Current Journal of Neurology

Clinical Note

Curr J Neurol 2020; 19(2): 93-5

An atypical form of chronic inflammatory demyelinating polyneuropathy presenting with initial isolated bulbar weakness

Received: 08 Dec. 2019
Accepted: 03 Feb. 2020

Malek Mansour, Asma Ouerdiene, Anis Riahi, Jamel Zaouali, Ridha Mrissa

Department of Neurology, Military Hospital of Instruction of Tunis, Tunis, Tunisia

Keywords
Polyradiculoneuropathy; Chronic Inflammatory Demyelinating Polyneuropathy; Glossopharyngeal Nerve; Vagus Nerve

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired inflammatory neuropathy, classically characterized by a slowly progressive or relapsing symmetrical sensorimotor dysfunction developing over at least 8 weeks. However, CIDP can have a heterogeneous presentation with atypical forms in about 54% of cases, leading to a delay in diagnosis and treatment.[1] Predominant cranial nerve involvement is a relatively unusual feature of CIDP described in only 5% of cases.[1] Oculomotor nerves (III, IV, and VI) are most often affected, followed by the VII and, more rarely, IX, X, and XI cranial nerves.[1] We report here a case of a particular severe form of CIDP presenting with initial predominant bulbar weakness including IX and X cranial nerves dysfunction.

A 56-year-old man, without any particular medical history, presented on October 13, 2018, 16 days following a flu, a rapidly progressive dysphagia with frequent choking associated to dysphonia and numbness of the posterior right half of the tongue, reaching a severe condition, in about a week, with total aphagia, laryngopharyngeal reflux, saliva stasis, aphonia, and dyspnea. He was explored with a laryngeal-nasofibroscopy showing unilateral paralysis of the right half of the pharynx; esophago-gastro-duodenoscopy, cavum and skull base tomodensitometry, and cerebral magnetic resonance imaging (MRI) were all normal. The evolution was then made 45 days after the beginning of the symptomatology, by the appearance of ascending paresthesia, associated with muscle weakness.

He was admitted to our department, on December 07, 2018, 55 days after the beginning of

How to cite this article: Mansour M, Ouerdiene A, Riahi A, Zaouali J, Mrissa R. An atypical form of chronic inflammatory demyelinating polyneuropathy presenting with initial isolated bulbar weakness. Curr J Neurol 2020; 19(2): 93-5.

Corresponding Author: Asma Ouerdiene
Email: ouerdiene.asma@gmail.com

the symptomatology. Physical examination showed flaccid tetraplegia of grade 4 based on the Medical on the Overall Disability Sum-Score (ODSS), Research Council (MRC) grading, and grade 7 based areflexia, and sensory ataxia with a positive Romberg sign, and loss of deep sensation in four limbs associated with dysfunction of IX and X cranial nerves and associating dysphonia with nasal speech, abolished emesis reflex, and uvula deviation to the right.

An electromyography (EMG) was performed, and showed sensory and motor demyelinating polyradiculoneuropathy with decreased motor conduction velocities, prolonged distal latencies, and prolonged F-wave latencies in the four limbs, with decreased sensory conduction velocities in four limbs, and decreased amplitude in the upper lumbs. No increment or decrement in repetitive nerve stimulation was found. Cerebrospinal fluid (CSF) analysis highlighted a discrete albumino-cytological dissociation with 0 white cell, and protein level of 0.43 mg/dl. Complete metabolic, infectious, autoimmune, and paraneoplastic workup was negative including glycated hemoglobin (HbA1c), glycemia, liver tests, renal tests, vitamin B12, free T4 (FT4), thyroid stimulating hormone (TSH), hepatitis B virus (HBV), hepatitis C virus (HCV), human immunodeficiency virus (HIV), Epstein-Barr virus (EBV), cytomegalovirus (CMV), herpes simplex virus (HSV), West Nile virus (WNV), Lyme, brucellosis, syphilis serologies, antinuclear antibodies (ANA), antineutrophil cytoplasmic antibodies (ANCA), anti-ganglioside antibodies, acetylcholine receptors antibodies, tumor markers, onconeuronal antibodies, and throracic-abdominal-pelvic computed tomography (CT) scan.

The diagnosis of definite CIDP was made according to European Federation of Neurological Societies/Peripheral Nerve Society 2010 guidelines.

