Neuromyelitis optica mimicking multiple sclerosis-presentation of two cases

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Abstract

Introduction: Neuromyelitis optica (NMO) or devic disease is a relatively uncommon neurologic disease that often mimicks multiple sclerosis (MS) in clinical practice. It usually presents with recurrent attacks of optic neuritis and myelitis but unlike MS the attacks are often associated with some degrees of sequelae and there is no involvement of nervous system outside of the optic nerves and spinal cord particularly in the earlier phase of the disease. the constellation of findings including characteristic clinical behavior, imaging findings of the brain and spinal cord and the recent finding of antibody against aquaporin-4 water channels differentiates this disease from MS. Response of the attacks to corticosteroid may be inadequate and we may need plasma exchange for better symptomatic remission of acute attacks. To prevent further attacks, prophylactic treatment with immunosuppressive agents may be helpful .

Two cases of NMO with characteristic clinical and neuroimaging features are presented that mimicked MS at first and with good response to plasma exchange despite the inadequate improvement following corticosteroid pulse therapy.

Keywords: **Neuromyelitis optica-multiple sclerosis-optic neuritis-myelitis-corticosteroid plasma exchange**

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Introduction

NMO or devic disease is a relatively uncommon neurologic disease that often mimicks MS at first. It presents with recurrent attacks of optic neuritis and myelitis in more than 90% of cases. Like MS it is more common in women but the average age of onset is somewhat more in the late fourth decade. Unlike MS it is more often seen in non Caucasians.(1-2)

The main clinical features of NMO are optic neuritis and myelitis which unlike MS are more severe and associated with more residual symptoms even after acute treatment . there is no prominent and early involvement of central nervous system outside of optic nerve and spinal cord and if present they are of mild severity and late in the course of disease.(1-2-6-9)

Added to the mentioned different clinical course, the characteristic MRI findings of the brain and spinal cord also help to differentiate this disease from MS. MRI of the brain is normal in the early stages of the disease or shows just few lesions which are not diagnostic for MS according to the defined criteria. It should be noted that after months to years of the onset of NMO the extent of brain lesions may fulfill the criteria of MS and so it is important to consider brain MRI in the early stages of the disease. (1-3-7)

Spinal MRI findings are the most helpful diagnostic test in NMO. Spinal cord involvement in 3 or more continuous segment in T2 images markedly increases the probability of NMO and so is a characteristic finding in the presence of suspected clinical picture. In contrary the spinal lesions of MS are usually small, acraniocaudal length of spinal cord.(7-8)

Analysis of cerebrospinal fluid particularly in severe attacks of myelitis may show marked pleocytosis (50-2000/mm) with neutrophilic predominance.(3)

Antibody of IgG type against aquaporin-4 water channels has a high index of sensitivity and specificity and helps a lot for diagnosis of NMO.(4-6)

Although for definite diagnosis of NMO the presence of optic neuritis and myelitis are both required but in the earlier phase of the disease there may be isolated optic neuritis or myelitis. Can we consider NMO in these patients?

In patients presenting with isolated optic neuritis the diagnosis of NMO should be considered in the following cases:

1/ The first attack of optic neuritis with no expected response to corticosteroid pulse therapy.

2/ Recurrent attacks of optic neuritis in the presence of normal MRI or what is nondiagnostic for MS.

3/ Simultaneous bilateral optic neuritis.

4/ Both optic nerves are involved consecutively with less than one month interval.

