Visual loss with papilledema in Guillain-Barre syndrome

P.S. Kharbanda, S. Prabhakar, V. Lal, C.P. Das

Summary

Papilledema and raised intracranial pressure have been reported in association with Guillain-Barre syndrome. Papilledema is usually asymptomatic or associated with mild visual field defects, without any visual loss. The cerebrospinal fluid protein is usually reported to be high. A case of a 35 year old lady is reported, who presented with headache, diplopia and progressive visual loss in both eyes and limb weakness with hyporeflexia. Optic fundus examination showed bilateral papilledema. She had features of pseudotumor cerebri. Nerve conduction studies were suggestive of polyradiculopathy. The unusual things in this case, were the profound visual loss, normal cerebrospinal fluid protein and the presentation of papilledema before the limb weakness.

Key words : Guillain-Barre syndrome, papilledema, Pseudotumor cerebri, visual loss.

Introduction

Guillain-Barre syndrome (GBS) or acute inflammatory demyelinating poly neuropathy, usually presents as an areflexic ascending quadriparesis. However atypical and incomplete manifestations are not uncommon. A number of cranial nerves can be involved, facial nerve being the commonest. Papilledema has been reported as a rare complication of GBS. Although blurred discs during the course of GBS were reported earlier, initial reports of definite papilledema appeared in 1936.1 Since then a number of case reports of this entity can be found in literature. Significant among them is a series of four cases by Morley et al.2 Some initial reports do not have the cerebrospinal fluid (CSF) pressure measurement, but later increased CSF pressure has been reported and these cases were classified as pseudotumor cerebri. Pseudotumor cerebri has also been reported in cases of GBS associated with human immunodeficiency virus infection.3 Communicating hydrocephalus has also been reported in GBS.4 Papilledema is usually seen after the limb weakness and is associated with elevated CSF protein. We are aware of only one report of GBS in which pseudotumor cerebri has been documented and the CSF protein was normal.5 The papilledema usually subsides gradually and visual loss has not been reported before.

Case Report

A 35 year old lady presented with continuous and bilateral headache for three weeks prior to admission. This was followed by binocular diplopia which was initially present on looking to the left, but after one week, diplopia was present in all directions. At the same time she started having progressive visual impairment in both the eyes. There was no retro orbital pain or pain on moving the eyes. There was total visual loss within one week. One week prior to admission, i.e., two weeks after the onset of symptoms, she developed progressive weakness of all the four limbs, which was symmetrical and was not associated with any radicular pain, wasting or fasciculations. Sensory symptoms were absent. There was no involvement of urinary bladder or bowel. She did not complain of any respiratory distress. On examination she had hypertension (BP-160/110mmHg), and mild tachycardia (pulse-108/min). Central nervous system examination revealed normal higher functions. She had bilateral ptosis, lateral rectus palsy and papilledema. Visual acuity was reduced to just projection of light in both the eyes. Other cranial nerves were normal. Speech was not involved. Motor examination revealed normal tone and bulk of all limbs. No fasciculations or any other abnormal movements were noted. Power was grade 3/5 in proximal and 4/5 to 4+/5 in distal muscles in both upper and lower limbs. Deep tendon jerks were absent and plantars showed flexor response. There was no sensory loss or signs of cerebellar dysfunction. Investigations revealed a normal hemogram and biochemical profile. Cerebrospinal fluid examination revealed raised pressure (420 mm of CSF) and no leukocytes. CSF protein was 20 mg/dl and sugar 76 mg/dl. Gram stain, bacterial culture, VDRL, malignant cytology, cryptococcal antigen, acid-fast bacilli, and flowcytometry were negative in the CSF. Antinuclear antibody, LE cell, antinuclear cytoplasmic antigen, and rheumatoid factor were negative in blood. Visual evoked responses did not show any wave formation and brainstem auditory evoked responses were normal. CT scan and MRI of the brain were unremarkable. Nerve conduction studies showed absence of F waves in upper and lower limbs. A diagnosis of acute inflammatory demyelinating polyradiculopathy with pseudotumor cerebri was made. She was started on treatment with acetazolamide and corticosteroids. Metoprolol was given for the autonomic dysfunction. Her headache
slowly subsided. Repeat optic fundus examination showed some features of anterior ischaemic optic neuropathy. This was thought to be secondary to the raised intracranial pressure. She started having a gradual recovery while she was in the hospital.

Discussion

The association of papilledema with acute inflammatory demyelinating poly neuropathy has been described in literature. In most of these reports the papilledema appeared after there was an established limb weakness, however, in the present case the papilledema preceded the limb weakness. The CSF proteins were elevated in the cases reported earlier. These elevated proteins were postulated to cause a defect in the proper absorption of CSF at the arachnoid villi, giving rise to raised intracranial pressure.6,7 Few cases of hydrocephalus associated with GBS have also been explained on the basis of the same theory. Joynt, however, postulated that the papilledema may be secondary to cerebral edema.8 Edema of the spinal nerve rootlets seen in GBS has been implicated in causing decreased absorption of proteins at this level and contribute to the raised CSF protein.4 Pseudotumor cerebri has also been reported with acute inflammatory demyelinating polyradiculoneuropathy (AIDP) but there is only one reported case of raised intracranial pressure in a patient of AIDP, where the CSF protein was normal.5 Our case had all the features of a pseudotumor cerebri, but with normal CSF proteins. In the earlier reported cases the papilledema was an asymptomatic feature and was detected only during the routine examination of the optic fundus, but in the index case this progressed to a near total visual loss. This prompted us to think that elevated CSF protein may not be the only mechanism for the increased intracranial pressure in patients of Guillain Barre syndrome and this raised intracranial pressure can even lead to visual loss. This also means that patients of GBS, especially the ones who develop papilledema, need to be closely followed up to detect any field defects or visual loss, so that timely intervention can be initiated.

References