He received an immunoglobulin therapy, and was put on methylprednisolone; but his condition continued to deteriorate with worsening of the muscular weakness to MRC of grade 2 in the upper limbs, with an ODSS of grade 12. Plasmapheresis was indicated. The evolution was then made of a drastic improvement. He was discharged on January 26, 2019 on methylprednisolone 60 mg daily, associated with an immunosuppressive agent (azathioprine), and continued to improve with a full recovery (MRC of 5 and ODSS of 0).

CIDP usually presents as a progressive or relapsing symmetrical flaccid tetraparesis with areflexia and sensory dysfunction. Diagnosis is based on the clinical presentation, supported by CSF and electrophysiological analysis. Multiple atypical forms showing distinct features have been described in literature in 54% of cases[1] leading to a delay in diagnosis and treatment. This includes predominantly distal features in 17% of cases, exclusively sensory polyneuropathy in 15% of cases, asymmetric disease in 8% of cases, associated CNS demyelination in 8% of cases, restless legs syndrome in 1% of cases, and predominant cranial nerve involvement in 5% of cases.[1] III, IV, VI, and VII cranial nerves were the most affected. IX and X cranial nerves involvement is infrequent in patients with CIDP,[1] presented by our patient, too.

Lux et al.[2] described a case of a 72-year-old man with initial progressive sensorimotor limb disturbance, followed 18 months later by the onset of a dysarthria, dysphonia with nasal speech, dysphagia, and bifacial weakness associated to a muscle weakness. Despite improvement in limb strength and recovery of palatal and facial weakness with intravenous immunoglobulin (IVIG), hypoglossal neuropathy persisted. Teramoto et al.[3] described a case of a 69-year-old Japanese woman who presented initially with limbs weakness that disappeared spontaneously; and then after 5 months, quadriplegia with slight dysphagia developed, which disappeared after IVIG. She presented then a relapse comprising dysphagia alone which responded as well to IVIG, but not to corticosteroids.

There have been few descriptions of cases with cranial nerve involvement presenting as the initial symptom preceding the development of a more generalized polyneuropathy, and especially IX and X cranial nerves involvement, like our patient. Rotta et al.[1] described a case of a 43-year-old woman characterized by an initial onset made of dysgeusia, dysphagia, and bilateral facial numbness and weakness, preceding by 5 months typical symptoms with imbalance and tingling in her fingertips. She was treated with IVIG and corticosteroids, and she improved. Malka et al.[4] described a case of a patient presenting with initial progressive ataxia and bulbar weakness with dysarthria and dysphagia over a 6-month period, followed by limb weakness and respiratory dysfunction which responded well to plasmapheresis like our case. Donaghy et al.[5] described cases of CIDP presenting with initial ocular palsy preceding the onset of a chronic relapsing polyneuropathy, with relapses including dysphagia and facial weakness. Complete recovery was not obtained despite

immunosuppressant treatment.

There have been few descriptions of cases with cranial nerve involvement presenting as the initial symptom preceding the development of a more generalized polyneuropathy in CIDP, and especially IX and X cranial nerves involvement, which was rarely described, like in our case. The aim of this case presentation is to highlight some atypical forms of CIDP, especially with the glossopharyngeal and vagus nerve involvement presenting as an initial isolated symptom.

CIDP can have a wide spectrum of phenotypes leading to a delay of diagnosis and treatment. It should be considered in patients with predominant cranial nerve involvement even with bulbar weakness presenting as an initial isolated symptom.

Conflict of Interests

The authors declare no conflict of interest in this study.

Acknowledgments

None.

References

1. Rotta FT, Sussman AT, Bradley WG, Ram AD, Sharma KR, Shebert RT. The spectrum of chronic inflammatory demyelinating polyneuropathy. J Neurol Sci 2000; 173(2): 129-39.
2. Lux D, Zarkali A, Harikrishnan S. Cranial neuropathy in chronic inflammatory demyelinating polyradiculoneuropathy: A consideration with variable response to treatment. J Neurol ScI 2017; 381: 781.
3. Teramoto H, Morita A, Hara M, Ninomiya S, Shigihara S, Kusunoki S, et al. Relapse with dysphagia in a case of chronic inflammatory demyelinating polyradiculoneuropathy. Intern Med 2015; 54(14): 1791-3.
4. Malka R, Muntean E. Chronic inflammatory demyelinating polyneuropathy presenting with recurrent ataxia, bulbar weakness, and respiratory dysfunction. Neurology 2012; 78(1 Suppl): P06.135.
5. Donaghy M, Earl CJ. Ocular palsy preceding chronic relapsing polyneuropathy by several weeks. Ann Neurol 1985; 17(1): 49-50.