Case Report

A rare combination of acute inflammatory demyelinating polyradiculoneuropathy (AIDP) AMSAN variety in hypothyroidism – A case report

V. Shanthan[1], E.A. Ashok Kumar[2*]

[1]Post Graduate Student, [2]Professor of Medicine
Department of General Medicine, Mallareddy Institute of Medical Sciences, Hyderabad, India
[*]Corresponding author email: **ashokedla@gmail.com**

International Archives of Integrated Medicine, Vol. 7, Issue 3, March, 2020.

Available online at http://iaimjournal.com/
ISSN: 2394-0026 (P) **ISSN: 2394-0034 (O)**
Received on: 22-02-2020 **Accepted on:** 28-02-2020
Source of support: Nil **Conflict of interest:** None declared.
How to cite this article: V. Shanthan, E.A. Ashok Kumar. A rare combination of acute inflammatory demyelinating polyradiculoneuropathy (AIDP) AMSAN variety in hypothyroidism – A case report. IAIM, 2020; 7(3): 104-110.

Abstract

Acute inflammatory demyelinating polyradiculoneuropathy (AIDP) is an acute-onset, monophasic, immune-mediated polyneuropathy. Guillain barre syndrome and Miller Fisher Syndrome are variant forms of AIDP. The concurrence of AIDP with immune disorders of thyroid is known. In hypothyroidism with Hashimoto's thyroiditis and hyperthyroidism with Grave's disease, concurrence is known in AIDP, which is rare and infrequent. The diagnosis of AIDP relies heavily on the clinical impression obtained from the history and examination, although cerebrospinal fluid analysis and nerve conduction studies usually provide evidence supportive of the diagnosis. The calculated coincidental concurrence of AIDP (in both variants, MFS and GBS) and autoimmune thyroiditis was extremely low (0.0004%), thus suggesting a common pathogenic mediator. We report a case with a rare combination of AIDP, Acute Motor–Sensory Axonal Neuropathy (AMSAN) variety in a Multi Nodular Goitre patient with P.O. Subtotal Thyroidectomy and hypothyroidism without any associated autoimmune disorder.

Key words

Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP), Acute Motor–Sensory Axonal Neuropathy (AMSAN), Multi Nodular Goitre, Hypothyroidism, P.O. Subtotal Thyroidectomy.

Introduction

The association between Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP) and autoimmune to one or other auto-immune disorders of other systems is frequently reported. Importantly, the autoimmune thyroid disease has been shown to coexist with other autoimmune processes which may potentially cause neurological symptoms such as myasthenia, AIDP (Guillain-Barre syndrome) or pernicious anemia. Such conditions have to be considered as differential diagnosis in patients presenting with neurological signs and symptoms associated with thyroid disease [1-4].

GBS has five distinct subtypes: Acute inflammatory demyelinating polyradiculoneuropathy (AIDP), Acute motor axonal neuropathy (AMAN), Acute motor sensory axonal neuropathy (AMSAN), Miller Fisher syndrome, and Acute pan autonomic neuropathy [5]. The axonal forms are generally have poor prognosis, indicating a need for determining the specific subtype. These subtypes are distinguished by electro diagnostic features and pathological features.

But suffice to say, the association of Guillain-Barre syndrome which is Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP), AMSAN variety with hypothyroidism without auto immune disorder is a rare clinical situation discovered incidentally.

Case report

A 46 year old male patient was admitted to Mallareddy Institute of Medical Sciences, Hyderabad, with chief complaint of weakness of all four limbs. On admission, patient was conscious, well oriented with weakness of all the four limbs. On Examination, Pupils bilaterally equal reacting to light; Pulse Rate was 80 beats per minute, regular, normal volume. Blood pressure was 110/70 mm of Hg. Temperature was 98.4 $^{\circ}$F. Respiratory rate was 16 cycles per minute. SpO_2 – 97 % at room air, Height: 1.65 m, Weight: 58 kg. Examination of Cardiovascular, Gastrointestinal system and Respiratory system was normal. Nervous system examination – Intellectual function were normal, All cranial nerves function was normal. Hypotonia was present in all the four limbs. Power was 2/5 in all the limbs. Deep Tendon Reflexes (DTR) were all absent. Sensory loss was noted i.e. Loss of proprioception and hyperesthesia of both lower limb soles. Both Plantars were not elicitable. Bladder and bowel were not affected. Bilateral pedal edema was present pitting type.

Investigations:

Complete Blood Picture: Hb – 14.2 mg/dl, Total WBC Count – 11000 cells/mm^3, DLC: N: 72 %, E – 1%, B- 1%, L – 35%, M – 1%, platelets count – 3 lakhs/mm^3.