In patients with isolated or recurrent myelitis the diagnosis of NMO should be considered when there is severe transverse myelopathy involving three or

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more segments of the cord (more in cervical cord) and particularly in cases of respiratory compromise and hiccups as part of clinical picture. Again it is worth to say that myelpathy in MS patients is characteristically partial and involving dorsolateral cord usually presenting as asymmetric paraparesis and giving better response to corticosteroid pulse therapy comparing with NMO.[6-9-10]

The primary choice of treatment for acute attacks of optic neuritis and myelitis in NMO is methyl prednisolone (MPN) pulse therapy usually1 gram daily for 5 to 7 days but one should know that there may not be a good response to such a treatment. In theses cases we may switch to plasma exchange which may actually provide a better therapeutic response in many cases. Then prophylactic treatment with cytotoxic agents and especially azathioprine may be needed to prevent further disabling attacks.[8-9-10]

Case presentation

Case one: a 16 year old lady wad admitted with acute quadriplegia from one weeks ago in neurology department of Ahvaz Golestan hospital. In neurological exam there was flaccid quadriplegia , overflow urinary incontinence, T4 sensory level associated with secondary optic atrophy in right eye. There was a past history of admission for optic neuritis of right eye in the same center with just slight improvement after MPN pulse therapy. Cervicothoracic MRI showed a large heterogeneous high signal lesion in T2 images extending from almost upper thoracic to midcervical areas with slight enhancement after gadolinium injection (figs1-1 and 1-2). Brain MRI was normal at this time (fig1-3).

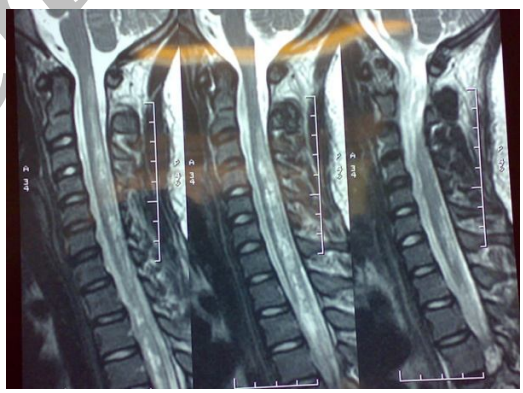

Figure (1-1): Heterogeneous high signal lesion extending from midcervical to thoracic cord

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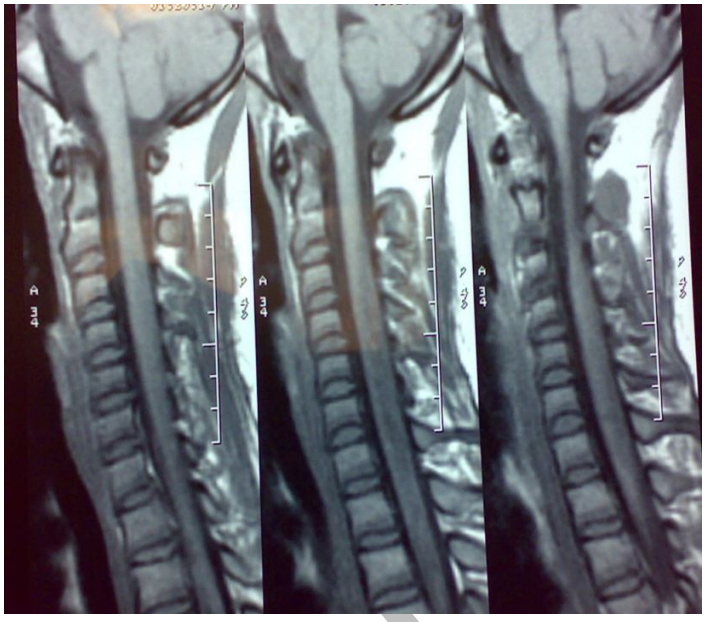

