Myelin Oligodendrocyte Glycoprotein (MOG) Associated Neuromyelitis Optica (NMO) Presenting with Isolated Absent F Wave

Shahpar Nahrir1*, Naim I Kajtazi2 and Jameela A. Saeedi2
1Heart of England Foundation Trust, Birmingham B5 9SS, UK
2National Neuroscience Institute, King Fahad Medical City, Riyadh 59046, KSA

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*Corresponding author: Shahpar Nahrir, Heart of England Foundation Trust, Birmingham B5 9SS, UK, E-mail: s_nahrir@yahoo.com

Abstract
We describe an unusual presentation of Neuromyelitis Optica (NMO). A young boy presented with two days history of acute onset flaccid paraplegia. Electrophysiological study showed only absence of F wave. Initial exam was suggestive of Gullian-Barre Syndrome (GBS). Nevertheless, MRI spine demonstrated a contrast enhancing lesion in conus medullaris. Few days later, patient developed bilateral optic neuritis. NMO was diagnosed then. Antibody to Aquaporin 4 was not found in the serum, but to Myelin Oligodendrocyte Glycoprotein (MOG) was present. Patient was refractory to IV IgG therapy. He continued to develop symptomatic new lesions in brain and spine. Later Rituximab therapy initiation ceased the disease. However, absent “F wave” in Nerve Conduction is quite a perplexing association. It may reflect concurrent peripheral nerve involvement which needs to be explored. This case is unique due to its atypical presentation and also to the best of our knowledge this is the first reported case of MOG positive NMO from Saudi Arabia.

Keywords: NMO; F wave; Paraplegia; GBS; MOG

Abbreviations
NMO: Neuromyelitis Optica; GBS: Gullian-Barre Syndrome; MOG: Myelin Oligodendrocyte Glycoprotein.

Introduction
Neuromyelitis Optica is a rare but a devastating disease if not managed timely and appropriately [1]. Hence, its knowledge is pertinent. The clinical features of NMO are manifold. Sero-negative group comprises about 5-40 % of all NMO [2]. Anti MOG antibody associated NMO is the commonest type of the sero-negative group. Its identification relies on accurate knowledge of its characteristic features and use of appropriate assay technique. This case illustrates a unique clinical feature of NMO which lends a doubt over its widely recognized sole involvement of central nervous system. Our patient not only had clinical features similar to GBS but also showed absence of F wave suggesting selective proximal block along with extensive central nervous system disease burden. One can argue that NMO can expand its involvement to peripheral nervous system with the presence of anti-MOG antibody. This case therefore allows us to ponder about such variation. Moreover, this is the first reported case of anti-MOG positive NMO from Saudi Arabia. Report of its existence will alert other physicians of this region to explore this entity.

Case Presentation
A 13-year-old previously healthy boy presented with two days history of bilateral leg weakness and numbness. There was no associated disturbance in speech, swallow, vision, bowel or bladder. He had no preceding history of fever, headache, diarrhea, or any heavy carbohydrate intake or raw milk ingestion. On neurological examination, he was awake, alert, and oriented. Lower limb examination showed hypotonia, muscle power 2/5, absent deep tendon reflexes, and unresponsive plantars. He had decreased light touch, pinprick, and temperature sensation over both legs but his joint position and vibration sense was intact. Lumbar puncture had normal opening pressure with 32 cells (lymphocyt-97%), normal protein and glucose, negative study for viral PCRs (EBV, HSV, CMV), MTB complex, cytology, and Brucella. Serum ANA/ENA, HIV (1&2), and Quantiferon TB Gold test were negative. Nerve Conduction Study (NCS) revealed intact CMAP but absent “F” wave (Figure 1). The overall clinical impression was of Gullian-Barre

