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CASE REPORT

Multiple Cranial Neuropathy (A Teaching Case)



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Abstract

There are few reports of the multiple cranial neuropathy variant of Guillain-Barré Syndrome (GBS). Patients usually present with facial diplegia, lower cranial nerve involvement and hypo or areflexia. It is crucial to identify promptly this unusual cranial variant but the clinical characteristics remain poorly defined. This GBS variant usually has a rapid progressive course with respiratory muscle paralysis. Most of the patients recover well, although the process is slow. We report a 54 year old man presenting with facial diplegia, progressive ophthalmoplegia, lower cranial nerve involvement, sensory ataxia and generalized areflexia. This GBS variant is very unusual and seldom described in the literature; it is oftenly misdiagnosed. The clinical features and nerve conduction studies (absent F-waves, motor conduction block) provide evidence to support a diagnosis of an acute demyelinating polyneuropathy consistent with a regional cranial variant of GBS.

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1. Case presentation

A 54-year-old right handed man was seen in the emergency room with a three day history of right ptosis, intermittent double vision and numbness of both hands. His symptoms progressed over a three day period, he developed bilateral ptosis, permanent diplopia, neck weakness, nasal voice, sialorrhea and dysphagia, initially for solids and then for

liquids. Two weeks before admission, he had had an upper respiratory tract infection that resolved spontaneously. The patient reported a past history of pulmonary tuberculosis and occupational exposure to tantalum, baryte and iron dust. He had recently traveled to Israel, denied alcohol abuse, smoking or recent immunizations. His mother died at 35 years of a ruptured brain aneurysm.

On examination his blood pressure was 135/85 mm Hg, the pulse 82 beats per minute, the respiratory rate 20 breaths per minute and the temperature 36.8 °C. The neurological examination revealed normal mental status and dysarthria. Pupils were 3 mm in diameter and reactive to light. There was a bilateral ptosis, palsy of extraocular muscles (video 1) and a facial diplegia (video 2). An

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asymmetric palate and reduced gag reflex was also noted. On protrusion the tongue was slightly deviated to the right. The patient was unable to elevate his shoulders or extend his neck. Additionally, he had sensory ataxia and deep tendon reflexes were absent. Muscle strength was normal in all limbs (video 3).

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A complete blood count, chemistry panel, and levels of serum electrolytes were normal. His condition rapidly deteriorated, requiring endotracheal intubation and close monitoring in the Intensive Care Unit for 15 days.

2. The case diagnosis

This 54-year-old man presenting with a facial diplegia, progressive ophthalmoplegia, lower cranial nerve involvement, sensory ataxia and generalized areflexia had a magnetic resonance imaging (MRI) scan with gadolinium which revealed no abnormalities. A lumbar puncture was performed with an opening pressure of 18 cm of water. Cerebrospinal fluid (CSF) analysis showed a protein concentration of 31.59 mg/dL, 0 white blood cells per mm³ and a CSF-blood glucose ratio of 0.63. Gram stain, india ink and CSF cultures were all negative. Erythrocyte sedimentation rate, C reactive protein, thyroid function tests, liver enzymes, blood glucose, vitamin B12 and folic acid levels were normal. Glycated hemoglobin was 5.8%. Thorax CT was negative for sarcoidosis. CSF-VDRL and ELISA for HIV were both negative.

Nerve conduction studies that included the median, ulnar and tibial nerves, revealed a motor conduction block (distal and proximal CMAP-amplitude of 7.2 mV and 3.5 mV, respectively), abnormal temporal dispersion (Fig. 1A), diminished conduction velocity and absent F waves (Fig. 1B), favoring a diagnosis of Guillain-Barré syndrome variant. No decremental response to repetitive nerve stimulation was observed. The serum anti-GQ1b antibodies were negative.

Treatment with intravenous immunoglobulin was initiated at 0.4 mg/kg daily over 5 days. However, no clinical improvement was noted after a second cycle of immunoglobulin therapy. After being discharged from the ICU, the patient developed a feeling of tightness around his chest, life threatening respiratory muscular weakness and severe dysphagia leading to aspiration. Tracheostomy and gastrostomy tubes were placed.

He received physical therapy and the symptoms partially resolved without further intervention. Enteral feeding was discontinued and patient was discharged 3 months after the admission with a Hughes scale of 2 (Videos 4 and 5).

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3. Discussion

Acute multiple cranial neuropathy, sensory ataxia and generalized areflexia suggest the diagnosis of a peripheral nerve disorder. Multiple cranial neuropathy associated with polyneuropathy, correspond to a Guillain-Barré syndrome

(GBS) variant called polyneuritis cranialis. This variant accounts for less than 5% of all cases of GBS (Gomez-Sanchez et al., 1999; Polo et al., 2002; Wang et al., 2011).

Myasthenia gravis should be considered in the differential diagnosis of patients with ptosis and diplopia. In this case, the patient presented initially with distal paresthesias and areflexia, favoring a diagnosis of Guillain-Barré syndrome variant (Yuki and Hartung, 2012). Additionally, a repetitive nerve stimulation test showed no significant decremental response.

Miller Fisher syndrome is characterized by the triad of ophthalmoplegia, areflexia, and ataxia. Antibodies to the GQ1b ganglioside are found in over 90% of cases (Levin, 2004). However, this diagnosis was excluded based on additional findings such as the presence of bulbar weakness and blood serum negative for anti-GQ1b antibodies. Furthermore, respiratory failure is not commonly found in this condition.

Two thirds of GBS cases are preceded by an acute infectious illness (Yuki and Hartung, 2012). As with other variants of GBS, this acute multiple cranial neuropathy form was associated with an upper respiratory tract infection, supporting the notion of molecular mimicry as an etiologic mechanism (Vucic et al., 2009).

As seen in many—but not all—patients with a Guillain-Barré syndrome variant, the nerve conduction study showed motor conduction block of >50% reduction in proximal to distal CMAP amplitude, abnormal temporal dispersion, diminished conduction velocity and absent F waves (Vucic et al., 2009).

Approximately half of patients with Guillain-Barré syndrome have albuminocytologic dissociation throughout the course of the first week of illness (Yuki and Hartung, 2012). For this particular patient, the CSF protein level was normal, presumably because the lumbar puncture was performed four days after the onset of symptoms.

When a diagnosis of Guillain-Barré syndrome variant is suspected, other causes of acute multiple cranial neuropathy must be ruled out (e.g. tumors of the skull base, space-occupying lesion in the brain stem, sarcoidosis, diabetes mellitus, Tolosa-Hunt syndrome and Lyme disease) (Lyu and Chen, 2004). For some authors splitting Guillain-Barré syndrome into several variants may not be that useful and instead these should be considered a spectrum of the same disease with common immune mechanisms. (Ter Brugge et al., 1998; Lehmann and Hartung, 2009).

Intravenous immunoglobulin (IVIg) is the standard treatment for patients with GBS; unfortunately the preferred therapy for patients with the multiple cranial neuropathy variant of GBS has not been well defined. The limited existing evidence suggests that IVIg is an effective treatment (Wang et al., 2011).

Although multiple cranial neuropathy is an unusual presentation of Guillain-Barré syndrome, from a practical standpoint it is considered a regional variant that may simulate various other disorders (Unal-Cevik et al., 2009; Wang et al., 2011). Further multicenter studies are needed to better understand this syndrome and to establish consensus for diagnostic criteria.

Conflicts of interest

None.

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