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ACUTE ONSET CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY: A CASE SERIES OF 4 PATIENTS FROM TERTIARY CARE CENTER

Neurology

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ABSTRACT

Acute onset CIDP(A-CIDP) is an often unrecognized CIDP variant which needs differentiation from Guillain -Barre syndrome(GBS) due to its different prognostic and treatment outcomes. METHODS: We report clinical course and investigations including electrophysiology of 4 patients of A-CIDP and comparison with Electrophysiology of 20 GBS patients. RESULTS: Four patients with initial GBS like clinical presentation were later diagnosed as A-CIDP. Three patients improved with immunotherapy but had exacerbation of weakness more than 8 weeks later while one patient had subacute onset progressive weakness. These patients were further managed with steroids and maintenance immunotherapy. CONCLUSION: GBS is diagnosed on the basis of clinical and electrophysiological criteria and managed with IVIg. Although some clinical features and initial electrorophysiology can suggest the possibility of A- CIDP, the diagnosis is confirmed on follow up only.

KEYWORDS

A-CIDP,GBS.

INTRODUCTION-

About 2% of initially diagnosed AIDP patients can develop more protracted course similar to CIDP especially in children and young adults, hence the term acute onset CIDP (A-CIDP). It constitutes 16% of CIDP cases.[1,2]

The two-inflammatory demyelinating polyradiculoneuropathies-AIDP and CIDP differ in various aspects. First difference lies in temporal evolution. In GBS the neurologic deficits evolve usually over less than 4 weeks, plateaus and then subsides slowly while for CIDP clinical criteria requires minimum progression of weakness of 8 weeks or more.[3] Up to 5% AIDP patients may have a recurrence but don't require maintenance therapy. Steroids are of therapeutic benefit in CIDP while they are ineffective in AIDP.[4]

A-CIDP patients have acute presentation of less than 4 weeks duration but may experience deterioration beyond 8 weeks, have 3 or more relapses more than 8 weeks after improvement and require maintenance immunotherapy.[3] A-CIDP patients have slower initial presentation and prominent sensory features but are less likely to have severe motor weakness, cranial nerve dysfunction or respiratory involvement requiring artificial ventilation than patients of GBS. [1,3] Conventional nerve conduction study (NCS) parameters are not useful to differentiate these patient groups in initial stages due to early nodal involvement in both conditions [5].Early institution of immune therapy including IV Ig and plasma exchange modify the disease course of both AIDP and CIDP and the diagnostic dilemma is resolved only on follow up. A-CIDP requires maintenance immunotherapy to prevent relapses and progression, thus emphasizing importance of early diagnosis.[3,6]

We report clinical and laboratory findings of 4 patients of A-CIDP with GBS like initial clinical presentation. We also describe a comparison of electrodiagnosis and clinical course of 20 GBS patients with the A-CIDP patients

Case report 1

An 18-year-old female was admitted with 8-10 days history of rapidly progressive ascending sensorimotor quadriparesis without cranial nerve involvement making her bedridden. Motor power was MRC grade 3/5 in lower limbs and 4-/5 in upper limbs with generalized hypotonia and areflexia. NCS revealed sensorimotor predominantly demyelinating polyneuropathy with albuminocytological dissociation in CSF. She was managed with 5-day IV Ig at dose of 0.4gm/kg/d. She improved and could walk independently but was admitted two and half months later with worsening of weakness in lower limbs for 8 days. She required support for walking. Motor power was MRC grade 4/5 in lower limbs and 5/5 in upper limbs with generalized areflexia. NCS again revealed predominantly demyelinating neuropathy with secondary axonal changes. Sural nerve biopsy was suggestive of CIDP like changes. She was managed with 5-day IV Methylprednisolone pulse therapy and maintenance therapy of oral steroids. She improved over next 1-2 weeks and could walk without support.

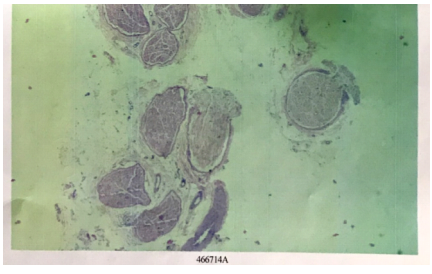

Fig1: Nerve biopsy of patient 1:H&E stained section; subperineurial and endoneurial edema with scattered lymphocytes and macrophages.

