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RESEARCH ARTICLE

SYRINGOMYELIA IN NEUROMYELITIS OPTICA SEROPOSITIVE FOR AQUAPORIN-4 ANTIBODY: A CASE REPORT

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Abstract

Neuromyelitis Optica (previously called Devic's disease) is an autoimmune-induced demyelinating disorder of the central nervous system that primarily affects the optic nerves and spinal cord. This manuscript presents a case of a 28-year-old female patient who clinically presented with acute lower extremity weakness and numbness associated with unilateral impairment of vision. Later in the course of the disease, magnetic resonance imaging of the cervico-thoracic spine showed multi-level abnormal non-enhancing and enhancing lesions from levels C3 to T8 levels which were identified to be combined transverse myelitis and cord syrinx. An anti-aquaporin-4 receptor antibody was obtained and yielded to be seropositive. Given the patient's clinical manifestations, combined with imaging and laboratory examinations, led to a diagnosis of Neuromyelitis Optica (NMO). The patient was managed with high-dose steroids. The significant discovery of anti-aquaporin-4 antibodies served as a key factor in the NMO immunopathogenesis. It is currently regarded as a specific biomarker of the diagnostic process, making it distinguishable from multiple sclerosis. This case highlights the mechanism of formation of a fluid-filled syrinx-like cavity in the cord in the setting of Neuromyelitis Optica.

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INTRODUCTION

Neuromyelitis Optica (NMO) is a rare autoimmune demyelinating disease that is severe and debilitating [1]. Clinical presentation and some radiological features are frequently mixed up with multiple sclerosis (MS). The groundbreaking discovery of an auto-antibody biomarker NMO-IgG (AQP4-Ab) has been used to delineate if from MS [2]. On the other hand, syringomyelia is a pathologic response of the spinal cord rather than a disease itself. The exact etiology remained unclear. It is characterized by an accumulation of CSF in the spinal cord that may be progressive, that causes dissociative paresthesiae mainly affecting pain and temperature sensations with preferential involvement of the upper extremities in a cape-like distribution [3].

Concomitant syringomyelia in multiple sclerosis (MS) has been described in various case series or reports. There is extremely limited literature describing syringomyelia coexisting with Neuromyelitis Optica [6-9]. To date, none have been encountered in the local literature. This paper aims to report the first case of a Filipino patient with NMO and coexisting syringomyelia.

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CASE PRESENTATION

Written and informed consent was obtained from the patient and the study was approved by the ethics review board. The patient is a 28-year-old female with no known comorbidities, was admitted to a tertiary hospital with a chief complaint of progressive bilateral lower extremities weakness and hypoesthesia for 1 week. Symptoms were persistent for 2 months, initially occurring intermittently 1-2 times per week, and accompanied with upper back pain characterized as electric-shock-like sensation, from the nape area radiating to the upper back over the shoulders then down the thoracic region. Progression of symptoms became notable after 1 month, associated with the onset of band-like truncal sensory deficit, urinary incontinence, and blurring of the right eye vision. She exhibited impairment of gait and difficulty maintaining an upright position.

Physical examination upon admission revealed normal vital signs as well as, cardiac, respiratory, and abdominal examinations. Neurologic examination suggested a normal mental status. Cranial nerves examination reported impaired visual acuity on the right eye and able to perceive hand movements with positive RAPD on swinging light test whereas, the left eye exam was all normal. The pupillary light reflexes were normal and has fully intact extraocular muscle movements. On ophthalmoscope examination, the disc border over the nasal area of the right eye was paler than the left. The facial muscles activity was symmetrical with no sensory deficit. The hearing, gag reflex, and tongue placement were essentially normal. There was a weakness of the bilateral lower extremities with a muscle strength grade of 3/5 over the proximal part and 1/5 over the distal part. Dermatome sensory examination revealed abnormal findings in the modalities of light touch, pain, and temperature with 50% deficit at the dermatomal areas T5 to T8 and 70% deficit at the dermatomes T9 to S3. Vibration sense was absent over the bony prominences of the lower extremities. Position sense was impaired on both the bilateral upper and lower extremities. Deep tendon reflexes revealed hyporeflexia. Examination of the pathological reflexes recorded a positive Lhermitte's sign and bilateral Babinski.

Complete blood count, blood chemistry, coagulation studies, electrocardiogram, and chest x-ray imaging were essentially normal. Magnetic resonance imaging of the spinal cord suggested the presence of demyelinating cord lesion in segments C3-C4, T1- T2 & T7-T8 (Fig. 1A-C) and syringomyelia from C3- C5 and T2-T6 (Fig. 1D-E). The optic nerve on the MRI was suggestive of optic neuritis (Fig. 2A-B). The fundoscopic examination was done and revealed optic atrophy of the right eye (Fig. 2C) with generalized depression of the visual perimetry (Fig. 2D). Serum NMO-IgG revealed a positive result.

