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A case of Miller Fisher syndrome with multiple cranial nerves enhancement on MRI

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Miller Fisher syndrome is a clinical variant of Guillain-Barŕe syndrome with classic triad of ophthalmoplegia, ataxia and areflexia which there were some reports of multiple cranial nerves enhancement on MRI. We report a case of 66-year-old male presenting with headache and numbness of hands and feet, then gradual developed bilateral diplopia, ptosis, decreased deep tendon reflex and impaired tandem gait. Post contrast MRI showed multiple cranial nerves enhancement. Miller Fisher syndrome was diagnosed based on clinical triad and electrophysiologic evidence of polyneuropathy involved axon and myelin. We suggested that thin slice section of the post contrast MRI is likely to be helpful for detection of these abnormalities.

Keywords : *Miller Fisher syndrome, multiple cranial nerves enhancement, Guillain-Barŕe syndrome variant*

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Miller Fisher syndrome เป็นกลุ่มอาการอย่างหนึ่งของ Guillain-Barŕe syndrome ซึ่งประกอบด้วย ophthalmoplegia, ataxia และ areflexia ซึ่งมีรายงานผู้ป่วยหลายฉบับที่พบว่ามี enhancement ของเส้นประสาทสมองจากการตรวจด้วย MRI รายงานผู้ป่วยชายอายุ 66 ปี มาด้วยอาการปวดศีรษะและชาที่มือและเท้าทั้งสองข้าง จากนั้นเริ่มมีอาการหนังตาตก เห็นภาพซ้อน เดินเซ และมี reflex ลดลง การตรวจ MRI หลังฉีดสารเพิ่มความแตกต่างของภาพพบว่ามี enhancement ของเส้นประสาทสมองหลายเส้น ผู้ป่วยได้รับการวินิจฉัยว่าเป็น Miller Fisher syndrome จากอาการแสดงและผลการตรวจการนำไฟฟ้าของระบบประสาทที่พบว่ามีพยาธิสภาพของเส้นประสาทส่วนปลายที่ axon และ myelin การใช้ thin slice section ในการตรวจ MRI หลังจากฉีดสารเพิ่มความแตกต่างของภาพน่าจะมีประโยชน์ในการตรวจหาความผิดปกติดังกล่าว

คำสำคัญ : *Miller Fisher syndrome, การ enhancement ของเส้นประสาทสมอง, Guillain-Barŕe syndrome variant.*

Miller Fisher syndrome (MFS) is a cranial nerve variant of Guillain-Barré syndrome (GBS). Its classic triad of ophthalmoplegia, ataxia and areflexia were first described by Fisher in 1956. It is often clinically recognizable, but some parts of the syndrome overlap with GBS and it is more limited form in involving only the components of the triad; these have also been widely described. Its pathogenesis is autoimmune neuropathy caused by infection-induced aberrant immune response that damages the peripheral nerves. Gadolinium enhancement of the cranial nerves on MRI has been reported in some literatures.

Case Report

A 66-year-old male presented with headache and numbness of hands and feet for two weeks. The symptoms included progressive headache and visual disturbance for a week. On examinations, there were normal general exam with decreased sensation of both hands. Blood chemistry and cell counts were normal. No abnormality was detected on the contrast enhanced CT scan of the brain on the first day of admission.

On the third day of admission, however, bilateral diplopia and gradual ptosis developed with decreased deep tendon reflex in both lower extremities and impaired tandem gait. There were also decreased sensations along the courses of bilateral ophthalmic and maxillary nerves. Cerebrospinal fluid examination revealed normal cell count with a protein concentration of 119 mg/dL. Electrodiagnostic studies showed evidence of sensorimotor polyneuropathy that involved axon and myelin. On MRI, thin slice section of the skull base (3-mm slice thickness and 0.5-mm intersection gap) were performed to look for any lesion in the cavernous sinus and abnormality of the cranial nerves. Enhancement of the cisternal portion of bilateral oculomotor nerves (Figure 1), bilateral facial nerves and bilateral Meckel's caves (Figure 2) were detected. Enhancement of the facial nerves was seen from the cisternal portion down to the mastoid segment.