Blood urea: 46 mg/dl, Serum creatinine: 0.6 mg/dl.

Na – 142 meq/dl, K – 3.8 meq/dl, Cl – 109 meq/dl

T3 -118 ng/dl, T4 – 7.4 mcg/dl, TSH – 16.99 micro IU/ml, Anti -TPO antibodies – 1.88 IU/mL, Anti – Thyroglobulin: 0.92 U/mL, Anti - ds DNA antibody: 10.4 IU/mL

CSF studies: CSF Cell Count less than 50 per uL

Ultrasound neck: impression: thyromegaly – changes of thyroiditis

FNAC - impression features suggestive of nodular goitre.

Treponema pallidum hemagglutination: 10 (negative)

P-ANCA – 2.67U/ml (negative)

C-ANCA – 1.77 U/ml (negative)

NCS reporting

Motor: Prolonged latency noted in left median, right median and right ulnar nerve. Decreased amplitude and conduction velocity noted in sampled nerves i.e. in bilateral median, ulnar, bilateral common peroneal nerve and tibial nerve, Absent F waves.

Sensory: Decreased SNAP noted in Bilateral median, ulnar and sural nerves.

Impression: features suggestive of symmetrical motor and sensory axonal polyradiculoneuropathy.

On admission, patient was diagnosed with quadriparesis (LMN type), peripheral neuritis, AIDP - AMSAN variety with multinodular goitre, p.o. partial thyroidectomy with hypothyroidism.

Treatment

Patient was treated with Inj. Solumedrol 1gram I.V. once a day for five days along with Tab. Eltroxin 100 mcg once a day followed by Tab. Wysolone 60mg per day tapering accordingly. A remarkable improvement was observed in the patient's motor activity as the days of treatment progressed.

Discussion

Guillain-Barré syndrome (GBS) is an eponym for a heterogeneous group of immune-mediated peripheral neuropathies. A feature common in all GBS variants is a rapidly evolving polyradiculoneuropathy preceded by a triggering event, most often a viral infection [3, 7]. GBS generally manifests as a symmetric motor paralysis with or without sensory and autonomic disturbances. The patient with GBS typically presents with weakness accompanied by tingling dysesthesias in the extremities. This weakness is prominent in the proximal muscles; legs are more often affected than arms. Paresthesias occur, spreading proximally but seldom extending beyond the wrists and ankles. Deep tendon reflexes disappear within the first few days of symptom onset [7].

The diagnosis of Guillain-Barré syndrome is based on typical clinical features, electrodiagnostic examination, and examination of the cerebrospinal fluid [7, 8]. Electrophysiology plays a determinant role in Guillain–Barré syndrome (GBS) diagnosis, classification of the subtypes and in establishing prognosis. In the last three decades, different electrodiagnostic criteria sets have been proposed for acute inflammatory demyelinating neuropathy (AIDP), acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN) [8].

Diagnostic Criteria for Typical Guillain-Barré Syndrome [7]
Features required for diagnosis
Progressive weakness in both arms and legs Areflexia
Features strongly supporting diagnosis
Progression of symptoms over days, up to four weeks Relative symmetry of symptoms Mild sensory symptoms or signs Cranial nerve involvement, especially bilateral weakness of facial muscles Recovery beginning two to four weeks after progression ceases Autonomic dysfunction Absence of fever at onset High concentration of protein in cerebrospinal fluid, with fewer than 10 cells per cubic millimeter Typical electrodiagnostic features.
Features excluding diagnosis
Diagnosis of botulism, myasthenia, poliomyelitis, or toxic neuropathy Abnormal porphyrin metabolism Recent diphtheria Purely sensory syndrome, without weakness

Subtypes of Guillain-Barré Syndrome [7, 9-14]
Acute inflammatory demyelinating polyradiculoneuropathy (AIDP)
Autoimmune disorder, antibody mediated is triggered by antecedent viral or bacterial infection Electrophysiologic findings demonstrate demyelination. Inflammatory demyelination may be accompanied by axonal nerve loss. Re-myelination occurs after the immune reaction stops
Acute motor sensory axonal neuropathy (AMSAN)
Wallerian-like degeneration of myelinated motor and sensory fibers. Minimal inflammation and demyelination Similar to AMAN except AMSAN affects sensory nerves and roots Typically affects adults
Miller Fisher syndrome
Rare disorder rapidly evolving ataxia, areflexia, mild limb weakness, and ophthalmoplegia Sensory loss unusual, but proprioception may be impaired. Demyelination and inflammation of cranial nerve III and VI, spinal ganglia, and peripheral nerves Reduced or absent sensory nerve action potentials, tibial H reflex is usually absent. Resolution occurs in one to three months.
Acute panautonomic neuropathy
Rarest of all the variants Sympathetic, parasympathetic nervous systems are involved. Cardiovascular involvement is common (postural hypotension, tachycardia, hypertension, dysrhythmias). Blurry vision, dry eyes, and anhidrosis Recovery is gradual and often incomplete. Often combined with sensory features