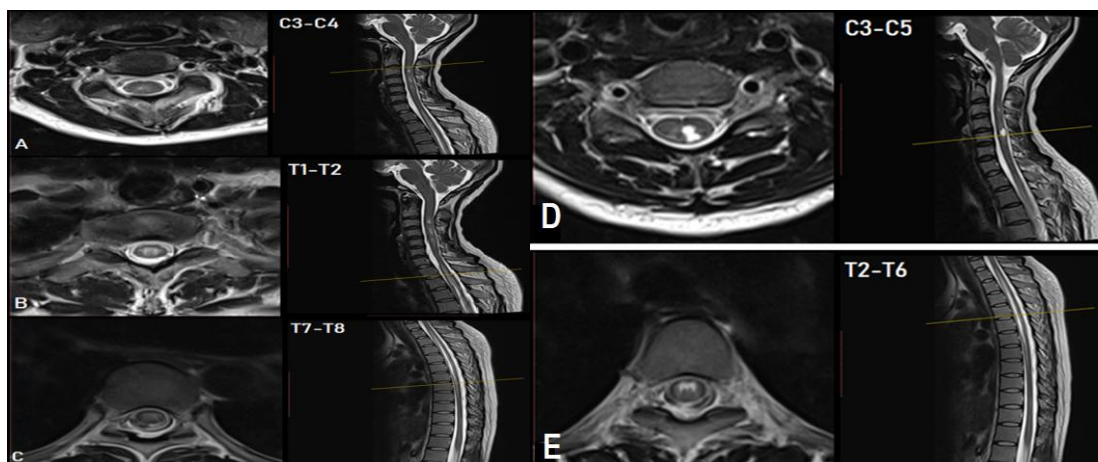


Figure 1. MRI of the cervicothoracic spine shows a T2 hyperintense, mildly non-enhancing intrinsic cord signal abnormality C3-C4 levels, T1-2, and T7-8 levels (A, B, C). Cord Syring C3-5 (D), T2-6 (E).

Based on the clinical manifestations, supported by the immunologic and radiographic examinations, a diagnosis of NMO was confirmed. The patient was managed pharmacologically and given 5-days treatment of high-dose intravenous methylprednisolone pulse therapy (1000 mg daily) and shifted to oral prednisolone at 20 mg daily thereafter, with subsequent improvement of symptoms. Azathioprine of 2.5 mg/kg was started and maintained even

after her being discharged after a month of confinement. On her follow-up 1 month after discharge, her vision recovered to normal, and able to do her usual daily activities. Her hematologic parameters and liver enzymes were normal. Azathioprine 50 mg/tab twice daily was continued and Prednisone was gradually tapered.