Figure 1: Nerve Conduction Study, at presentation showing absent “F wave”
Syndrome (GBS). Hence, IVlg 0.4g/kg/day for three days was given. He showed no response. Meanwhile, MRI-spine demonstrated localized expansion of conus-medullaris characteristics of myelitis. In addition, CSF oligoclonal band turned out to be positive along with high IgG index. Patient was then offered IV Methylprednisolone 1g/day for five days. He demonstrated substantial improvement. He started to walk using a walker. Two weeks later, he developed sudden visual loss in both eye to the extent of finger counting only. MRI-brain revealed multiple non-contrast enhancing supra and infratentorial parenchymal lesions. MRI-Spine showed interval progression of conus medullaris lesion to T11 and a new lesion around T3-T4. CT chest, abdomen and pelvis ruled out any occult malignancy or lymphadenopathy. There was prolonged P wave latency bilaterally in visual evoked testing. NMO/AQP4-IgG antibodies from serum and CSF were negative. Instead, antibody to Myelin Oligodendrocyte Glycoprotein (Anti-MOG) was detected in serum. The diagnosis was then reconsidered to be Neuromyelitis Optica (NMO). Plasma exchanges were commenced. Four days later he deteriorated, became completely paralyzed with only light perception. He developed new onset intractable vomiting. Repeat MRI-brain demonstrated enhancing lesion in medulla-oblongata. Rituximab (375mg/m²-weekly for 4 weeks) and maintenance of oral steroid were prescribed. The patient showed dramatic response after 4 doses of Rituximab with oral steroid with no further relapse (Figure 2). Currently, 18 months into his illness, he has regained normal muscle power and visual acuity. He is independent in ADL. He resumed regular schooling and takes part in sports with no difficulty.

Discussion

We describe a unique manifestation of anti-MOG positive NMO that is presenting as isolated absent ‘F’ wave in NCS. Historically, NMO is regarded as a variant of MS. Over the centuries, it has emerged as a distinct disease. Presence of AQP4 antibody confers definite NMO. However, sero-negative NMO has also transpired. Many case series has enumerated the features of sero-negative NMO. Some have been reported to posses Anti-MOG antibody. Comparative study by J Kitley et al. [1] showed sero-negative group having higher predilection for male, younger age, Devic’s phenotype (simultaneous/rapidly sequential optic neuritis, myelitis) and sphincter dysfunction. It is also known that sero-negative have lower median EDSS at the time of best recovery. MOG/NMO is more likely to have conus lesion with lower probability of relapse. Our patient bore many features common to these descriptions. The patient had incoercible vomiting reflecting the lesion in medullary area-prostema. This area is notoriously involved in AQP4/NMO, as AQP4 is heavily expressed here. Moreover, area-prostema lacks blood brain barrier allowing easy access to the antibody [3]. But its involvement in sero-negative NMO has not been highlighted before.

MOG is expressed on the outer most lamellae of myelin sheath [4]. Therefore, are believed to be vulnerable to antibody attack. Anti-MOG is not specific to NMO. Its association with other autoimmune conditions (MG, SLE, ADEM, and pediatric MS) is known for years. Recent studies illustrated that autoimmune response to MOG can induce a NMO-like disease in experimental animal models [5].

But concerns remain for the findings in NCS. “F wave” can be absent in demyelination, axonal degeneration, physiological conduction failure, or impaired excitability. Kuwabara S et al. [6] showed 10-27 % of GBS patients having only isolated absent F wave in first examination which they termed “selective proximal block”. Katchanov J et al. [7] described a patient with combined ADEM and demyelinating polyradiculoneuropathy. Furthermore, they perceived ADEM as monophasic illness believed to result from infection. Thus a theory of molecular mimicry as seen in GBS might take place in ADEM. Kitley J et al. [8] described a patient with Devic’s like phenotype to have actually an overlap of ADEM. Similarly, our patient may be ADEM-like thereby somewhat explaining “absent F wave” as well.

Conclusion

We could not elucidate the exact pathophysiology of “absent F wave” in our patient. But certainly, its presence in NMO is distinct that needs further research.
References


*Corresponding author: Shahpar Nahrir, Heart of England Foundation Trust, Birmingham B5 9SS, UK; E-mail: s.nahrir@yahoo.com

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