Chronic inflammatory demyelinating polyneuropathy with papillodema and response to immunoglobulins

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ABSTRACT

Chronic inflammatory demyelinating polyneuropathy is considered as a chronic form of Guillain-Barre syndrome differing from it in response to steroids, course and prognosis. Papillodema although rare is not an uncommon feature. Here we report a case of florid papillodema in a patient of chronic inflammatory demyelinating polyneuropathy, discuss the underlying mechanisms in relation to cerebrospinal fluid pressure and protein content and response to intravenous immunoglobulins.

Keywords: Polyneuropathy, papillodema, immunoglobulins.

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Chronic inflammatory demyelinating polyneuropathy (CIDP) is an idiopathic, acquired demyelinating neuropathy that shares many clinical, electrophysiological and pathological features with Guillain-Barre Syndrome (GBS) and differs chiefly by virtue of its rate of evolution, its prognosis and to some extent, by its response to treatment and hence can be considered as the chronic variety of GBS.[1,2] It begins insidiously and evolves slowly, but 15-20% of cases of CIDP have an acute presentation that mimics GBS. The clinical features are one of both sensory and motor abnormalities, though sensory abnormalities are often trivial. Among the uncommon features of CIDP, papilloedema has been reported in about 7% of cases[3], the same frequency seen in GBS. Although differences exists between CIDP and GBS in their responsiveness to treatment; CIDP being responsive to steroids whereas GBS does not, there are also many similarities. Both disorders appear to respond to plasma exchange (PE) and high dose intravenous immunoglobulins (IVIG). Many trails have shown that IVIG is effective in CIDP[4-6], though may require repeated infusions. Here we report a case of CIDP, who had bilateral gross papilloedema with haemorrhages along with other features and though had partial response to steroids, responded well to repeated IVIG infusions.

Case Report. A 48 year old male patient was admitted to the neurology unit with history of numbness, paresthesiae and weakness of lower limbs of 4 weeks, insidious onset, progressive and a few days before admission had weakness and paresthesiae of upper limbs and diplopia. He neither had a history of dyspnea, dysphagia or sphincteric disturbances nor preceding respiratory tract infection or diarrhoeal illness. Seven months prior to this admission he had a similar episode, was admitted to a local hospital, diagnosed as polyradiculopathy,

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Table 1 - Serial CSF study showing CSF pressure and proteins (days/weeks from admission).

Number of LP	CSF Pressure (mm of H_2O) Opening	Closing	Proteins (mg/dL)	Sugar (mg/dL)	Cells	Culture and Sensitivity
(3rd Day) Ist	425	325	860	71	Nil	Negative
(2 weeks) IInd	500	250	421	91	Nil	Negative
(4 weeks) IIIrd	400	350	450	74	Nil	Negative
(8 weeks) IVth	450	325	215	64	Nil	Negative
(13 weeks) Vth	425	250	206	60	Nil	Negative

treated with steroids, after which he made a good recovery, but only to relapse in a month's time, responded partially to steroids and was unable to walk unaided.

His neurological examination revealed normal higher mental functions and cranial nerves, fundii showed markedly congested angry looking disc with haemorrhages particularly around the disc with absence of venous pulsations. Motor system examination revealed wasting of small muscles of the hand, hypotonia in the lower limbs, muscle power of grade 3/5 in the upper limbs and in the lower limbs grade 3/5 proximally and grade 2/5 distally with dorsiflexors grade 0/5. His deep tendon reflexes were absent, plantars unresponsive, had bilateral footdrop and was unable to stand or walk. He was jittery and ataxic, his sensory was intact. Vital signs and systemic examination were unremarkable.

His blood counts, erythrocyte sedimentation rate, biochemistry, thyroid function tests were normal and rheumatoid factor, lupus erythromatosus cells, Hepititis B surface antigen, Hepititis C virus and human immunodeficiency virus were negative. In view of papillodema and small muscle wasting a magnetic resonance imaging scan of the brain, brainstem, and cervical spine was performed and was normal. Electromyogram and nerve conduction studiesusing concentric needle electrodes showed no distal responses and delayed latencies at the nerve trunks in all nerves of both lower limbs and prolonged distal latencies in the right median and ulnar nerves and prolonged F-wave latency in right ulnar nerve. Cerebrospinal fluid (CSF) analysis done on 5 different occasions during this admission (refer to Table 1) showed persistently high opening pressure and high protein content (860 mg/dL initially and 206 mg/dL in the last lumbar puncture), with normal sugars, no cells and culture negative.

He was managed initially with high dose IVIG 0.4 gms/kg/day for 5 days, responded appreciably, only to relapse in 2 weeks time, wherein he received 2nd and 3rd course of IVIG with 10 days interval and showed prolonged remission. Even though his muscle power improved and was able to stand and walk unaided, since his papillodema had not resolved and cerebrospinal fluid proteins were high, he received a 4th course of IVIG just prior to discharge. A month later, on follow-up, muscle power was near normal, but still was areflexic and CSF pressure remained high with an opening pressure of 425 mm of H_20) although the proteins content showed a marked fall to 206 mg/dL.

Discussion. Chronic inflammatory demyelinating polyneuropathy is a form of polyneuropathy characterised by chronic relapsing course, enlargement of nerves and responsiveness to steroids. Among the uncommon features of CIDP, papillodema is reported in about 7% of cases.[3] The precise pathogenesis of papillodema is still unclear and the proposed mechanisms include: (i) a high CSF protein concentration interfering with resorption of CSF by arachnoid villi[7], (ii) normal production rates of CSF with reduced outflow, a situation similar to patients with pseudotumor cerebri.[8,9] In our patient the most striking feature was florid papillodema with haemorrhages and although initially it correlated with high CSF protein and pressure, subsequently inspite of protein levels dropping down markedly, the CSF pressure remained high (refer to Table 1) suggesting that both mechanisms may be possibly operating together.

The therapeutic options in CIDP are prednisone, PE and IVIG. All three options are equally effective, however complexity and costs make PE the last choice. Prednisolone, although cheap and beneficial, the nature of the disease usually requires long term treatment at high doses with the inevitable adverse effect and hence IVIG is now widely used in the treatment. Despite occasional report[2] showing no significant advantage between IVIG and placebo group, several controlled trails[4-6] have shown to be beneficial in the treatment of CIDP, albeit for only several weeks to months, after which infusion have to be repeated. Our experience has been no different, in

that although he responded to steroids initially, the response was only partial in the 2nd attack. Hence repeated infusions of IVIG were given with a remarkable outcome.

In conclusion that: papillodema although described, the severity as seen in this case is unusual. The underlying mechanism may be multifactorial-possibly due to coexistence of high CSF pressure and high protein content as illustrated. The case also illustrates the usefulness of repeated IVIG infusions in this conditions.

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