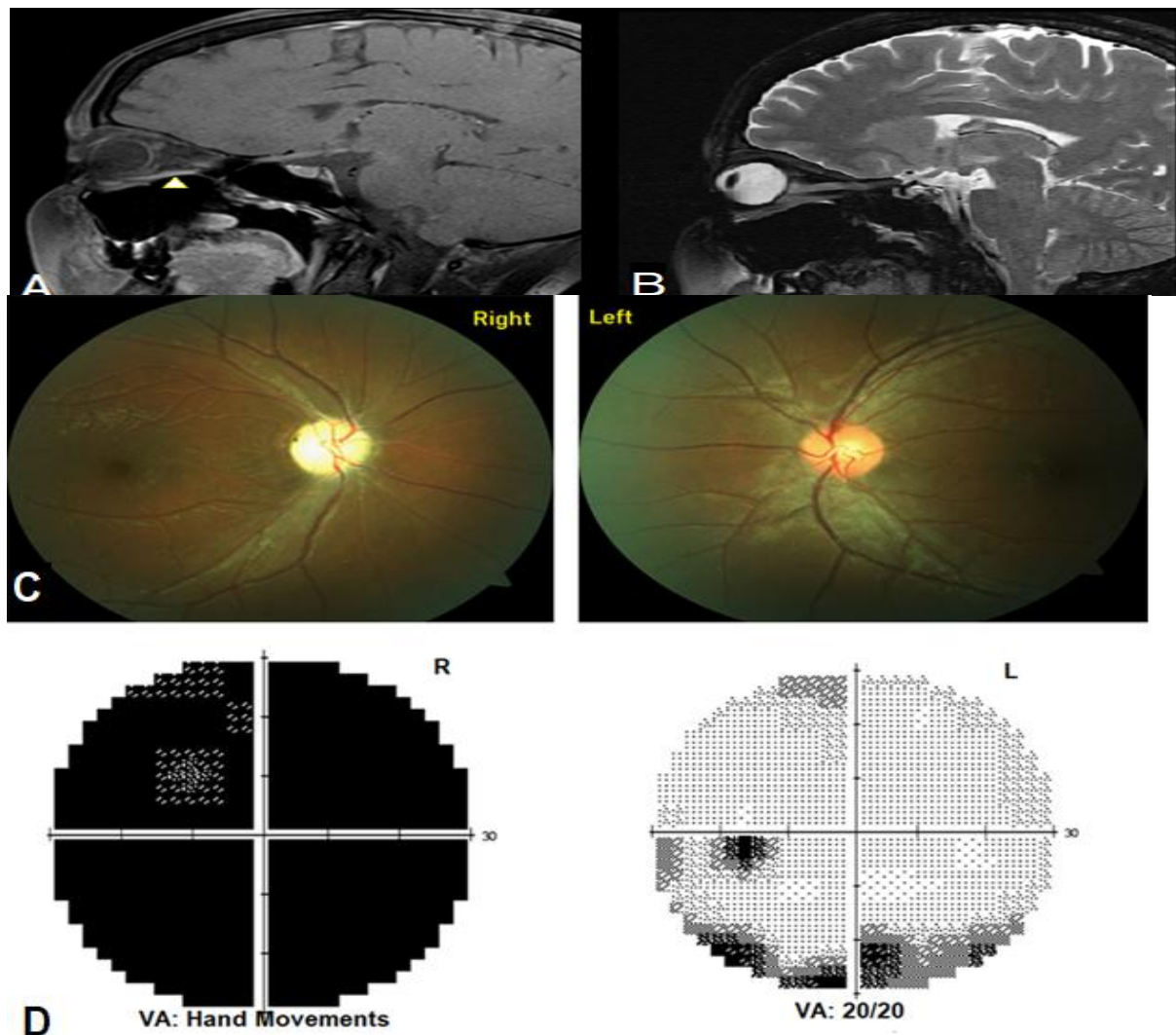


Figure 2. Contrast-enhanced T1 (Fat suppressed-orbital cuts) images show mild enhancement of the optic nerve on the right with arrowhead (A) and T2 orbital cuts show hyperintensity involving the right optic nerve (B). Disc photo of the right eye shows characteristic pale, “flat white” optic disc, reduced blood vessels on the surface, and attenuation of peripapillary blood vessels, suggestive of optic atrophy (C). Visual field map showing a generalized depression on the right eye and a normal field map on the left eye, take note that the VA of the patient on the right eye is HAND movement (D).

DISCUSSION

The case presented arrived at a diagnosis of NMO, by following the revised criteria proposed to have been commonly used for years, including 2 of the absolute symptoms of optic neuritis (ON) and acute myelitis, and at least two of three supportive criteria including contiguous spinal cord lesion of no less than 3 vertebral segments via MRI, brain MRI demonstrating lesions inconsistent of multiple sclerosis, and seropositivity of NMO-IgG [4]. In the

previous study, the detection of the AQP4 serum autoantibody yields a sensitivity of 91% and specificity of 100% for NMO and is positive in up to 80% of patients [5].

The patient presented also developed syringomyelia in the cervical and thoracic spine. The combination of the extensive demyelinating lesion (mild non-enhancing intrinsic cord signal on C3-C4 levels, T1-2, and T7-8 levels) and the formation of cord syrinx (enhancing lesion on C3-5 and T2-6) may not be just coincidental and considered to have a connection in the mechanism of syringomyelia formation in the setting of NMO.

There were previous studies reported on patients with radiological findings of syringomyelia in the setting of NMO [6-9]. Previous studies suggested that AQP4-IgG antibody binding to astrocytic AQP4 and localization to the central gray matter with high expression of AQP4, significantly contribute to the pathogenesis of NMO and may facilitate the formation of syrinx through cascade activation including complement-dependent cytotoxicity (CDC), the infiltration of inflammatory cells, blood-brain barrier disruption, oligodendrocyte injury, and demyelination

CONCLUSION

This report described the first case of a patient in the Philippines with NMO combined with syringomyelia from C3 to T8 vertebral segments. Neuromyelitis Optica is a severely disabling condition, if undetected may succumb to incomplete recovery and irreversible disability. In this perspective, utilizing the diagnostic criteria and detection for anti-aquaporin-4 antibodies are of paramount importance to the diagnosis and management of NMO. Furthermore, our case report exemplifies that radiologic findings of syringomyelia should warrant clinicians to further investigate and consider the possibility of NMO.

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