J. Dhaka National Med. Coll. Hos. 2018; 24 (02): 50-53

Case Report

Chronic inflammatory demyelinating polyneuropathy maymimic with Motor neuron disease

Nurul Amin Khan[1], Shaheen Wadud[2], Torikul Islam[3], Liton Chandra Ghosh[4], Kanaj Kumar Barman[5]

[1]Associate Professor, Department of Neuromedicine, Dhaka National Medical College, [2]Assistant Professor, Department of Neuromedicine, Dhaka National Medical College, [3]Registrar, Department of Neuromedicine, Dhaka National Medical College, [4]Assistant Professor, Dept. of Nephrology, Dhaka National Medical College, [5]Associate Professor, Department of Neurology, BSMMU.

Abstract:

Chronic inflammatory demyelinating polyneuropathy (CIDP) is characterized by symmetrical weakness, involving both proximal and distal muscles with sensory impairment. Muscle wasting is rarely pronounced in CIDP. When a patient present with CIDP and muscle wasting may mimicking with motor neuron disease like spinomuscular atrophy (SMA), Progressive muscular atrophy (PMA). It is very important to distinguish between CIDP and motor neuron disease (MND) by clinical, laboratory and histological feature because of different effective therapeutic strategies. Our patient 35 years old male presented with tingling, numbness and weakness in all four limbs. On examination there was muscle weakness and wasting more in the distal than the proximal. Electrophysiological investigation showed mixed sensory, motor demyelinating and axonal polyradiculoneuropathy and on Electromyogram (EMG)- fibrillation was absent, CSF examination there was albuminocytological dissociation. We established this case as a CIDP though it was mimicking with MND and treatment was started with steroid and other disease modifying drugs.

Introduction:

Chronic inflammatory demyelinating polyneuropathy is a common, albeit underdiagnosed, and potentially treatable disease with an estimated prevalence of about 0.5 per 100,000 children[1] and 1 to 2 per 100,000 adults.[2,3] Classic features of CIDP are symmetrical weakness in proximal and distal muscles, that progressively increases for two months. The condition is associated with impaired sensation, absent or diminised tendon reflexes, an elevated CSF protein level, demyelinating nerve conduction studies and sign of demyelination in nerve biopsy.[4-6] The course can be relapsing or chronic and progressive, the former being much more common in young adult. In CIDP pathologic abnormality is inflammatory demyelination and for this reason muscle wasting is less pronounced. A group of CIDP patient may present with muscle wasting of the limbs and electrophysiology and nerve biopsy showed axonal loss.[7] This group of patient has clinical similarity with motor neuron disease like SMA or PMA.

Case Report:

A 35 years old, male,non diabetic, normotensive, son of nonconsanguinal parent presented to us with the complaints of tingling sensation,numbness in all four limbs for 5 months. Weakness and wasting of all four limbs for 4 months, unable to walk for 2months. According to patient's statement he was reasonably alright 5 months ago, then he developed tingling sensation, numbness in all four limbs which was insidious in onset and gradually progressive, involved ulnar aspect of the right hand first then he started to experience numbness, tingling sensation in the left hand as well. He also noticed the same problem in both feet within 2 months. Subsequently, he also noticed weakness and wasting in his both upper limbs, involving both proximal and distal group of muscles. Then he experienced weakness and wasting in the both lower limbs with same pattern of involvement. Along with this he also developed progressive gait impairment. He developed difficulties in using stairs, and he started using a cane. He commented that his hands feel like "sand paper". He was unable to do pushups and curls. There were no chewing or swallowing difficulties orspeech impairment. There were no respiratory difficulties or bowel, bladderinvolvement. His weakness had no diurnal variation. He denied any other constitutional symptoms. He denied any exposure to toxins, tickbite, arthalgia, or skin rash. Patient did not give any significant past history and drug history. But he was a smoker. On examination patient appeared well, he was not in apparent distress. He was attentive, pleasant and cooperative and was able to provide appropriate medical history. His pulse was 74 b/m, BP-110/70 mmHg, not anemic, nonecteric, there was no lymphadenopathy. During neurological examination, we found that his highermental function including speech was normal,

cranial nerves including fundus were normal. Motor examination revealed, reduced muscle bulk and tone in both upper and lower extremities. There were muscle wasting more in the distal than the proximal groups of muscle (Figure-1). Muscle power was reduced more in the proximal than the distal group of muscle. Muscle power in the upper limbs were grade 3, on the other hand in the lower limbs were grade 2. All deep tendon reflexes were absent. Planter reflexes were bilaterally flexor. All modalities of sensation deminishedexcept position and vibration sense. Romberg's sign negative and there were no cerebeller sign.

