Neurology Asia 2018; 23(2) : 189 – 191

Headache associated with polyneuritis cranialis

Sang-Soo Lee *MD*, Soyeon Kwon *MD*

Department of Neurology, Chungbuk National University College of Medicine, Cheongju, Korea

Abstract

Polyneuritis cranialis (PNC), a variant of Guillain-Barré syndrome (GBS), is a very rare disorder of multiple cranial nerve palsies without any peripheral nerve involvement or ataxia. While pain is frequent in GBS, headache is not known in the patient with PNC. We report a 72-year-old man who presented acute bilateral multiple cranial neuropathy, including neuropathies of cranial nerve III-XII and severe and prolonged headache. The brain MRI, cerebrospinal fluid examination, and peripheral nerve conduction studies showed no abnormalities. The headache improved in five weeks. All cranial neuropathies resolved completely 8 months later. We suggest that headache should be considered as an additional cardinal feature of PNC along with multiple cranial neuropathies.

Keywords: Polyneuritis cranialis; Guillain-Barré syndrome; headache

INTRODUCTION

The various forms of Guillain-Barré syndrome (GBS) have been considered by some to be a continuum and by others to be distinct entities. Polyneuritis cranialis (PNC) has been used to describe some patients who develop multiple cranial neuropathies at once and exhibit clinical features consistent with GBS, but do not fulfill diagnostic criteria for Miller Fisher syndrome (MFS) or other GBS subtypes with prominent cranial nerve involvement.[1] Recently, a new diagnostic classification for GBS and MFS based on the clinical features was proposed. However, a case of PNC has fallen out of the new classification and this term has not appeared in the recent definition.[2] The incidence of PNC remains unknown, but it is thought to be much smaller than other forms of GBS, as there are few cases in the literature. The rarity of the PNC has made it more difficult to characterize the PNC.

Pain is not uncommon in GBS, being present in 89% of patients[3], but much less is known about headache as a part of pain syndromes in GBS. Its etiology is poorly understood. Moreover, headache was not known in PNC patients. We report a case of prolonged headache with PNC without ataxia or any other peripheral nerve involvement.

CASE REPORT

A previously healthy 72-year-old man was hospitalized because of diplopia for 3 days. His prodromal symptoms included watery diarrhea for 5 days. On admission, he was alert but had bilateral blepharoptosis, bilateral facial palsy, severe difficulty in speaking and swallowing and inability to protrude the tongue and open the mouth. Pupillary function was intact. Eye movements were completely limited in all directions of gaze. Trapezius weakness was also noted. However, neither limb nor truncal ataxia was present. He was able to walk independently without any limb weakness. All deep tendon reflexes were preserved, and no pathological reflexes were elicited. Sensory and autonomic functions were intact. Routine blood chemistry tests including vitamin B_{12} and folic acid and thyroid function test yielded normal results. The cerebrospinal fluid examination showed 12 cm H_2O opening pressure, no cells, a 43 mg/dl protein concentration on admission, and follow-up findings after 1 week remained normal. Contrast enhanced MRI of the brain showed no abnormalities. Anti-ganglioside GM-1 IgM and IgG antibody levels were negative, as were anti-ganglioside GQ1b and GD1b, and anti-Ach-R antibodies. Blink reflex test showed no visible waves. Direct facial nerve stimulation revealed markedly decreased amplitudes of the compound action potentials, whereas nerve conduction studies in the extremities yielded normal results. Brainstem auditory evoked potentials study showed poorly consistent wave I bilaterally. In addition to neurological symptoms, he complained of headache from the day after hospitalization. He had no prior history of headaches. It radiated

Address correspondence to: Sang-Soo Lee, MD, Department of Neurology, Chungbuk National University College of Medicine, 1 Chungdae-ro, Seowon-gu, Cheongju 28644, Korea. Tel: +82-43-269-6336, E-mail: sslee@chungbuk.ac.kr

up from the neck to the front of the head. The headache was persistent and the intensity was variable (8~10 on visual analog scale). At times, headache was so severe that he could not fall asleep. There was no photophobia, nausea, or vomiting. Examination of his fundi revealed no abnormality. Nonsteroidal anti-inflammatory drugs or narcotics did not provide significant relief. After that, gabapentin was administered. Clinical improvement started at 4 weeks. The headache gradually improved, being controlled by gabapentin, and it resolved completely after 5 weeks. The patient underwent intravenous immunoglobulin treatment for five days, 0.4 g/kg. Six weeks after the onset of illness, his symptoms gradually diminished. Eight months after his initial symptoms he completely recovered.

