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Case Report

Chronic Inflammatory Demyelinating Polyneuropathy with Atypical Symptoms: A Rare Case of Tongue Fasciculation and Facial Muscle Weakness

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Abstract

This case report presents a 55-year-old man, with a history of hypertension, who developed an unusual and atypical presentation of chronic inflammatory demyelinating polyneuropathy (CIDP). Over 3 months, the patient experienced progressive limb weakness ascending from lower limbs to upper limbs with marked tongue fasciculations and facial muscle weakness without dysarthria or dysphagia. While cranial nerve involvement has been documented in CIDP, cases featuring the hypoglossal nerve are much less frequent than those with the facial nerve. This makes the occurrence of both happening at the same time quite rare. Diagnosing atypical presentations of CIDP can be tricky, and offer a challenge for neurologists in daily practice. A comprehensive diagnostic workup that could include blood work, cerebrospinal fluid analysis, and a nerve conduction study is required to confirm the diagnosis and rule out potential other causes. Effective management in this case included several inpatient plasmapheresis sessions and administration of steroids, which led to significant clinical improvement in the patient's symptoms. These breakthroughs are essential for clinicians to promptly diagnose and treat patients suffering from the same diseases.

Key words: Fasciculation, facial paralysis, middle-aged, polyradiculoneuropathy, chronic inflammatory demyelinating.

Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an uncommon autoimmune disorder characterized by an abnormal immune response that destroys the myelin sheath and damages peripheral nerve axons.[1] The incidence of CIDP varies from about 1.2 to 8.9 cases per 100,000 people globally, and it can start at any age but is more common around 50 years old, while about 10% of cases involve children.[2] CIDP is characterized by a pattern of relapse and remission, with symptoms lasting for more than two months. The clinical manifestations of this condition vary widely, with predominant presentation often involving sensorimotor disorders that symmetrically affect both the proximal and distal areas and are frequently accompanied by areflexia and conduction slowing. Although less common, CIDP may also present with involvement of the cranial nerves, autonomic symptoms, and neuropathic pain.[3] Cranial nerve involvement in CIDP is uncommon, but it can still occur in 5–20% of cases, resulting in various cranial neuropathies, including facial muscle weakness and tongue fasciculations associated with hypoglossal and facial nerve involvement. While most presentations feature classic symptoms such as muscle weakness and sensory disturbances, these atypical manifestations can complicate the diagnosis of CIDP.[4,5] Facial palsy is the most common cranial nerve involvement in patients with CIDP. In contrast, hypoglossal nerve involvement is exceptionally rare, highlighting its uncommon presentation.[6-8]

This case report highlights the rare occurrence of tongue fasciculation and facial muscle weakness in a patient diagnosed with CIDP. This

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Authors Contribution

SR & SZ conceptualized the project. SR also did the literature search, statistical analysis and data collection. Drafting, revision & writing of manuscript were done by SZ & M.

study aimed to outline the patient's unique clinical presentation, challenges encountered during the diagnostic process, and subsequent treatment.

Case Report

We present the case of a 55-year-old man from Afghanistan, with a history of hypertension. The patient presented to the outpatient department of Liaquat National Hospital, Karachi, three months after the symptom's onset, with a complaint of progressively worsening bilateral symmetrical weakness. This weakness initially appeared in the lower limbs and gradually extended to involve the bilateral upper limbs, ultimately causing the patient to be bedridden. Activities of daily living, such as getting up from bed, standing, walking, and even buttoning his shirt, became exceedingly challenging.

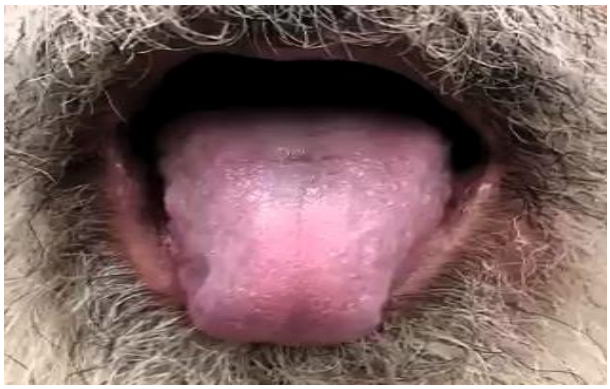

Figure 1a: Tongue atrophy and fasciculations.

