

A Caes Report of Atypical Acute Inflammatory Demyelinating Polyneuropathy



Medical Science

KEYWORDS :

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ABSTRACT

Acute demyelinating inflammatory polyneuropathy (AIDP) is a general classification of pathologies that could affect secondary the peripheral nervous system. It is characterized by an autoimmune process directed towards myelin. Clinically it is characterized by symmetrical ascending progressive weakness and / or mild sensory changes. Acute inflammatory demyelinating polyneuropathy is often referred to as Guillain-Barré syndrome (GBS). GBS is the major cause of acute nontraumatic paralysis in healthy people and it is caused by autoimmune response to viral agents (influenza, coxsackie, Epstein-Barr virus, or cytomegalovirus) or bacterial infective organisms (Campylobacter jejuni, Mycoplasma pneumoniae). A detailed history, with symptoms of progressive ascending symmetrical bilateral weakness, areflexia, with a typical demyelinating EMG pattern supports the diagnosis. Progressive affection of respiratory muscles and autonomic instability coupled with a protracted and unpredictable recovery normally results in the need for ICU management. We present a case report of a patient with a atypical AIDP with asymmetrical limb involvement and asymmetrical facial involvement which was confirmed by CSF examination and EMG NCV report.

INTRODUCTION

Acute inflammatory neuropathies encompass groups of heterogeneous disorders characterized by pathogenic immune-mediated hematogenous leukocyte infiltration of peripheral nerves, nerve roots or both, with resultant demyelination or axonal degeneration or both, and the pathogenesis of these disorders remains elusive.

The recent isolation and characterization of human endoneurial endothelial cells that form the blood-nerve barrier provides an opportunity to elucidate leukocyte-endothelial cell interactions critical to the pathogenesis of inflammatory neuropathies at the interface between the systemic circulation and peripheral nerve endoneurium.

Acute inflammatory demyelinating polyneuropathy is believed to be caused by an immunologic attack that is directed against myelin components. This results in a demyelinating polyneuropathy. Both cellular and humoral immune mechanisms appear to play a role. Early inflammatory lesions consist of a lymphocytic infiltrate that is adjacent to segmental demyelination. Macrophages are more prominent several days later.

The peripheral nerve changes consist of varying degrees of perivascular oedema, accumulations of mononuclear cells, and paranodal and less commonly, segmental demyelination. They are often multifocal with some predilection for the nerve roots, sites of entrapment, and distal ends. In the axonal variant of Guillain-Barre syndrome, axonal degeneration often predominates. Severe Guillain-Barre syndrome is often associated with axonal degeneration as well, which results in wallerian degeneration. Axonal degeneration occurs either as a primarily axonal process or as a bystander-type axonal degeneration, associated with demyelination. Rarely, the pathologic process extends into the central nervous system.

As the regeneration occurs, nerve sprouting and increased scarring often results.

With electron microscopy, macrophages are observed stripping off the myelin sheath. Humoral molecules such as antimyelin antibodies and complement likely contribute to the process by directing macrophages to Schwann cells by opsonization. Indeed,

complement and antibodies have been found to coat the myelin sheath. The changes are observed in nerve roots, peripheral nerves, and cranial nerves. In acute motor axonal neuropathy (AMAN, an AIDP variant), deposited complement is found at the nodes of Ranvier, while myelin often is left undamaged.

Damage to the myelin sheath leads to segmental demyelination. This results in decreased nerve conduction velocity and, at times, conduction block. In this current review, AIDP refers to the more common demyelinating form unless otherwise specified.

Typical case AIDP: Febrile antecedent especially campylobacter jejuni, CMV, EBV, mycoplasma, coxsackie, hepatitis virus. Other non-infectious causes include Hodgkins disease, and events such as surgery, childbirth, and immunization. Within 2 weeks, get symmetric leg paraesthesia, tingling, crawling sensations, pain especially in back (50%), ascending weakness. 50 % develop weakness diaphragm and cranial nerve. > 50 % have autonomic findings. CSF dissociation acellularity, increased protein.

Variants: asymmetric; pure motor; prominent sensory loss; preserved DTR's; regional presentations (pharyngeal/brachial/cervical); paraparetic; facial diplegia with paraesthesia; pure sensory neuropathy; pure autonomic neuropathy; Miller Fisher variant; axon loss variants (AMAN, AMSAN). Miller Fisher syndrome-- triad ophthalmoplegia, areflexia, ataxia (follows sensory loss). NCV show loss of SNAP amplitude. Course is often benign, may not require IVIG/Plasmapheresis. It May include facial weakness, dysarthria, dysphagia, abnormal pupils, limb weakness. 95 % have ANTI GQ1B antibodies. AMAN-- Chinese disease--NCV show motor amplitude loss without demyelination. High correlation with campylobacter jejuni. Antibodies to GD1a and GM1 at node of Ranvier. Involves complement activation and macrophage infiltration

AMSAN-- cannot differentiate from AMAN until enough time has elapsed to show NCV findings. Typically this does not respond to IVIG/plasmapheresis.

CASE PRESENTATION.