Electro diagnostic criteria for AIDP and AMAN [8, 20-23]

Criteria for AIDP	**Albers, et al. (1985) [19]**	**Cornblath (1990) [20]**	**Ho, et al. (1995) [21]**	**Hadden, et al. (1998) [23]**
	Must have one of the following in two nerves	Must have one of the following in two nerves	Must have one of the following in two nerves	Must have one of the following in two nerves
Conduction velocity	<95% LLN < 85% if d-amp <50% LLN	<80% LLN < 70% if d-amp <80% LLN	<90% LLN < 85% if d-amp <50% LLN	<90% LLN < 85% if d-amp <50% LLN
Distal latency	>110% ULN >120% if d-amp <LLN	>125% ULN >150% if d-amp <LLN	>110% ULN >120% if d-amp <LLN	>110% ULN >120% if d-amp <LLN
Temporal dispersion	Unequivocal or >20% prox-dist NP area or PP amp decrease;>15% prox-dist	>20% prox-dist NP area or PP amp decrease;>15% prox-dist	Unequivocal	Not considered
Conduction block	<0.7 prox.dist amp ratio	>20% prox-dist NP area or PP amp decrease;<15% prox -dist	Not considered	<0.5 prox-dist amp ratio and d-amp >20% LLN

F-wave latency	>120% ULN	>120% ULN >150% if d-amp <80%LLN	>120% ULN	>120% ULN

d-amp = distal CMAP amplitude; amp = CMAP amplitude; dur = CMAP duration; LLN = lower limit of normal; ULN = upper limit of normal; prox = proximal; dist = distal; NP = negative peak; PP = peak-to-peak; demyel = demyelination

The role of imaging

Diagnostic imaging, specifically MRI, does not play a key role in the diagnosis of GBS as GBS is mainly diagnosed via clinical features and supportive electrophysiological and CSF studies; however, MRI can be used as a supplemental diagnostic modality when other supportive studies are ambiguous [24].

The role of steroids

The value of corticosteroid treatment in acute polyneuritis is still uncertain, due to natural outcome of the untreated disease is recovery in most of the cases [25]. Corticosteroids given alone do not significantly hasten recovery from GBS or affect the long term outcome and very low quality evidence suggests that, oral corticosteroids delay recovery [26]. However we found a remarkable improvement in this patient with steroid treatment, and the patient was able to walk on his own on 7th day onwards and regained a complete motor power in all the limbs, and the sensory symptoms and signs disappeared on the 15th day of treatment.

Conclusion

It is evident that there is a rare concurrence between AIDP and one or other auto-immune disorders. The association of Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP), with immune disorders of thyroid is known.

AIDP with hypothyroidism of Hashimoto's thyroiditis [1] is reported. AIDP IN hyperthyroidism with Grave's disease [6], which is rare and infrequent [3] is also reported. We reported a rare combination of acute inflammatory demyelinating polyradiculoneuropathy (AIDP) in hypothyroidism, which is non-immunological disorder, Acute Motor–Sensory Axonal Neuropathy (AMSAN) variety in a multi nodular goitre patient P.O. Subtotal Thyroidectomy with hypothyroidism which is non-immunological disorder. The patient completely recovered from his motor and sensory deficit with steroid treatment.

References

1. Polizzi A, Ruggieri M, Vecchio I, Genovese S, Rampello L, Raffaele R. Autoimmune thyroiditis and acquired demyelinating polyradiculoneuropathy. Clin Neurol Neurosurg., 2001; 103(3): 151-4.
2. Adel Olshansky. Diagnosis and Treatment of Guillain-Barre Syndrome. American Medical Association Journal of Ethics, 2007; 9(8): 552-554.
3. M. Toudou Daouda, N.S. Obenda, L. Maazou, D. Camara, Z. Souirti, A. Elmidaoui, M. F. Belahsen. Guillain–Barre syndrome and Hashimoto's thyroiditis. QJM: An International Journal of Medicine, 2016; 547–548.
4. Yuanyuan Huang, Zhajian Ying, Zhibo Chen, WeiWei Quan, Yiyun Weng, Xu Zhang. Thyroid hormone level is associated with the frequency and severity of Guillain–Barré syndrome. International Journal of Neuroscience, 2017; 127(10): 893 - 899.
5. Gorson KC, Ropper AH. Gullain-Barre Syndrome (Acute Inflammatory Polyneuropathy) and Related Disorders. In: Katirji, B, Kaminski, HJ, Preston, DC, Ruff, RL, and Shapiro, BE (eds): Neuromuscular Disorders in Clinical

Practice, Butterworth Heinmann, Boston, 2002, p. 544-566.

