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CASE REPORT

A 24 YEAR OLD FEMALE WITH NEURO MYELITIS OPTICA FROM ETHIOPIA

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ABSTRACT

A 24 year old female patient from Bedelle, Oromia region, Ethiopia presented with progressive and recurrent quadriparesis of a five year duration and was diagnosed to have acute myelitis and neuritis, Neuromyelitis optica (NMO). To our knowledge NMO has never been reported from Ethiopia. Hence, we discuss the case presentation in light of the available literature on the topic.

Key words: *Neuro Myelitis Optica, Ethiopia.*

INTRODUCTION

Neuromyelitis optica (NMO) is an autoimmune inflammatory disorder in which a person's own immune system preferentially attacks the optic nerves and spinal cord resulting in optic neuritis and myelitis. This condition is also called Devic's disease or Devic's syndrome. NMO is up to nine times more prevalent in women with a median age of onset of 39 years. In contrast to Multiple Sclerosis (MS), NMO is relatively common in non-whites (1-3). There are few case reports from Africa (4,5), perhaps because it is actually challenging to diagnose in resource limited settings.

CASE REPORT

Patient X was a 24 year old female from Bedelle, Oromia region, Ethiopia who worked as a cashier. She presented to Tikur Anbessa Teaching Hospital on May 06, 2015 with a progressive quadriparesis involving the left arm, left leg, right arm, and right leg in that order, over a period of two weeks. She had lost her left eye vision progressively one week prior to the weakness. She also had associated tingling, numbness and burning sensation over her left upper limb. She had experienced similar attacks five years ago and another, just six months earlier, with fecal and urinary incontinence which improved spontaneously over subsequent months. She was married and nulliparous.

Her past medical history showed no personal or family history of diabetes mellitus, hypertension, or cardiac illness. She did not have exposure to toxins or ever travelled out of Ethiopia. On physical examination she was noted to have complete visual loss of the left eye with prominent retinal vessels and pale background on fundoscopic examination. All four limbs were flaccid with quadriparesis, with positive Babinski sign bilaterally. There was also sensory (touch, temperature and pain) loss of the left side of the body up to the mandible but there was no gait disturbance. The rest of the systemic examination was unremarkable. Her laboratory investigations revealed normal complete blood count, liver function test, renal function test, serum electrolytes, and plasma glucose. She was also negative for syphilis (VDRL) and HIV tests. Her brain CT scan and MRI findings were normal but cervical MRI (Figure1) revealed a hyper intense signal intensity (arrow) intramedullary lesion on T2W1 extending from C1-C7 vertebra. It was hypo intense on T1W1. The spinal cord was enlarged but the cervico-medullary junction was preserved.

The MRI of the patient also showed a left side atrophied optic nerve.

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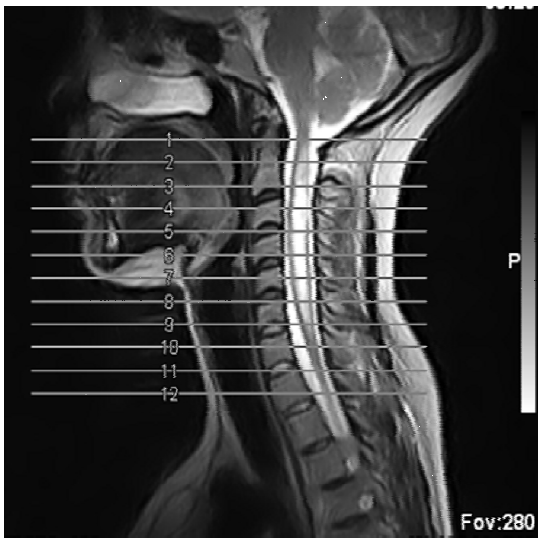

Figure 1. T2W1 MRI of cervical spine of the patient -hyper intense signal intensity (arrow) intramedullary lesion extending from C1-C7 vertebra

DISCUSSION

Neuromyelitis optica (NMO) is considered as a clinical variant of Multiple Sclerosis (MS). Whereas MS is a CD4+T-cell mediated autoimmune disease, NMO is a humorally mediated autoimmune disease. (1)

The serum auto-antibody marker, neuromyelitis optica-immunoglobulin G (NMO-IgG), appears specific for NMO. The condition predominantly affects females, and has a median age of onset of 39 years. It is relatively more common in non-whites compared to MS. In NMO, attacks of optic neuritis occur more commonly unilaterally than bilaterally. MRI findings of the brain of NMO patients are usually normal at the onset of the disease (1-3).

Prognosis of NMO is determined by a number of factors (1,7). These are:

- The number of relapses in the first 2 years of disease activity
- The severity of the first attack.
- Concomitant Systemic Lupus Erythematosus or a related non-organ specific autoimmune disorder.
- High titer auto-antibodies (NMO IgG).

Diagnosis of NMO requires two absolute criteria, plus at least two of three supportive criteria. (1,6)

Absolute criteria:
1. Optic neuritis
2. Acute myelitis

Supportive criteria:
1. Brain MRI not meeting criteria for MS at disease onset
2. Spinal cord MRI with contiguous T2-weighted signal abnormality extending over 3 or more vertebral segments, indicating a relatively large lesion in the spinal cord
3. NMO-IgG seropositive status.

A case, which fulfills the above criteria, is 99% sensitive and 90% specific for Neuromyelitis optica.