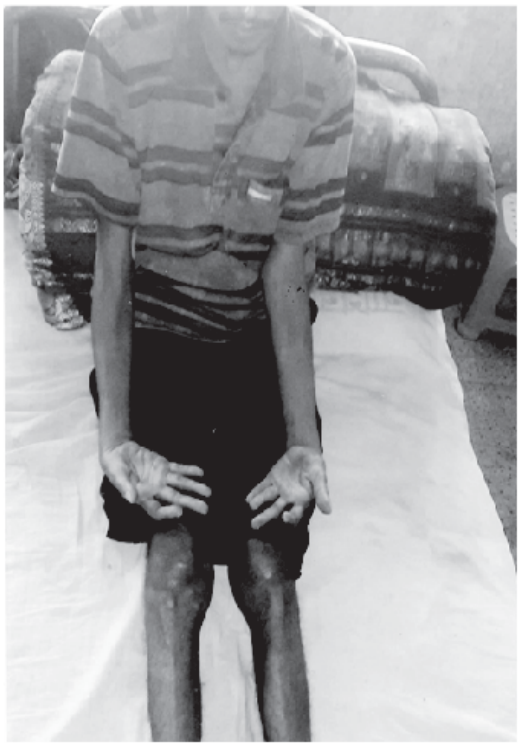

Figure 1: Wasting of all four limbs,where the distal muscle goups more wasted than the proximal group.

Investigation reports were as follows: CBC-WBC:10500/cmm, N:53%, L:39%, Hb%:16.2gm/dl, Platelet: 280X109/L. Urine RME-Normal, CSF study:Glucose: 4.2mmol/L, Protein: 130 mg/dl, Cell count:02/cmm, mostly lymphoc yte, ChestXray: Normal, Xray Lumbosacral spine both view: normal. CT scan of Brain: Normal. NCS: Mixed sensory motor demyelinating and axonal polyradiculoneuropathy. EMG: No feature of denervation that is positive sharp wave and fibrillation.Nerve biopsy was next plan but patient denied to do it. On the basis of above clinical findings as well as investigations our clinical diagnosis was CIDP and we exluded other mimicking diseases like SMA, PMA and we started steroid therapy and regular physiotherapy. Patient was improving gradually. Patient was discharged and kept under regular follow up.

Discussion:

Classic CIDP is characterized by a symmetric proximal and distal phenotype. When diagnostic criteria for CIDP were initially proposed, weakness of proximal and distal limbs was a mandatory inclusion criterion.[5,8,9] CIDP may begin insidiously and evolves slowly, attaining its maximum severity after several months or even a year or longer.[8] The symptoms and signs of CIDP may be asymmetrical initially and have ascending involvements. But it usually progresses slowly to symmetric weakness, loss of deep tendon reflex, and impaired sensation in hands and feet.[8,10,11] Antecedent infections can be identified far less regularly in patients with CIDP than those with acute inflammatory demyelinating polyneuropathy (AIDP).[12] Elevated concentration of CSF protein and evidence of demyelination on electrodiagnostic examination are found in most patients with CIDP.[8,10] CIDP ,though a demyelinating polyneuropathy is associated with concomitant axonal loss attributed to the primary demyelinating process.[7,13] This finding appear to be important, since this may create confusion with MND such as SMA and PMA. So, we have to exclude MND by clinical examination and other relevant investigations.

On the other hand SMA is an autosomal recessive disease characterized by degeneration of anterior horn cells of the spinal cord leading to progressive symmetrical weakness and atrophy of the proximal muscles.[14] Clinical classification of SMA is based on age at onset and maximum motor function acquired, with the following categories: 1) severe (type I, severe SMA or Werdnig- Hoffmann disease); 2) intermediate (type II or chronic SMA); 3) mild (type III, juvenile SMA or Kugelberg-Welander disease); and 4) type IV (adult SMA).[15] Type III SMA can be classified as type IIIa (Age of onset before three) and type IIIb (Age of onset after 3).[16] Patients with Type IIIa are able to walk until they are,[20] while Type IIIb patients will be able to walk for their whole lives.[17] In SMA, α motor neurons are lost progressively, only motor function is compromised and sensory neurons are unaffected. This loss of function leads to weakness and to progressive symmetrical atrophy of the proximal voluntary muscles of the legs,

arms and, sometimes, the trunk, as the disease progresses.[18] Proximal muscles are more involved than distal muscles, legs are more affected than arms and arms are more affected than the face anddiaphragm.[18] Our patient clinically mimicking SMA but electrophysiologycally did not show any evidence of denervation and features of MND.

Another CIDP mimicking motor neuron disease is PMA,which was first described by Aran in 1850,[19] is a disease that exclusively involves LMNs during its entire clinical course and comprises approximately 4% of all adult-onset motor neuron diseases.[20] A common presentation is that of focal asymmetrical muscle weakness in the distal extremities with gradualspread to other contiguous muscles without sensory impairment.The weakness and muscle atrophy is purely LMN in type and eventually involves both the upper and the lower extremities. A less common presentation is that of proximal rather than distal muscle weakness. PMA is a dignosis of exclusion. CIDP, SMA have to be ruled out first with the help of electrophysiological examination.[21] Our patient presented to us with weakness and wasting of all four limbs associated with sensory impairment. Weakness and wasting was started from the both upper limbs then gradually involved both lower limbs. Weakness was more profound in the proximal group of muscle but wasting was more in the distal group of muscle. Clinical presentation more or less similar to SMA or PMA except distribution pattern of muscle involvement and sensory involvement. When we performed electrophysiology, it showed mixed sensory motor demyelinating and axonal polyradiculoneuropathy and absence of denervation and fibrillation. CSF study showed there was albuminocytologicaldissociation. With the help of electrophysiology and CSF study, we went for definite diagnosis that this was a case of CIDP though clinical feature mimicking with the MND like SMA or PMA.

