A Case of Guillain-Barre Syndrome with Significant Lymphocytic Pleocytosis

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ABSTRACT

Guillain-Barre syndrome (GBS) is an immune-mediated polyneuropathy. In the cerebrospinal fluid (CSF) characteristic increased protein without pleocytosis i.e., albuminocytologic dissociation is seen. But cases of GBS with significant CSF lymphocytosis have rarely been reported. Here, we present a 38-year-old man with dysarthria, dysphagia, bilateral facial palsy, and limb weakness with the diagnosis of GBS. Nerve Conduction Study confirmed the diagnosis but interestingly in the CSF study increased protein and lymphocytic predominant pleocytosis were found. Other possible differential diagnoses were ruled out by appropriate paraclinical studies. The patient underwent treatment with intravenous immunoglobulin and showed favorable response.

Keywords: Guillain-Barre syndrome; lymphocytosis; cerebrospinal fluid

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INTRODUCTION

Guillain-Barre syndrome (GBS) is an immune-mediated acute polyradiculoneuropathy characterized by motor weakness, areflexia, paresthesia with minor sensory loss, and increased protein in the cerebrospinal fluid (CSF) without pleocytosis (albuminocytologic dissociation) [1]. The diagnosis of GBS depends on clinical criteria supported by electrophysiological studies and CSF findings. It is known as a heterogeneous disorder with different variants with characteristic demyelinating pattern [2]. The most common form is acute inflammatory demyelinating polyneuropathy (AIDP), while other variants including acute motor axonal neuropathy (AMAN), acute motor and sensory axonal neuropathy (AMSAN), and Miller Fisher syndrome are less common [1,2]. Over fifty percent of the patients with GBS may show cranial nerve involvement usually as facial weakness. CSF analysis and electrodiagnostic (EDX) studies are critical for confirming or excluding the diagnosis of GBS. Other laboratory studies have limited value [3,4]. Here, we report a 38-year-old man as a rare and atypical case of GBS.

CASE PRESENTATION

A 38-year-old man referred to emergency room of Shiraz Nemazie Hospital with problem in speaking, eating and performing facial expression such as laughing, and also distal extremities paresthesia and burning sensation since 10 days prior to admission.

The patient did not mention any sign of dysphagia, dyspnea and extremity weakness and urinary problem. During the last few weeks the patients showed no sign or symptom of upper respiratory infection or gastroenteritis. Vital signs of the patient were normal, without any respiratory depression or respiratory difficulty. The neurological exam revealed normal mental status. Bilateral severe peripheral facial palsy was detected but other cranial nerves were normal. The patient's muscle power was normal except very mild weakness in the proximal muscles of the lower extremities. However, all of the upper and lower extremities deep tendon reflexes were decreased. The plantar reflex was downward. The sensory

examination revealed hypoesthesia of the distal upper and lower limbs, but no sensory level was found. Cerebellar tests and patient's gait were normal. The patient had no neck rigidity and meningeal signs were negative. The other components of physical examination were normal.

With impression of Guillain-Barre syndrome, EMG-NCV was performed for the patient. The EMG-NCV showed prolonged F latency, decreased conduction velocity (26 and 31 meter/second) in both tibial nerves and inconsistency of the right proneal F wave and significantly prolonged distal latency of both facial nerves, all of them being in favor of GBS. Then, lumbar puncture was done. CSF analysis revealed significant pleocytosis (total WBC count 170, 90% lymphocyte), high protein (268 mg/dl) and normal glucose. The viral markers including HIV, HBs Ag, HCV Ab and CSF polymerase chain reaction of HSV, EBV and CMV were negative. Also, vasculitis work ups including ANA, ANCA and DSDNA were normal. All other laboratory work ups including complete blood count and routine work ups were normal.

At the third day of hospital admission, treatment with IVIG was started. The patients received a total dose of 2gr/kg IVIG divided in 5 consecutive days. Then, the patient was discharged with improvement in distal extremities paresthesia and burning sensation. After a 3-month follow up, all the patients' symptoms were relieved.

DISCUSSION

Abnormalities of basic CSF analysis including cell count and CSF protein level in GBS are commonly caused by dysfunction of blood-CSF barrier which can be shown as an elevated CSF protein concentration. However, raised cell count or intrathecal produced immunoglobulin as depicted by oligoclonal IgG or by IgG-Index usually cannot be found [1,3]. Akbayram and his colleagues reported the results from 35 patients with GBS for whom lumbar puncture was performed. The CSF cell count was normal (<10 cells/mm^3) in all the patients [5]. In this case, we presented a 38-year-old man with significant CSF pleocytosis. As mentioned in Asbury's et al. study in GBS, prominent elevation of cell counts or positive polymorph nuclear granulocytes in CSF could raise concern about different diagnoses [6]. Myelitis, caused by HSV 1 and 2, EBV, VZV and CMV can clinically mimic GBS, and in the early disease course CSF may show mild to moderate pleocytosis with a significant proportion of polymorphonuclear granulocytes [6].

In this case, 10 days after initial symptoms when the patients developed severe bilateral facial palsy, lumbar puncture was done and increased cell count with lymphocyte dominancy was found. Erythrocyte sedimentation rate and C-reactive protein as the indicators of infection or inflammatory process were all within normal ranges. Viral markers were measured for additional evaluation, all of which were negative. So based on the history, physical exam and also the laboratory data, other infections which were mentioned above were ruled out. As Rauschka and his colleagues concluded that in classic manifestations of GBS, atypical CSF finding including pleocytosis more than 50 cells or polymorphonuclear granulocytes could not exclude GBS [7]. However, infectious and autoimmune disorders should be considered in atypical CSF findings. Likewise, in our study we concluded that the abnormal findings in CSF do not contribute to other suspected disorders and do not rule out GBS.

CONCLUSION

GBS is an acquired multi-focal and segmental demyelinating polyradiculoneuropathy. CSF usually shows cell-protein dissociation, but rarely significant CSF pleocytosis can be seen as an atypical variant.

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