6. Anirban Majumder, Sagar Basu. Guillain–Barré Syndrome developing in a Patient with Graves' Disease. JAFES, 2019; 34(1).
7. Dana L. Newswanger, LCDR, MC, USNR, National Naval Medical Center, Bethesda, Maryland Charles R. Warren, LCDR, MC, USNR, Naval Hospital Jacksonville, Jacksonville, Florida, Guillain-Barré Syndrome. American family physician, 2004; 69(10): 2405-2410.
8. A. Uncini, S. Kuwabara. Electrodiagnostic criteria for Guillain Barre syndrome: A critical revision and the need for an update. Clinical Neurophysiology, 2012; 123: 1487–1495.
9. Hahn AF. Guillain-Barré syndrome. Lancet, 1998; 352: 635-4.
10. McKhann GM, Cornblath DR, Griffin JW, Ho TW, Li CY, Jiang Z, et al. Acute motor axonal neuropathy: a frequent cause of acute flaccid paralysis in China. Ann Neurol., 1993; 33: 333-42.
11. Ho TW, Li CY, Cornblath DR, Gao CY, Asbury AK, Griffin JW, et al. Patterns of recovery in the Guillain-Barré syndromes. Neurology, 1997; 48: 695-700.
12. Griffin JW, Li CY, Ho TW, Tian M, Gao CY, Xue P, et al. Pathology of the motor-sensory axonal Guillain-Barré syndrome. Ann Neurol., 1996; 39: 17-28.
13. Mori M, Kuwabara S, Fukutake T, Yuki N, Hattori T. Clinical features and prognosis of Miller Fisher syndrome. Neurology, 2001; 56: 1104-6.
14. Zochodne DW. Autonomic involvement in Guillain-Barré syndrome: a review. Muscle Nerve, 1994; 17: 1145-55.
15. T. W. Ho, B. Mishu, C. Y. Li, et al. Guillain-barré syndrome in Northern China relationship to Campylobacter jejuni infection and anti-glycolipid antibodies. Brain, 1995; 118(3): 597–605.
16. R. A. C. Hughes, D. R. Cornblath. Guillain-Barré syndrome. The Lancet, 2005; 366(9497): 1653–1666.
17. S. S. Gunatilake, R. Gamlath, H. Wimalaratna. An unusual case of recurrent Guillain-Barré syndrome with normal cerebrospinal fluid protein levels: a case report. BMC Neurology, 2016; 16(1).
18. Eric Lombardi, Ryan Misek, Krishna Patel. An Unusual Presentation of Acute Weakness: Acute Inflammatory Demyelinating Polyneuropathy in a Patient with Psychiatric Illness. Case Rep Emerg Med., 2018; 2018: 4065342.
19. Albers JW, Donofrio PD, McGonagle TK. Sequential electrodiagnostic abnormalities in acute inflammatory demyelinating polyneuropathy. Muscle Nerve, 1985; 8: 528–39.
20. Cornblath DR. Electrophysiology in Guillain–Barré syndrome. Ann Neurol., 1990; 27(suppl): S517–20.
21. Ho TW, Mishu B, Li CY, Gao CY, Cornblath DR, Griffin JW, et al. Guillain–Barré syndrome in northern China: relationship to Campylobacter jejuni infection and anti-glycolipid antibodies. Brain, 1995; 118: 597–605.
22. Ho TW, Willison HJ, Nachamkin I, Li CY, Veitch J, Ung H, et al. Anti-GD1a antibody is associated with axonal but not demyelinating forms of Guillain–Barré syndrome. Ann Neurol., 1999; 45: 168–73.
23. Hadden RD, Cornblath DR, Hughes RAC, Zielasek J, Hartung HP, Toyka KV, et al. Electrophysiological classification of Guillain-Barré syndrome: clinical associations and outcome. Ann Neurol., 1998; 44: 780–8.
24. O. Alkan, T. Yildirim, N. Tokmak, M. Tan. Spinal MRI Findings of Guillain-Barré Syndrome. Journal of Radiology Case Reports, 2009; 3(3): 25-28.

25. Darryl C, De Vivo, W. King Engle. Remarkable recovery of a steroid – responsive recurrent polyneuropathy. J. Neurol. Neurossurg. Psychiat., 1970; 33: 62-69.

26. W.B. Matthews, D. AHowell, R.C. Hughes. Relapsing corticosteroid dependent -only neuritis. J. Neurol. Neurossurg. Psychiat., 1970; 33: 330-337.