Clinical response with steroid and other disease modifying drugs are excellent in CIDP but later needs intravenous immunoglobulin. Our patient was respond bestly with steroid only. Most of the CIDP patient expired by either respiratory failure or other secondary infection.

Conclusion:

CIDP is a disease of the peripheral nerve presenting with wasting and weakness of limbs. It may occasionally create confusion with MND like SMA or PMA needs extensive clinical and electrophysiological evaluation for diagnosis. As because CIDP is a partially treatable disease and responds to intravenous immunoglobulin, plasma exchange and steroid therapy. Further research should provide further insight into the underlying mechanisms of nerve damage and may facilitate the development of more effective treatments.

References:

1 . Connolly AM. Chronic inflammatory demyelinating polyneuropathy in childhood. Pediatr Neurol 2001;24:177-82.

2 . McLeod JG, Pollard JD, Macaskill P, Mohamed A, Spring P, Khurana V. Prevalence of chronic inflammatory demyelinating polyneuropathy in New South Wales, Australia. Ann Neurol 1999;46:910-3.

3 . Lunn MP, Manji H, Choudhary PP, Hughes RA, Thomas PK. Chronic inflammatory demyelinating polyradiculoneuropathy: a prevalence study in south east England. J Neurol Neurosurg Psychiatry 1999;66:677-80.

4 . Dalakas MC, Engel WK. Chronic relapsing (dysimmune) polyneuropathy: pathogenesis and treatment. Ann Neurol 1981;9:Suppl:134-45.

5 . Barohn RJ, Kissel JT, Warmolts JR, Mendell JR. Chronic inflammatory demyelinating polyradi culoneuropathy: clinical characteristics, course, and recommendations for diagnostic criteria. Arch Neurol 1989;46:878-84.

6 . Research criteria for diagnosis of chronic inflammatory demyelinating polyneuropathy (CIDP): report from an ad hoc subcommittee of the American Academy of Neurology AIDS Task Force. Neurology 1991; 41:617-8.

7 . Bouchard C, Lacroix C, Plante V, et al.Clinicopa thologic findings and prognosis of chronic inf lammatory demyelinating polyneuropathy. Neuro logy 1999;52:498-503.

8 . Saperstein DS, Katz JS, Amato AA, et al. Clinical spectrum of chronic acquired demyelinating polyneuropathies. Muscle Nerve 2001;24:311-24.

9 . Dyck PJ, Lais AC, Ohta M, et al. Chronic inflamma tory polyradiculoneuropathy. Mayo Clin Proc 1975;50:621-37.

10. Sander HW, Latov N. Research criteria for defining patients with CIDP. Neurology 2003;60:S8-S15.

11. Koski CL. Therapy of CIDP and related immune-mediated neuropathies. Neurology 2002;59:S22-7.

12. Victor M, Ropper AH. Diseases of the peripheral nerves. In: Adams and Victor's Principles of Neurology 2001: 1410-3.

13. Dalakas MC. Advances in chronic inflammatory demyelinating polyneuropathy: disease variants and inflammatory response mediators and modifiers. CurrOpin Neurol 1999;12:403-9.

14. Dubowitz, V. The muscular dystrophies. In: Dubowitz, V. (ed.), Muscle Disorders in Childhood, 2nd ed. London: WB Saunders Co;1995.P.34-133.

15. Russman BS. Spinal muscular atrophy: clinical classifications and disease heterogeneity. J Chil Neurol. 2007;22:946-51.

16. Wirth B, Brichta L, Hahnen E. Spinal muscular atrophy: from gene to therapy. SeminPediatr Neurol. 2006;13:121-31. Review.

17. Zerres K, Rudnik-Schöneborn S. Natural history in proximal spinal muscular atrophy. Clinical analysis of 445 patients and suggestions for a modification of existing classifications. Arch Neurol. 1995;52:518-23.

18. Wang CH, Finkel RS, Bertini ES, Schroth M, Simonds A, Wong B, et al. Consensus statement for standard of care in spinal muscular atrophy. J Child Neurol. 2007;22:1027-49.

19. Visser J,deJongJM,de Visser M.The history of Progressive muscular atrophy:Syndrome or disease?Neurology2008;70:723-7.

20. MH, Ganesalingam J, et al. Natural history and clinical features of the flail arm and flail leg ALS variants. Neurology 2009;72:1087-94.

21. Krivickas LS, Carter GT. Motor neuron disease. In: DeLisa JA, Gans BM, Walsh NE, Bockenek WL, Frontera WR, Geiringer SR, editors. Physical medicine and rehabilitation: principles and practice. 4th ed. Philadelphia: Lippincott Williams &Wilkins; 2005: 931-52.