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Gullain-Barre Syndrome as Ropper's Variant

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Abstract

Gullain-Barre syndrome commonly present as ascending flaccid paralysis with progressive respiratory involvement. There are number of variants with different presentations like Pharyngeal-cervical-brachial variant. Here we are describing 26 year old male who presented with bilateral ptosis with bilateral lateral recti and bilateral seventh nerve involvement with progressive brachial involvement. Nerve conduction study showed conduction block in right posterior tibial nerve, the right common peroneal CMAP and conduction velocity were normal with absent F waves. The left common peroneal and left posterior tibial nerve shows absent F wave. Intravenous Immunoglobulins were started and patient showed improvement in ptosis and slight improvement in facial and brachial involvement. **Keywords:** GBS, Ropper's variant, AIDP.

Introduction

Gullain-Barre syndrome is an acute onset, monophasic illness characterized by ascending paralysis, hyporeflexia or areflexia with mild sensory involvement. AIDP classically present as distal paraesthesia with weakness beginning in distal muscles and progressin ascending fashion. Diaphragmatic and cranial muscle weakness involve in significant number of patients. Atypical AIDP manifest as distinct variants. Ropper variant presenting as bilateral sixth and bilateral seventh nerve involvement is rarely described in literature. Here we are describing similar presentation that is diagnosed with NCS. Our purpose is to describe atypical presentation that mimics other condition like Myaesthenia.

Case Report

A 26 year old male, Farmer by occupation presented with history of double vision first noticed in early morning at his usual time of awakening. This double vision was increasing on looking either side and hampered his movement across home. He went to sleep again. This was associated with difficulty in opening his eyelids as noticed by family members. History of difficulty in eating was present due topooling of food in buccal cavity but there was no history of drooling of saliva. Next morning patient noticed weakness

JMSCR Vol||09||Issue||08||Page 93-95||August

in his bilateral upper limbs in form of difficulty in holding objects in hands and simultaneously not able to comb his hair but no difficulty in standing or walking. On examination positive findings were bilateral ptosis. Pupils were of equal size and normally reacting to light. Bilateral Sixth and Seventh nerve involvement (Fig.1a,b) was present. Single breath count was 8.The power in upper limbs was 3/5 at shoulder and 3/5 at elbow extension and 4/5 at elbow flexion and 3/5 at wrist bilaterally and areflexia at upper limbs and 5/5 power in lower limbs with normal reflexes and normal plantar reflexes bilaterally.



Fig.1a Left lateral rectus palsy



Fig 1b Right lateral rectus weakness

In acute presentation, with such LMN type weakness possibility of Myaesthenia gravis and snake bite was also kept beside GBS. Ice pack test and Neostigmine challenge test was done to exclude Neuromuscular junction pathology. No obvious history of any bite and no fang marks were found on examination. To rule out central cause CT head was also done.

Nerve Conduction study showed conduction block in right posterior tibialnerve, the right common peroneal CMAP and conduction velocity are normal with absent F waves. The left common peroneal and left posterior tibial nerve showed normal CMAPs and conduction velocity with absent F waves.

CSF analysis was done on fifth day of weakness and showed paucicellular smear with occasional lymphocytes with total protein 52 mg/dL.

Patient was started on intravenous immunoglobulins with dosage according to weight. There was improvement in ptosis and single breath count after 4 days of Ivig but no significant improvement in brachial weakness during hospital stay.

Discussion

GBS has varied clinical presentations depending upon variants. First description was given by J.B.O. Landry in 1859. A.H. Ropper in 1986 described unsusual variant as severe ptosis with minimal or no ophthalmoplegia and mild facial weakness.^[1] In 1994 he described patient with right sixth nerve palsy with minimal facial weakness. Power loss is mostly proximal with hyporeflexia^{.[2]} The regional involvement may be due to antigenic differences between peripheral nerves or their endothelium. Multiple cranial nerve involvemet is common in GBS .B. Amita et al showed Bulbar palsy as most common involvent (49.2%) in their study of GBS patients and Ophtalmoplegia was in 6.5%.^[3] Very often such presentation are initially misdiagnosed as having myasthenia gravis or brainstem stroke.

Myaesthenia gravis is an autoimmune disorder caused by autoantibodies against the nicotinic acetylcholine receptor on postsynaptic membrane at the neuromuscular junction. Typically patients present with weakness and fatigability of muscles on sustained or repeated activity that improves after rest. Ptosis that is often partial and may be unilateral, is a common presenting feature.^[4]

JMSCR Vol||09||Issue||08||Page 93-95||August

Venomous snakes inhabit the foothills and mountain ranges. Raina S et. al. observed that among the symptomatic venomous snake bites in Himachal Pradesh, common were neuroparallytic followed by heamatotoxic.^[5]

In our patient there were acute rapid progressing symptoms and signs consistent with Ropper's variant and unsual thing was involvement of third nerve and brachial weakness

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