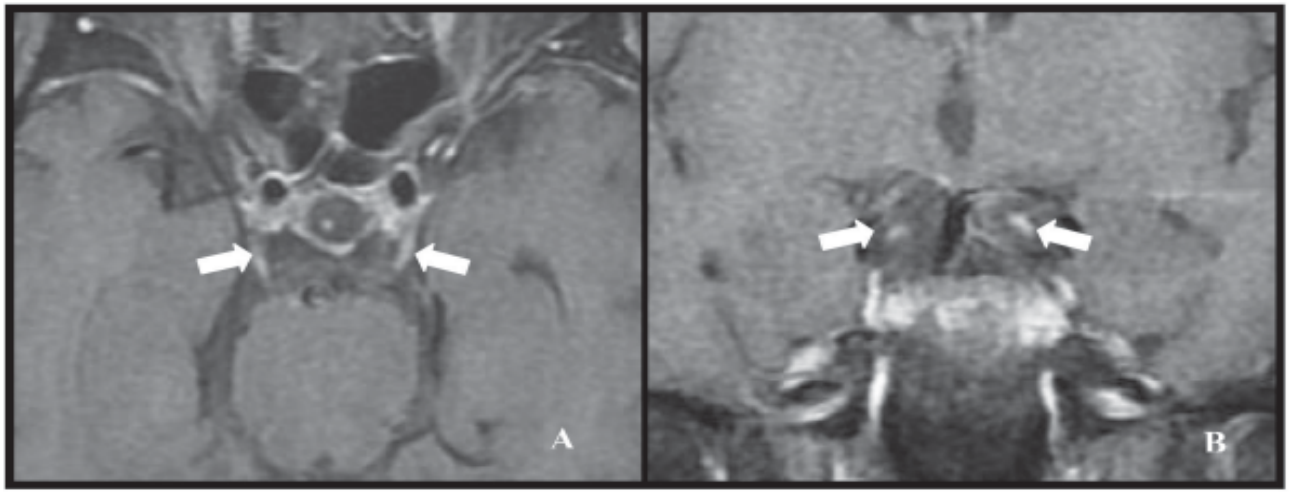

Figure 1. Thin slice axial (A) and coronal (B) FSE T1W with fat suppression shows enhancement of bilateral oculomotor nerves (white arrows).

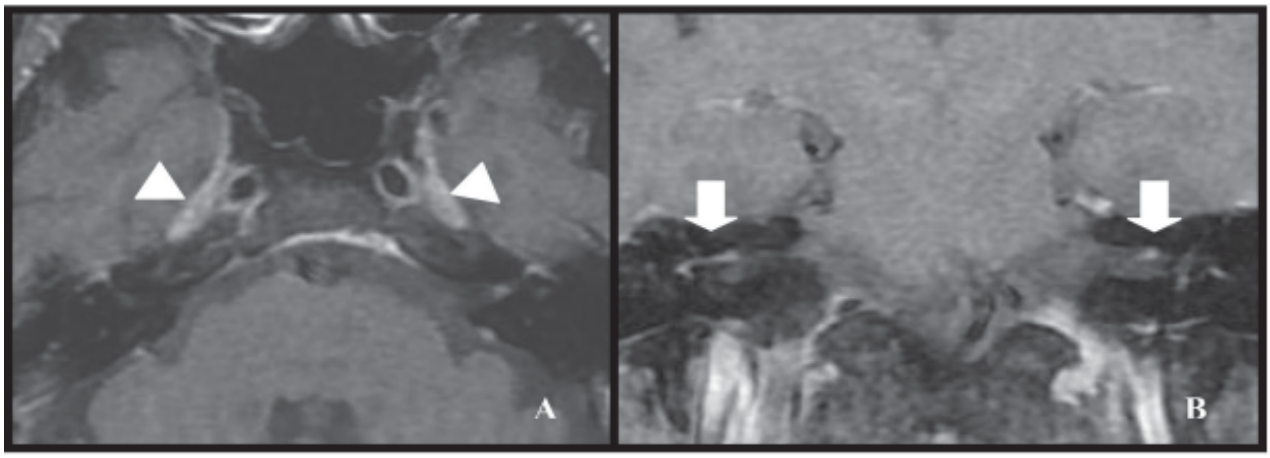

Figure 2. Thin slice axial (A) and coronal (B) FSE T1W with fat suppression shows enhancement of bilateral Meckel's caves (white arrow heads) and facial nerves in bilateral internal acoustic canals (white arrows).