DISCUSSION

There are two notable features in this patient. One is a widespread cranial neuropathy, including neuropathies of cranial nerve III-XII. The other is a prolonged headache during the course of the illness. Our patient has some characteristics of GBS, such as symmetric weakness, monophasic clinical course, favorable outcome, and history of antecedent infectious symptoms. Other disease processes affecting the meninges or cranial nerves were excluded. Classification of PNC is made difficult because it appears at the borderland between GBS and MFS.[1] PNC could be an overlapping form of MFS and pharyngeal-cervical-brachial weakness (PCB) or suggested as an oculopharyngeal variant of GBS if the facial nerve was spared.[4] However, there remains some ambiguity as to how best classify a small set of patients, who develop multiple cranial neuropathies in the absence of ataxia or limb weakness. Phenotypically, the majority of patients diagnosed with PNC displayed facial weakness, which has only rarely been reported in association with MFS, and not reported with acute pharyngeal weakness. Therefore, the pattern of weakness in PNC extends beyond what would be predicted from the overlap between MFS and PCB.[2] Furthermore, facial weakness in these patients cannot be attributed to bifacial weakness with paresthesias, which is caused by demyelinating neuropathy and not associated with antiganglioside antibodies.[5] For the above reasons, PNC was proposed as a separate subtype altogether, which lies at the interface between MFS and GBS.[2]

By far the most frequent location of pain in GBS patients was in the extremities[6], whereas orbital pain was reported in the three patients out of 27 patients with MFS.[7] The distribution of muscle weakness seems to be related to the location of the pain. However, surprisingly, the largest prospective study of pain to date in GBS did not include headache as a type of pain location, further suggesting its lesser prominence in GBS.[6,8] The review of 50 consecutive patients with MFS did not describe headache or pain.[9] Moreover, no reports exist to our knowledge that describe headache with other GBS variants. The precise mechanisms of headache in GBS remain uncertain and are thought to be diverse. Several clinical settings where headache in GBS occurs were known: the posterior reversible encephalopathy syndrome, the rare cases where papilloedema and increased intracranial pressure occur after GBS, and MFS.[8] Other potential explanation for headache is an activation of the trigeminovascualr pain pathway resulting from the serum autoantibodies that cause the disease.[10] However, our patient's headache cannot be explained by any of the above mechanisms, although antecedent pathogens and target molecules of autoantibodies still remain unknown in most PNC cases. The severe headache in PNC seems to have a different mechanism. Headache may be considered as an additional cardinal feature of PNC along with multiple cranial neuropathies and needs further evaluation and proper management.

DISCLOSURE

Conflict of interest: None

REFERENCES

1. Wakerley BR, Yuki N. Polyneuritis cranialis: oculopharyngeal subtype of Guillain-Barré syndrome. *J Neurol* 2015;262:2001-12.
2. Wakerley BR, Yuki N. Polyneuritis cranialis-subtype of Guillain-Barré syndrome? *Nat Rev Neurol* 2015;11:664
3. Moulin DE, Hagen N, Feasby TE, Amireh R, Hahn A. Pain in Guillain-Barré syndrome. *Neurology* 1997;48:328-31.
4. Dosi R, Ambaliya A, Patel N, Shah M, Patell R. Acute multiple cranial neuropathy: An oculopharyngeal variant of Guillain-Barré syndrome. *Australas Med J* 2014;7:376-8.
5. Susuki K,koga M, Hirata K, Isogai E, Yuki N. A Guillain-Barré syndrome variant with prominent facial diplegia. *J Neurol* 2009;256:1899-905.
6. Ruts L, Drenthen J, Jongen JL, *et al*. Pain in Guillain- Barré syndrome: a long-term follow-up study. *Neurology* 2010;75:1439-47.

7. Koga M, Yuki N, Hirata K. Pain in Miller Fisher syndrome. *J Neurol* 2000;247: 720-1.
8. Farmakidis C, Inan S, Milstein M, Herskovitz S. Headache and pain in Guillain-Barré syndrome. *Curr Pain Headache Rep* 2015;19:40-6
9. Mori M, Kuwabara S, Fukutake T, Yuki N, Hattori T. Clinical features and prognosis of Miller Fisher syndrome. *Neurology* 2001;56:1104-6.
10. Friedman DI, Potts E. Headache associated with Miller Fisher syndrome. *Headache* 2007;47:1347-8.