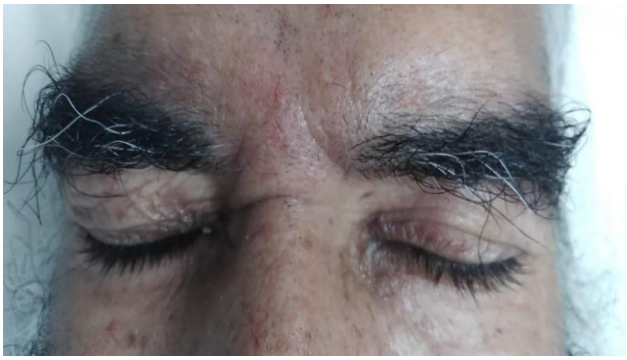

Figure1b: Bilateral facial weakness and inability to forcibly close the eyes.

Video Link: Tongue fasciculations-video mp4

On neurological examination, the patient remained well-oriented to time, place, and person, with no preceding history of infection and no association with bladder or bowel dysfunction. Notable clinical findings included marked fasciculations in the tongue (Figure-1a) and facial muscle weakness. (Figure-1b) Importantly, there was no evidence of dysarthria or dysphagia, and the fasciculations were localized to the tongue. There was bilateral lower motor neuron facial weakness, as seen in the figure below, he was unable to close his eyes completely. Generalized muscle wasting was observed in all four limbs, with a Medical Research Council grade of power 2/5 distally and proximally in the upper limbs and 0/5 in the lower limbs both distally and proximally. In addition, all deep tendon reflexes were absent. There was a glove-stocking sensory loss. The joint position and vibration were intact.

The patient's initial assessments included a comprehensive blood analysis, which included a full blood count, erythrocyte sedimentation rate, C-reactive protein, and UCE (urea, creatinine, electrolytes) all of which yielded unremarkable results. Furthermore, following informed consent, a lumbar puncture (LP) was performed. Cerebrospinal fluid (CSF) analysis from the LP revealed a sugar level of 97 and a protein concentration of 284, with no observed cellular abnormalities. Subsequently, an ANA panel by immunoblot and Anti HIV1/2 was performed, both of which returned negative results. In addition, the patient's serum B12 concentration was quantified at 1043 pg/mL, and serum protein electrophoresis was normal. Electromyography (EMG) and nerve conduction studies (NCS) showed diffuse sensory-motor demyelinating neuropathy with secondary axonal loss, supporting the diagnosis of CIDP (Table-1: a-c).[9] Based on the comprehensive assessment, a diagnosis of CIDP was -established, and secondary causes were excluded. A treatment plan was initiated, including a series of six plasmapheresis sessions on alternate days. He was started simultaneously on the oral steroid prednisolone at a dose of 1 mg/kg/day. In addition, limb physiotherapy was administered twice daily to support the patient's rehabilitation. His power improved from 0/5 to 3/5. We discharged the patient on slow-tapering doses of steroids with follow-up in his own country.

We obtained written informed consent from the patient for the publication of this case report, ensuring their privacy and voluntary agreement. No personal identification details were included.

Discussion

Our case report highlights an uncommon manifestation of chronic inflammatory demyelinating polyneuropathy (CIDP), which is characterized by limb weakness and cranial nerve involvement. The results from the nerve conduction velocity tests matched established electrodiagnostic criteria for

Table 1a: Sensory nerve conduction study.

Nerve	*Recording Site*	*Stimulation Site*	*Latency (ms)*	*Distance (cm)*	*Amplitude (µv)*	*NCV (ms)*
Rt. Median	F2	W			No response	-
Rt. Ulnar	F5	W				-
Lt. Median	F2	W	9.3	14.0	15.2	14.9
Lt. Ulnar	F5	W			No response	
Lt. Sural	A	C	4.8	14.0	10.0	29.1
Rt. Sural	A	C	4.6	14.0	15.2	38.2

Table 1b: Motor nerve conduction study.

