

OCULOPHARYNGEAL VARIANT OF GUILLAIN-BARRÉ SYNDROME: A DIAGNOSTIC DILEMMA

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Introduction

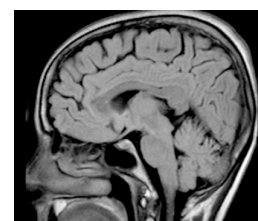
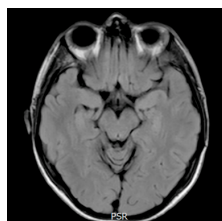
Multiple cranial neuropathy as a variant is rare and occurs in only 5% of patients. It can occur in autoimmune, neuromuscular, vascular, infectious and neoplastic disease. Hence the differential diagnosis could be difficult.

Case report

A 49-years-old woman was admitted for acute onset of dysarthria, rhinolalia and dysphagia without limb weakness. She denied prodromal symptoms and any past medical history.

These symptoms quickly worsened within twenty-four hours with bilateral facial palsy, diplopia and absence of gag reflex without strength or sensitive impairments.

In the emergency room she underwent cerebrospinal fluid analysis that excluded an albumin-cytologic dissociation but revealed oligoclonal bands. Brain magnetic resonance excluded vascular disease, electrophysiological studies revealed normal peripheral nerve conductions and repetitive nerve stimulation (RNS).



In consideration of the rapid development and clinical severity she was treated with plasmapheresis (six sessions) and oral corticosteroids, within 48 hours of illness onset.

During the recovery laboratory findings, including antibody testing (anti-GQ1b, anti-AchR and anti-MuSK) were found to be normal. On day 14 of the illness CSF analysis reconfirmed previous results and ENG study showed reduction of sensory conduction velocity at the ulnar and sural nerves with normal RNS.

Nerve	Latency (ms)	Amplitude (µV)	Velocity (m/s)	Velocity (m/s)	Nerve	Latency F min(ms)	Latency F max(ms)	Latency F med (ms)	Latency F med (ms)
R-Ulnar	2,50	6,80	<u>44,00</u>	57,90	S-Peroneal	44,10	47,15	45,63	43,58
S-Ulnar	2,10	12,30	52,40	54,10	S-Ulnar	29,85	31,55	30,52	27,33
R-Sural	3,20	15,10	<u>43,80</u>	54,20					
S-Sural	3,00	18,00	<u>43,30</u>	56,00					

Clinical course gradually improved after 10 days and she completely recovered within 21 days. ENG/EMG performed 1 month later proved a complete resolution. Follow up 6 months later showed no evidence of recurrence.

Discussion

In this case the severity and rapidity of clinical impairment, restricted only to cranial nerves without sensory and motor involvements complicated the diagnosis. In the critical phase plasmapheresis stabilized the patient in the event of diagnostic immune-mediated disease.

However, instrumental studies, such as MRI and ENG, in association to a monophasic clinical course and follow-up allowed us to confirm the diagnostic suspect of a possible oculopharyngeal variant of Guillain-Barré syndrome without limb involvement.

References

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