Table1: Motor conduction studies in case 1

	1-NCS			2-NCS		
	Right			Right		
Motor	Latency m-sec	Amp mV	Vel. m/sec	Latency m-sec	Amp mV	Vel. m/sec
Median	12	1.5	36	12.5	1.5	32.49
Ulnar	4	1.8	37.2	5	1.4	34.49
Peroneal	7	2.8	27	8.02	99.6µV	23.81
Tibial	14	1.6	22.8	20.42	119µV	13.74

F waves and SNAPs were not recordable(NR)

Case report 2

A 78-year-old male developed acute onset sensorimotor quadriparesis with bulbar and respiratory involvement. NCS revealed sensorimotor predominantly demyelinating polyneuropathy. He was treated as GBS with mechanical ventilation and plasma exchange.

Patient had improved and could walk independently but 10 months later again developed progressive sensorimotor LMN quadriparesis without cranial nerve involvement. Motor power was MRC grade 4/5 in all limbs with generalized hypotonia and areflexia with sensory loss (both exteroceptive and proprioceptive) in glove and stocking pattern. He had prominent sensory ataxia. Romberg's test was positive. He required support for walking. Repeat NCS revealed predominantly demyelinating polyneuropathy with secondary axonal changes. Sural nerve biopsy was suggestive of CIDP like changes. He was managed with 5-day IV Methylprednisolone pulse therapy and maintenance therapy of oral steroids .He started walking without support over next 2 weeks although with persistent paresthesias in hands.

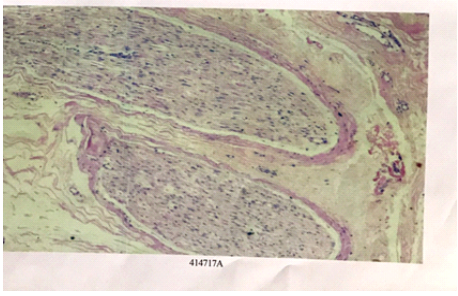

Fig2:Nerve biopsy of patient marked subperineurial and endoneurial edema with scattered lymphocytes and axonal degeneration

Table 2: Table1: Motor conduction studies in case 2

	I-NCS			II-NCS		
	Right			Right		
Motor	Latency m-sec	Amp mV	Vel m/sec	Latency m/sec	Amp mV	Vel. m/sec
Median	3.85	3.7	40	3.96	9.4	45.2
Ulnar	3.21	6.6	44.8	3.4	6.4	41.8
Peroneal	11.67	0.4	27.2	5.1	0.4	26.4
Tibial	5.5	1.4	34.11	4.9	4.9	36.80

F waves and SNAPs were not recordable

Case report 3

A 13-year-old male was admitted with 1-week history of rapidly progressive ascending type sensorimotor quadriparesis with bulbar involvement,making him bedridden. Motor power was MRC grade 3/5 in both lower limbs and 4-/5 in both upper limbs with generalized hypotonia and areflexia. NCS revealed sensorimotor predominantly demyelination with secondary axonal changes and CSF study was suggestive of albuminocytological dissociation. He was managed with 5-day IV Ig therapy at dose of 0.4gm/kg/d. Patient started improving and could walk with support, but 3 months later patient developed worsening of weakness. Motor power was MRC grade 4-/5 in lower limbs and 4/5 in upper limbs with generalized hypotonia and areflexia. He required support for walking. Repeat NCS revealed predominantly demyelinating polyneuropathy with secondary axonal changes. Sural nerve biopsy was suggestive of CIDP like changes. He was managed with 5-day IV Methylprednisolone pulse therapy and maintenance therapy of oral steroids . He started walking without support over next 2 weeks.

Table 3: Motor conduction studies in case 3

	I-NCS			II-NCS		
	Right			Right		
Motor	Latency m-sec	Amp mV	Vel m/sec	Latency m/sec	Amp mV	Vel. m/sec
Median	12.7	1.6	32.03	27.7	0.6	9.04
Ulnar	9.9	0.6	27.06	20.78	0.9	6.19
Peroneal	17.50	0.2	45.7	NR	NR	NR
Tibial	11.15	0.3	31.72	NR	NR	NR

F waves and SNAPs were not recordable(NR)

Case report 4

A 50-year-old male developed acute onset sensorimotor quadriparesis without cranial nerve or autonomic involvement. Motor power was MRC grade 4/5 in all limbs. His weakness progressed over next 2 weeks with motor power MRC grade 3/5 and generalized hypotonia and areflexia. He had become bedridden. He had sensory loss (both exteroceptive and proprioceptive) in glove and stocking pattern with prominent sensory ataxia and positive Romberg's test. Patient presented late as he didn't consult any physician and had become bedridden within 2 weeks. NCS revealed predominantly demyelinating polyneuropathy with secondary axonal changes. Sural nerve biopsy was suggestive of CIDP like changes. He was managed with 5-day IV Methylprednisolone pulse therapy and maintenance therapy of oral steroids.He improved over next 2-3 weeks and could walk with support.