45 year old married female from middle socioeconomic class residing at Ahmedabad came to emergency ward with complain

of suddenly developed bilateral lower limb weakness, left upper limb weakness and bilateral LMN type of facial weakness which was more on left side then right side since 6 hours. She also had complain of slurring of speech associated with facial weakness. She was unable to stand and walk. She had no complain of tingling numbness, no bowel or bladder involvement, no fever, no trauma, no convulsion, no neck rigidity, no unconsciousness, no neck pain, no back pain, no electric shock like sensation on back.

Patient had no past history of diabetes mellitus, hypertension, ischemic heart disease, tuberculosis, jaundice, anaemia, recurrent fracture of any limb, or any same complain In past. Patient had no significant family history. Patient had adequate sleep and appetite, no complain regarding micturition and defecation, and no addiction. Patient was in post-menopausal stage. Obstetric history was not significant.

Patient was conscious, cooperative and oriented to time, place and person. Body temperature was normal, pulse rate was 92/min, regular, normal in volume, blood pressure was 150/92mmHg which was raised, respiratory rate was 14/min, no cyanosis, clubbing, no lymphadenopathy.

On neurological examination patient was conscious, cooperative and oriented to time, place and person, speech was slurred with preserved comprehension. Memory and intelligence were normal. Tone was decreased in left upper limb and bilateral lower limb. Power of muscle as follows.

Muscle	Bicep	Tricep	Knee	Ankle	Hip
Right	5	5	1	1	1
Left	3	3	1	1	1

All deep reflexes were absent, bilateral planter were present and were extensor. Bilateral facial muscle weakness which was LMN type and more on left side which suggestive of 7th asymmetrical cranial nerve involvement, and slurring of speech which might be due to brain insult lead to cranial nerve involvement or local pathology lead to cranial nerve involvement.

INVESTIGATION

Hb- 11.2 gm/dl, WBC- 11300/cu mm, platelet- 4,22,000/cu mm, urea- 28.1 mg/dl, S.creatinine- 0.51 mg/dl, SGPT- 56.1 U/L, SGOT - 68.9 U/L, ALP- 54.3 U/L, S.total bilirubin- 1.93 mg/dl, PT INR - 1.6, S. sodium- 138 meq/L, S. potassium- 3.8 meq/L.

ESR- 8(after 1 hour), CPK total- 123.8, CRP- 1.20, HIV- nonreactive, HBsAg- nonreactive. HbA1c- 10.5, urine R/M- sugar- appx 1000 mg/dl. CSF examination s/o protein-259.9, glucose- 68.0, chloride- 112, total cells-02, lymphocytes- 100%, polymorphs- 0%, which suggestive of albumin/cytological dissociation.

Urine porphobilinogen- negative
Serum protein electrophoresis- absent M band, raised alpha 2 globulin fraction suggestive of inflammation.

CSF protein electrophoresis- oligoclonal band not seen. Normal pattern with single major spike of albumin.
ANA - negative

EMG NCV suggestive of axonal and demyelination type of sen-

sory motor polyneuropathy.
NCCT brain- normal

MRI brain with whole spine screening suggestive of mild posterior disc bulge at L4-L5 and L5-S1 intervertebral disc, brain study was normal.

Patient was not affordable so we have not done anti GQ1 and anti GM1 antibody.

2D ECHO was normal with LVEF- 55%, RVSP- 29 mmHg.

Ultrasonography of abdomen was suggestive of liver enlarged(17.5 cm), fatty changes seen.

DIAGNOSIS

Newly detected hypertension and diabetes mellitus presented with atypical AIDP.

TREATMENT

As patient was diagnosed as atypical AIDP, she was treated with methylprednisolone 1 gm OD for three days and then 5 cycle of plasmapheresis on alternate day. Patient was found to be hypertensive and diabetic so we started antihypertensive drug and injection insulin as per requirement of the patient.

DISCUSSION

Patient was presented with sudden onset bilateral limb weakness with left sided upper limb and facial palsy,so we were thinking of initially cerebrovascular accident but point against that whose presence of bilateral facial weakness and it was LMN type. Though we had done NCCT brain to rule out any cerebrovascular accident which was normal. Another differential diagnosis was that might be compression of cervical cord with isolated cranial nerve involvement so patient further investigated with MRI brain with whole spine screening, result of which again ruled out any possibility of brain or spine pathology as cause of quadriplegia.we have performed protein electrophoresis, which shows absent M BAND which ruled out multiple myeloma. we have performed CSF routine microbiology. It showed albumin-cytological dissociation. we had also performed CSF electrophoresis to rule out multiple sclerosis and it suggestive of absent OLIGOCLONAL BAND. So we had further investigated for EMG NCV which had confirmed the diagnosis of AIDP. Patient's blood pressure was continuously at higher level and HbA1c was high and no past history of hypertension and diabetes mellitus so we diagnosed as newly detected hypertension and diabetes mellitus.

CONCLUSION

Normally patient of AIDP present with ascending bilateral symmetrical flaccid paralysis with or without cranial nerve involvement with areflexia and absent planter. Whenever cranial nerve nerve involved, 50% cases present with 7th cranial nerve involvement. But here patient had atypical presentation of AIDP. He presented with bilateral asymmetrical limb involvement with bilateral planter extensor and 7th,9th and 10th cranial nerve involvement which was improved by corticosteroid and 5 cycle of plasmapheresis. So we can conclude that sometimes patient's of AIDP may present with atypical presentation.

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