Figure (1-2) :Low signal lesion in T1 sequence as described

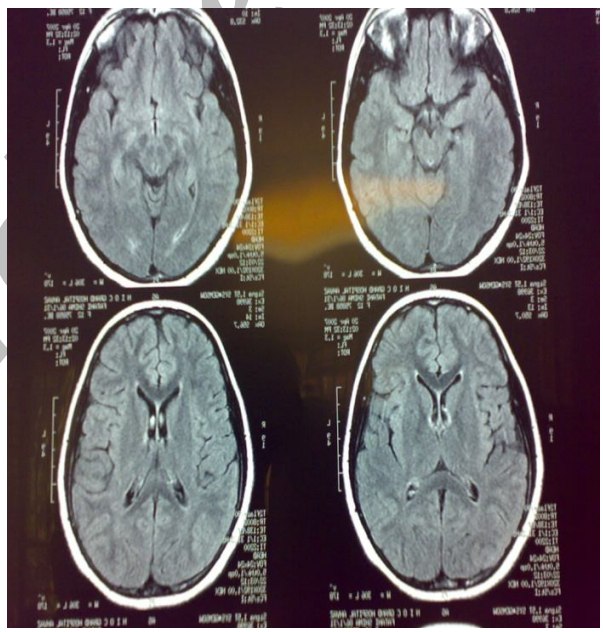

Figure (1-3): Brain M.R.I was normal

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There was no systemic symptoms and signs and laboratory findings including vasculitic and infectious tests were normal. She was given a 7 day course of MPN pulse therapy one gram daily with no motor improvement after 2 weeks. According to tha clinical and MRI features, plasma exchange 2 liters per day was started for 7 days and we noted beginning of improvement after just 1 week. She was able to walk unaided after 3 weeks and now she is able to do sport activities with good quality after about 10 months of her attack. She is now on azathioprine 125 mg per day with no further attack.

Case 2: A 40 year old woman referred with acute painful left sided vision loss. The examination showed marked afferent papillary defect and normal fundoscopy. There was also evidence of chronic myelopathy such as spastic paraparesis, sphincter disorder, hypereflexia and bilateral babinski sign. There was a history of admission in ICU about 1 year ago with quadriplegia and respiratory failure requiring mechanical ventilation treated as the diagnosis of severe myelitis in cervical cord based on characteristic MRI findings (fig 2-1) with MPN pulse therapy in that time with substantial partial improvement.

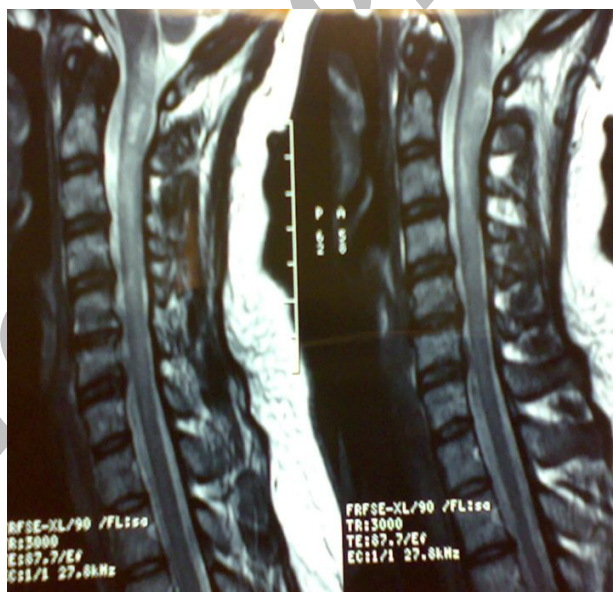

Figure (2-1): Severe extensive myelitis in long segment of cervical cord in T2 sequence

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She was again treated as retrobulbar optic neuritis with MPN pulse therapy. Brain MRI was normal but cervical MRI showed residual marked cord atrophy as the sequelae of the previous severe cervical myelitis (fig 2-2).