Nerve-Muscles	*Stimulus site*	*Latency (ms)*	*Distance (cm)*	*Amplitude (mv)*	*NCV (ms)*	*F Wave LATENCY (ms)*
Rt Tibial (AH)	A					
Rt Peroneal (EDB)	A			No response		
Rt Peroneal (TA)	DK					
Rt Tibial (Gastroc)	Popliteal fossa	11.4	-	60uv	-	-
Rt. Median (APB)	W	7.6	7.0	2.7	-	66.6
`` `` `` `` ``	E	18.1	29.0	1.1	27.6	-
Rt. Ulnar (ADQ)	W	5.1	7.0	5.5	-	53.9
`` `` `` `` ``	DE	13.3	26.0	3.3	31.7	-
`` `` `` `` ``	PE	16.3	10.0	3.2	33.3	-
Lt. Peroneal (EDB)	A			No response		
Lt Peroneal (TA)	DK	9.8	10.0	250uv	-	-
`` `` `` `` ``	PK	14.6	10.0	50uv	20.8	-
Lt. Tibial (AH)	Popliteal fossa			No response		
Lt Tibial (Gastroc)	PK	10.5	-	280uv	-	-
Lt. Median (APB)	W	7.7	7.0	2.9	-	53.7
`` `` `` `` ``	E	18.1	29.0	1.9	25.0	-
Rt. Ulnar (ADQ)	W	6.3	7.0	3.1	-	55.5
`` `` `` `` ``	DE	18.6	26.0	1.0	18.8	-
`` `` `` `` ``	PE	25.1	10.0	440uv	15.3	-
Rt. Facial	Mastoid	6.5	-	640uv	-	-
Lt. Facial	Mastoid	5.7	-	910uv	-	-

Table 1c: Electromyography.

	Spontaneous Activity			*Motor Units*			
	Fibs.	*Psw*	*Others*	*Amplitude*	*Duration*	*Polyphasic*	*Recruitment*
Rt FDI	Nil	Nil	RFR	Few High	Few Broad	No	Reduced
Rt. TA	+	+	None	High	Broad	No	Reduced

Abbreviations: NCV-Nerve conduction velocity, EDB- Extensor Digitorum Brevis, gastroc-gastrocnemius, AH-abductor hallucis, TA- tibialis anterior, APB-abductor pollicis brevis, ADQ- adductor digiti quinti, FDI- First Dorsal Interosseous, psw-positive sharp waves, fibs-fibrillation potential

CIDP while showing a positive change following immunomodulatory therapy. Thorough clinical and laboratory evaluations effectively ruled out alternative underlying causes, consistent with the diagnostic criteria set forth by the European Academy of Neurology and Peripheral Nerve Society guidelines, categorizing the patient's condition as a definitive case of CIDP.[9] The typical clinical presentation of Chronic Inflammatory Demyelinating Polyneuropathy often includes a symmetric sensorimotor polyneuropathy. However, this case shows that CIDP can have many symptoms. Some atypical features were noted, like tongue fasciculation and weakness in facial muscles, highlighting the challenge of diagnosing cases with unusual presentations.

The most striking aspect of this case was the simultaneous involvement of the hypoglossal nerve along with bilateral facial nerve involvement. CIDP has been known to cause cranial nerve palsies, including those impacting cranial nerves II, III, IV, VI, and VII as well as cranial nerves IX, X, and XII.[5-8] Facial muscle weakness is reported in around 13% of cases.[4] In another study, 92 CIDP patients were examined. Among them, 15% had facial weakness, 6% had bulbar palsy, and 4% exhibited ophthalmoplegia.[10] However, involvement

of the hypoglossal nerve is much less common, with reports indicating a prevalence of only 0–3.8%.[4,5]

After an extensive literature review based on PubMed, we could identify only two cases at least to our knowledge that described the association of the presentation of tongue fasciculations with bilateral facial paresis.[8,11]

The presence of atypical clinical features, such as tongue fasciculation and facial muscle weakness, poses a challenge to the conventional symptomatology of Chronic Inflammatory Demyelinating Polyneuropathy (CIDP). Clinicians must recognize its broad symptom spectrum since it can present diversely with cranial neuropathy among others making it essential to consider CIDP during differential diagnoses. Incorporating nerve conduction studies into diagnostic evaluation is essential for identifying demyelination and sensory involvement, thereby facilitating precise and timely diagnosis.

