 110 mg%.). CSF was negative for gram stain, culture, Ziehl-Nelson stain for acid fast bacilli, TB PCR, Gene Xpert, fungal smear, India Ink, cryptococcal antigen, and cytology for malignant cells. Nerve conduction studies and contrast MRI brain/orbit were normal. Anti-GQ1b IgG antibody was positive. He was administered intravenous methylprednisolone (1 gm/day for 5 days) followed by oral prednisolone at 1 mg/kg for 4 weeks and tapered off over the next 4 weeks. He recovered completely within this period.

The association of anti-GQ1b antibodies with acute isolated ophthalmoplegia (AIO) without ataxia and areflexia, termed atypical Miller Fisher syndrome, was first described by Chiba *et al*.[1] and belongs to the spectrum of anti-GQ1b antibody syndrome.[2]

The pathogenesis involves molecular mimicry between capsular antigens on campylobacter jejuni and peripheral nerve myelin, with resultant pathogenic anti-ganglioside antibodies. GQ1b gangliosides are densely concentrated in extramedullary paranodal regions of oculomotor, trochlear, and abducens nerves, which explains their involvement in anti-GQ1b antibody mediated diseases. CSF protein levels were normal in our patient. Albuminocytologic dissociation is a characteristic feature in GBS and MFS. In the largest retrospective study of 34 patients of anti-GQ1b syndrome by Lee *et al*., 11 had AIO, 13 had MFS, 6 had GBS with ophthalmoplegia, and one had BBE.[3] Normal CSF protein was present in 8 out of 11 patients with AIO which may be due to inflammation restricted to cranial nerve roots. Odaka *et al*. in 2001 proposed diagnostic criteria for acute ophthalmoplegia:

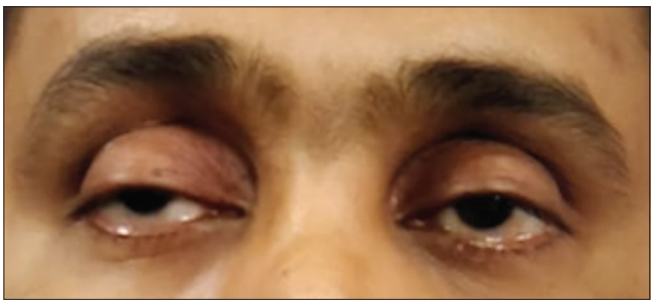

Figure 1: At presentation, the patient had bilateral ptosis and external ophthalmoplegia

progressive, relatively symmetric acute ophthalmoplegia by 4 weeks without ataxia or limb weakness.[2] Features that strongly support the diagnosis of AIO includes history of infectious symptoms 4 weeks prior to the onset of neurological symptoms, CSF albuminocytological dissociation, and presence of anti-GQ1b IgG antibody. AIO has been treated with intravenous immunoglobin (IVIG) and plasma exchange[4] but spontaneous recovery has also been reported.[5,6] Literature does not mention use of steroids for AIO. Our case responded to steroids with complete recovery.

AIO with positive anti-GQ1b antibody is a rare immune-mediated syndrome, which can be treated cost-effectively with steroids.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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