Miller Fisher syndrome was diagnosed based on the clinical triad of ophthalmoplegia, ataxia and areflexia and electrophysiologic evidence of polyneurapathy that involved axon and myelin. The patient underwent intravenous immunoglobulin treatment for five days (total dose of 2 gm/kg). There were much improvement of the ophthalmoplegia and ataxia after the treatment. A follow-up MR was not obtained.

Discussion

Miller Fisher syndrome (MFS) is a cranial nerve variant of Guillain-Barre syndrome (GBS). [1] It has classical triad of ophthalmoplegia, ataxia and areflexia as described by Fisher in 1956. It is often clinically recognizable. The syndrome also overlaps with GBS and it is more limited form involving only components of the triad which have also been widely described. [2] Another related syndrome is Bickerstaff's brainstem encephalitis with alteration of conscious or long tract signs are seen in addition to ophthalmoplegia and ataxia.

Typically, GBS and MFS are usually preceded by infection of the respiratory or gastrointestinal tract, particularly *Campylobacter jejuni* enteritis. [1] Many reports have documented the occurrence after a vaccinations, operations or stressful events.[2]

Its pathogenesis is autoimmune neuropathy, an infection-induced aberrant immune response that damages peripheral nerves. [1] Serum antibodies to various gangliosides have been found in these patients [1], and anti-GQ1b IgG antibodies are sensitive and specific tests for MFS.[2] A previous study demonstrated that anti-GQ1b antibody affected both presynaptic neuronal membranes and perisynaptic Schwann cells of the neuromuscular junction, causing injury by complement mediated complex. [1,2]

Gadolinium enhancement of the cranial nerves has been reported in some literatures.[3 - 5] Coronal and axial thin slice sections or 3D images of the post gadolinium study are likely to be helpful for detection of these abnormalities[4], which thick slice thickness might be hardly detect these findings.

Differential diagnosis of multiple cranial nerves enhancement includes leptomeningeal disease which could be from inflammation, infection or neoplasm in origin.[6] Granulomatous inflammatory diseases that can cause multiple cranial nerves and diffuse leptomeningeal enhancement are Wegener granulomatosis, Behcet disease and sarcoidosis. However, there could be nodular thickening enhancement of cranial nerves and also nodular thickened meningeal enhancement as well as in metastatic process. Infectious meningitis, pyogenic and tuberculous, can also cause multiple cranial nerves and diffuse leptomeningeal enhancements. Herpes zoster can cause multiple cranial nerves enhancement with frequent presence of cutaneous lesions. Metastasis and lymphoma are the differential diagnoses of neoplastic process involving multiple cranial nerves.

References

1. van Doorn PA, Ruts L, Jacobs BC. Clinical features, pathogenesis, and treatment of Guillain-Barre syndrome. Lancet Neurol 2008 Oct; 7(10): 939 - 50
2. Overell JR, Willison HJ. Recent developments in Miller Fisher syndrome and related disorders. Curr Opin Neurol 2005 Oct;18(5): 562 - 6
3. Garcia-Rivera CA, Rozen TD, Zhou D, Allahyari P, Niknam R, Dougherty D, Morgan J, Flanders A. Miller Fisher syndrome: MRI findings. Neurology 2001 Nov; 57(10): 1755
4. Tanaka H, Yuki N, Hirata K. Trochlear nerve enhancement on three-dimensional magnetic resonance imaging in Fisher syndrome. Am J Ophthalmol 1998 Aug; 126(2): 322 - 4
5. Fulbright RK, Erdum E, Sze G, Byrne T. Cranial nerve enhancement in the Guillain-Barre syndrome. AJNR Am J Neuroradiol 1995 Apr; 16(4 Suppl): 923 - 5
6. Adams ME, Linn J, Yousry I. Pathology of the ocular motor nerves III, IV, and VI. Neuroimaging Clin N Am 2008 May; 18(2): 261- 82