Table 4: Motor conduction studies in case 1

	1-NCS			II-NCS		
	Right			Right		
Motor	Latency m-sec	Amp mV	Vel. m/sec	Latency m-sec	Amp Mv	Vel. m/sec
Median	19.9	2.6	29.27	NR	NR	NR
Ulnar	6.25	1.9	35.02	NR	NR	NR
Peroneal	10.42	0.4	37.96	NR	NR	NR
Tibial	16.04	0.6	32.87	NR	NR	NR

F waves and SNAPs were not recordable(NR)

DISCUSSION-

We report four patients of A-CIDP with clinical presentation simulating GBS. Patient 1,2 and 3 were initially managed as GBS due to rapidly progressive weakness, attaining a nadir within 2-3 weeks. Unlike the clinical features of typical A-CIDP, patient 2 had respiratory failure requiring artificial ventilation and patient 3 had bulbar involvement but all the four patients had prominent sensory features. Patient 2 was managed with artificial ventilation and plasma exchange, patient 1 and 3 received IV Ig while patient 4 didn't receive any treatment for first episode of illness. Patient 1,2 and 3 improved in terms of regaining ambulation before experiencing exacerbation of weakness more than 8 weeks later while patient 4 had progressive worsening of weakness and ataxia after an acute onset of illness making him bedridden within 2 weeks of onset of illness. All the four patients fulfilled the criteria of CIDP -Electrodiagnostic, CSF and nerve biopsy and improved with steroid therapy.

The initial NCS of patients 1,2 and 3 had distal latency >150% of Upper limit of normal ,conduction velocity <70% of Lower limit of normal in motor conduction studies,absent senory nerve action potential(SNAPs) ,absent F waves and Conduction block in two or more motor nerves and these parameters exacerbated in subsequent NCS of relapse episode .NCS of patient 4 had similar parameters .NCS of these four patients was compared with NCS of 20 GBS patients admitted between June 2017 to December 2018.The criteria by Asbury et al. was used to diagnose these patients as GBS.NCS findings of these 20 GBS patients were : absent SNAPs in 3 patients of sensory ataxic variant ; absent H reflex in 2 patients ;motor axonal changes in 4 patients of AMAN variant; sensorimotor axonal changes in 2 patients of AMSAN variant and sensorimotor demyelinating changes in 9 patients of AIDP. None of the 16 patients managed as GBS with IV Ig or conservative management met the NCS criteria of CIDP. These 16 patients improved and didn't report any exacerbation in follow up.

In addition to Asbury et al criteria,other electrodiagnostic criteria were used to diagnose GBS(Albers and Kelly,1989) and CIDP(Ad Hoc Subcommittee of American Academy of NeurologyAIDS Task Force 1991). Sural sparing pattern and sensory ratio may also be used to differentiate axonal dying back neuropathy from acquired demyelinating polyneuropathies. Conventional NCS parameters don't differ significantly between A-CIDP and AIDP patients and nerve excitability test parameters may have potential utility in differentiating between A-CIDP and AIDP[5,8]

A closer scrutiny of NCS of four patients of A-CIDP reported by us reveals that patients fulfilling electrodiagnostic criteria of CIDP should be managed as CIDP with steroids even though their clinical

presentation may be acute of less than 4 weeks duration. All of four A-CIDP patients had markedly prolonged distal latency with severe reduction in conduction velocity even at first clinical presentation which exacerbated during relapse. This simple observation can help us save expensive therapy like IVIg in our resource scarce setup as well as emphasizes the need to be more vigilant in follow up of such patients of A-CIDP. Electrodiagnostic studies should be repeated in such patients of GBS with predominant demyelinating features to distinguish them from persisting or new demyelinating features in electrodiagnostic studies which would suggest a possibility of A-CIDP .Further prospective studies are needed to establish electrodiagnostic criteria differentiating GBS from A-CIDP.

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