In light of the above, the case presented here fulfilled the criteria for a diagnosis of NMO; the sole criteria which was not met was a positive serostatus for NMO-IgG. The test was not done because it was not available in the country at the time of diagnosis.

Intravenous corticosteroid therapy is commonly the initial treatment for acute attacks and plasmapheresis is recommended for patients with NMO having severe cervical myelitis, or for those refractory to steroids. Maintenance immunosuppressive therapy for reducing relapses of NMO with azathioprine (2·5-3·0 mg/kg/day) plus oral prednisone (1·0 mg/kg/day) is the recommended treatment. Mitoxantrone, intravenous immunoglobulin, and rituximab can induce clinical remission of NMO in patients who are treatment-naive or who have a relapse despite other treatments (1,8).

Our patient was treated with azathioprine and prednisolone after 5 days of IV methylprednisolone and recovered some strength in her limbs although there was no improvement in visual loss. Her follow up after 7 months of treatment revealed that all extremity strength had significantly improved; the muscle power had reached 4+ out of 5 on the right lower limb and 3+ out of 5 on the left lower limb with deep tendon reflexes, 2 out of 4 bilaterally. There was still no gait disturbances but she had fecal and urinary incontinence, which caused emotional stress to the patient.

A similar case has been reported in a 17-year-old girl from a study in Greece who presented with acute-onset cervical pain, followed by left arm weakness and gait disturbances. A diagnosis of spinal cord astrocytoma was made using MRI performed at an outpatient facility. The patient was admitted to the neurosurgery department for a spinal cord biopsy (9). However, a second neurologic evaluation of this patient indicated neuromyelitis optica (NMO) as the

most likely diagnosis; this was confirmed by NMO–IgG seropositivity. The patient was then treated with rescue plasmapheresis with substantial clinical and radiologic improvement (Fig. 2 and 3).

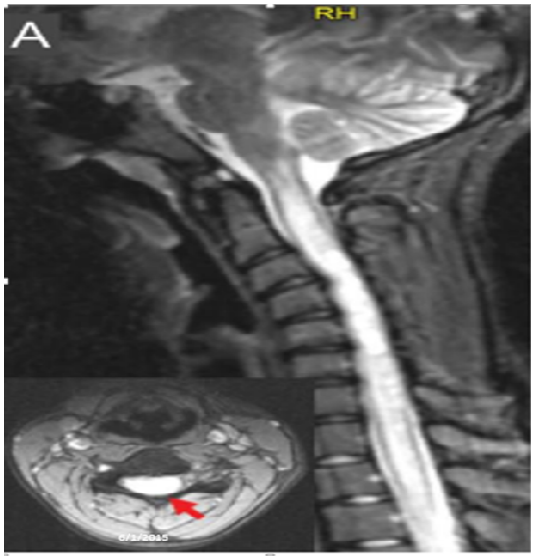

Figure 2. T2W1 MRI of the patient from the Greek study, diagnosed as an extensive (C2-T4) hyper intense lesion involving nearly the whole cervical cord cross section.

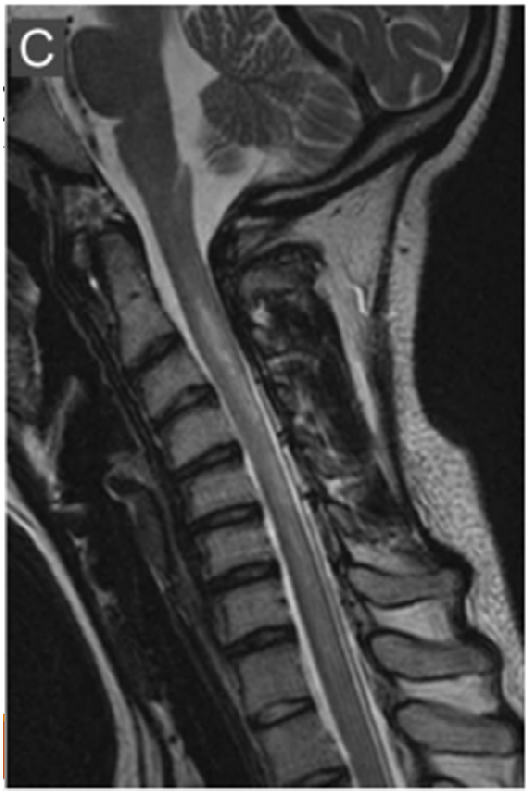

Figure 3. T2W1 MRI of the cervical spine of Patient from the Greek study with radiologic improvement after plasmapharesis.

An NMO case report has also been described from Uganda (4). The case was a 24-year-old woman of Bantu origin who presented initially with bilateral loss of vision and weakness of the lower limbs in 2010 that resolved completely after a few days. Eight months later, she presented with bilateral lower limb weakness and urinary incontinence. Spinal MRI T2 weighted magnetic resonance imaging showed hyper intense ill-defined lesions from T4–T8, similar to our patient and the case from the USA. Her symptoms improved completely following steroid treatment. However, she developed an episode of quadriparesis four months later, which improved with steroids and azathioprine with some residual weakness requiring a walking aid.

From these cases, it is wise to consider NMO in patients (especially females) with unexplained recurrent quadriparesis and optic neuritis. Accordingly spinal MRI and serologic tests should be considered before starting patients on a specific treatment regimen.

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