Case Report

Facial Nerve Palsy With Chronic Inflammatory Demyelinating Polyneuropathy

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ABSTRACT

Chronic inflammatory demyelinating polyneuropathy is an autoimmune disease characterized by progressive symmetrical motor and sensory dysfunction. There is a known association of chronic demyelinating polyneuropathy with diabetes mellitus. Cranial nerve palsies are also reported with CIDP. Here we are going to present a case of 40 years old diabetic who presented to us with progressive parapresis and left facial nerve palsy which subsequently turned out to be a case of CIDP on CSF examination and nerve conduction studies. We started oral steroids and Azathioprine to the patient and he showed dramatic response to steroids.

INTRODUCTION

Chronic inflammatory demyelinating polyneuropathy is an autoimmune disease which involves an autoimmune response against peripheral nerve myelin and is characterized by motor or sensory dysfunction in more than one limb for more than 2 months with hyporeflexia and characteristic CSF and electrodiagnostic test results[1]. Postulated mechanisms include antigen mimicry, triggering self-reactive T-cell clones, and cytokine upregulation that may induce aberrant MHC class II expression [2.] There is a known association of chronic inflammatory demyelinating polyneuropathy with diabetes mellitus[3, 4, 5, 6]. Cranial nerve palsies are also reported with CIDP like occulomotor nerve palsy [7, 8]. CIDP and GBS may be treated with intravenous immunoglobulin and plasmapheresis, and CIDP may also be treated with steroids. There is less evidence for steroid use but studies have failed to demonstrate a difference in efficacy among these three treatments; consequently, the choice is often based on availability, side effect profile and individual patient response[9]

CASE REPORT

A 40 year old man who is a known diabetic for the last 2 years taking oral hypoglycemic agents with poor compliance and control presented to us through out patient department with complaints of progressive weakness of legs and arms with difficulty in walking for three months and facial deviation for the last 20 days. The examination revealed symmetrical distal muscle weakness [Medical Research Council (MRC) grade 2-3] more in the lower limbs as compared to the upper limbs. All cranial nerves on examination were intact except left seventh cranial nerve which showed a lower motor neuron type facial palsy. There was distal dominant sensory deficit in both lower limbs, and reduced deep tendon reflexes. Albuminocytologic dissociation in CSF (total protein: 92 mg/dL; cell count: 3 lymphocytes/ mm3) was present. Nerve conduction study showed prolonged distal latency, slowed conduction velocity (10-20 m/s), delayed or absent F waves, and partial conduction block in the 4 limbs. These findings fully satisfy the diagnostic criteria for "definite CIDP" proposed by American Academy of Neurology[10]. The patient was started high dose oral corticosteroids i.e. Prednisolone 60 mg per day for the first one month and on weekly follow ups power was checked and recorded. Subjective assessment of the patient on subsequent visits showed dramatic improvement in the symptoms. The power in lower limbs gradually improved and the patient was able to walk independently after one month treatment. The improvement in upper limb followed and the patient was able to carry out his daily houseful activities after

about two months. The facial weakness of left side also showed improvement gradually. After high dose treatment with steroids for about 6 weeks the dose of steroids was tapered. At the same time immunosuppressant drug Azathioprine was added, initially at a dose of 50 mg which was gradually increased to 100mg per day. The tapering of steroids was continued fortnightly. Currently the patient is taking 10 mg of Prednisolone and 100 mg Azathioprine per day at which the patient's response was optimal. The patient is still on our follow up after about one year of continuous treatment.

DISCUSSION

Chronic inflammatory demyelinating polyneuropathy is a relatively rare disease which is due to an autoimmune response of our own lymphocytes against the myelin component of nerves. As it is an autoimmune disease it has an association with other autoimmune diseases as well. Association of CIDP with diabetes is well documented[4, 5]. Especially there is a strong association of CIDP with type one diabetes mellitus[3]. Our patient was also a diabetic. One important point to be emphasized here is that due to high dose steroids given in the treatment of CIDP the glycemic control of the patient may be disturbed so this should always be kept in mind treating a diabetic patient with corticosteroids. In our patients we closely monitored the blood sugar levels and his oral hypoglycemic dose had to be increased accordingly. Hattori N, Misu K, Koike H, et al. described that the age of onset does affect the disease course as well as the response in CIDP suggesting that if the patient is young at the onset of CIDP his disease will be of less severity as well as his chances of remission with treatment are more as compared to elderly population[11]. Our patient was 40 years old and he had no past history of such illness, still at this age the response of our patients to systemic steroids was quite satisfactory. There are three main treatment modalities for CIDP available i.e. steroids, plasmaphresis and I/V immunoglobulins. Though there is not enough evidence that systemic steroids are better than plasmaphresis and I/V immunogobulins [9], in our case the response of oral steroids has been very dramatic.

REFERENCES

1. Koller H, Kieseier BC, Jander S et al. Chronic inflammatory demyelinating polyneuropathy. N Engl J Med 2005; 352:1343–55.
2. Albert LJ, Inman RD. Molecular mimicry and autoimmunity. New England Journal of Medicine(Abbrev)?? 1999; 341: 2068–74.
3. Alves C, Braid Z, Públio N. Chronic inflammatory demyelinating polyneuropathy in an adolescent with type 1 diabetes mellitus. Arq Neuropsiquiatr 2009; 67:311-313.
4. Sharma KR, Cross J, Farronay O, Ayyar DR, Shebert RT, Bradley WG. Demyelinating neuropathy in diabetes mellitus. Arch Neurol 2002; 59:758-765.
5. Stewart JD, McKelvey R, Duncan L, Carpenter S, Karpati G. Chronic inflammatory demyelinating polyneuropathy (CIPD) in diabetics. J Neuro Sci 1996; 142:59-64.
6. Kalita J, Mistra UK, Yadav RK. A comparative study of inflammatory demyelinating polyradiculoneuropathy with and without diabetes mellitus. Eur J Neurol 2007; 14:638-643.
7. Tsuda E, Imai T, Hozuki T, Yamamoto D, Harada K et al. Transient Oculomotor Palsy Correlated with Nerve Enhancement on MRI in Chronic Inflammatory Demyelinating Polyneuropathy. The Japanese Society of Internal Medicine. Intern Med. 2009; 48:1985-7.
8. Alwan AA, Mejico LJ. Ophthalmoplegia, ptosis, and lid retraction caused by cranial nerve hypertrophy in chronic inflammatory demyelinating polyradiculoneuropathy. J Neuro-Ophthalmol 27: 99-103, 2007.
9. Toothaker TB, Brannagan TH. Chronic inflammatory demyelinating polyneuropathies: current treatment strategies. Current Neurology and Neurosciences. Rep 2007; 7: 63–70.
10. Cornblath DR, Asbury AK, Albers JW, et al. Report from an Ad Hoc Sucommittee of the American Academy of Neurology AIDS Task Force: research criteria for the diagnosis of chronic inflammatory demyelinating polineuropathy (CIPD). Neurology 1991; 41:617-618.
11. Hattori N, Misu K, Koike H, et al. Age of onset influences clinical features of chronic inflammatory demyelinating polyneuropathy. J Neurol Sci 2001; 184:57-63.

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