This case report underscores the importance of considering CIDP as a potential diagnosis, even with uncommon clinical features such as tongue fasciculation and facial muscle weakness. This case serves as a reminder to clinicians to maintain a broad differential diagnosis of neuropathic presentations, especially when faced with a multifaceted disorder such as CIDP. The comprehensive diagnostic workup, characterized by neurophysiological assessments and the exclusion of alternative etiologies, demonstrates the complexity of diagnosing CIDP. Ultimately, prompt recognition and tailored treatment are essential for improving the patient's quality of life and prognosis.

Conflict of interest: None declared.

References

1. Kuwabara S, Misawa S. Chronic Inflammatory Demyelinating Polyneuropathy. In: Sango K, Yamauchi J, Ogata T, Susuki K, editors. Myelin [Internet]. Singapore: Springer Singapore; 2019: 333–43. (Advances in Experimental Medicine and Biology; vol. 1190). (Accessed on 10th December 2024) Available from URL:http://link.springer.com/10.1007/978-981-32-9636-7_21
2. Mathey EK, Park SB, Hughes RAC, Pollard JD, Armati PJ, Barnett MH, et al. Chronic inflammatory demyelinating polyradiculoneuropathy: from pathology to phenotype. J Neurol Neurosurg Psychiatry 2015; 86(9): 973-85.
3. Lewis RA. Chronic inflammatory demyelinating polyneuropathy. Curr Opin Neurol 2017; 30(5): 508.
4. Barohn RJ, Kissel JT, Warmolts JR, Mendell JR. Chronic inflammatory demyelinating polyradiculoneuropathy. Clinical characteristics, course, and recommendations for diagnostic criteria. Arch Neurol 1989; 46(8): 878-84.
5. Rotta FT, Sussman AT, Bradley WG, Ram Ayyar D, Sharma KR, Shebert RT. The spectrum of chronic inflammatory demyelinating polyneuropathy. J Neurol Sci 2000; 173(2): 129-39.
6. Shibuya K, Tsuneyama A, Misawa S, Suichi T, Suzuki Y, Kojima Y, et al. Cranial nerve involvement in typical and atypical chronic inflammatory demyelinating polyneuropathies. Eur J Neurol 2020; 27(12): 2658-61.
7. Hemmi S, Kutoku Y, Inoue K, Murakami T, Sunada Y. Tongue fasciculations in chronic inflammatory demyelinating polyradiculoneuropathy. Muscle Nerve 2008; 38(4): 1341-3.
8. Zhao H, Zheng Y, Meng L, Yu M, Zhang W, Lv H, et al. Chronic inflammatory demyelinating polyneuropathy with hypoglossal nerve involvement and inverted Beevor's sign: case report. BMC Neurol 2021; 21(1): 244.
9. Van den Bergh PYK, van Doorn PA, Hadden RDM, Avau B, Vankrunkelsven P, Allen JA, et al. European Academy of Neurology/Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint Task Force-Second revision. J Peripher Nerv Syst 2021; 26(3): 242-68.
10. McCombe PA, Pollard JD, McLeod JG. Chronic inflammatory demyelinating polyradiculoneuropathy. A clinical and electrophysiological study of 92 cases. Brain J Neurol 1987; 110 (Pt 6): 1617-30.
11. Roberto KT, Antonio AKD, Fernandez MLL, Damian LF. Chronic inflammatory demyelinating polyneuropathy with tongue fasciculation: A case report. J Clin Neurosci Off J Neurosurg Soc Australas 2020; 71: 297-9.