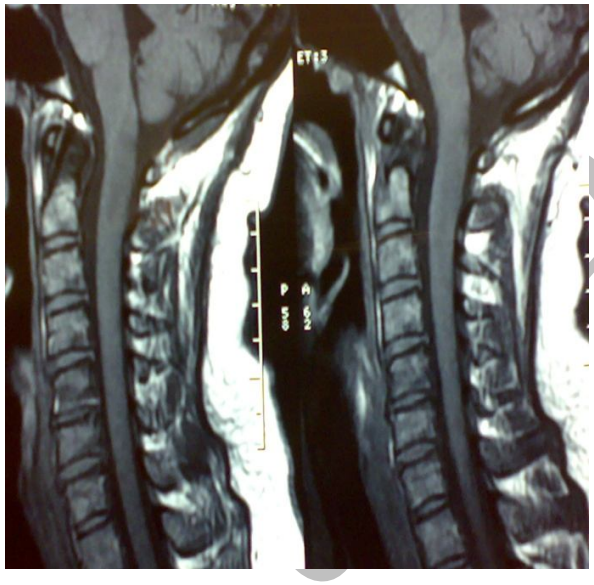

Figure (2-2): Post-inflammatory atrophy of cervical cord after several months

Since she didn't get good response of optic neuritis to pulse therapy she was started on plasma exchange with favorable response beginning after 2 weeks and continued for the next 2 months.

Discussion:

NMO which had been introduced as a form of MS in past years is now recognized as a seperate entity with characteristic clinical and neuroimaging features and different therapeutic implications. The role of autoimmune response against water channels has been recognized in the past years and there is growing evidence that it is a separate entity different in many features with MS. Rapid diagnosis and early treatment which can be different compared with MS can lead to better therapeutic response and improve the outcome of these patients.

We presented two cases of NMO with particular emphasis on the differentiating clinical and radiologic features helpful in early diagnosis of such patients and also the probability of good response to plasma exchange in cases not responsive to corticosteroid

In the first case the association of acute myelitis, previous optic neuritis and characteristic spinal cord lesions in the presence of normal brain MRI and

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exclusion of other diagnoses was enough to make a diagnosis of NMO and to start treatment protocols accordingly.

In the second case the patient had presented before with a typical attack of severe cervical myelitis culminating in respiratory failure and after the recent attack of optic neuritis the true nature of the disease became clear .

Similar in both cases was the good response to plasma exchange despite the previous unsatisfactory improvement with corticosteroid pulse therapy.

Conclusion:

Although optic nerve and spinal cord involvement in NMO superficially resembles MS and even it had been considered as a variant of MS before, the following features seperates it from MS:

1/ Sparing of nervous system other than optic nerve and spinal cord in the earlier stages

2/ The severity of the attacks and tendency to put residual sequelae

3/ Continuous involvement of 3 or more segments of spinal cord in MRI

4/ Normal or nondiagnostic brain MRI at the onset of disease

5/ positive NMO-IgG antibody against aquaporin-4 water channels.[7-9]

Overall in patients referred with optic neuritis or myelitis, one should think of NMO in the following cases:

1/ The first attack of severe optic neuritis with no good response to corticosteroid pulse therapy as we expect in MS patients.

2/ Recurrent attacks of optic neuritis in the presence of normal MRI or the one that is not diagnostic for MS.

3/ Recurrence of optic neuritis in the opposite eye in less than one month which is not typical for MS.

4/ Severe and complete transverse myelitis with involvement of 3 or more esegments in spinal cord MRI .

5/ Myelitis associated with respiratory failure or hiccup which indicated the severity of involvement.

It is worth to say that diagnosis of NMO is by exclusion and other disorders such as vasculitis, postviral autoimmune encephalomyelitis, infectious disorders , celiac disease and others should be ruled out .

The importance of correct diagnosis of NMO is in different prognostic and therapeutic implications of this chronic disabling disorder. Plasma exchange may provide substantial improvement for acute attacks in patients not responsive to corticosteroids. Prophylactic treatment with interferons have not shown hopeful efficacy in the previous studies although it needs more studies to judge in this regard. Cytotoxic therapy with or without prednisolone are recommended as prophylactic therapy. In this regard azathioprine has shown the best efficacy.[9-10]

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معرفی دو مورد نورومیلیت اپتیکا بعنوان مقلد بیماری M.S

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چکیده

سابقه و هدف: نورومیلیت اپتیکا (NMO) یا بیماری Devic یک بیماری نسبتاً ناشایع نورولوژیک است که اغلب بیماری MS را تقلید می‌کند. این بیماری معمولاً خود را با حملات راجعه نوریت اپتیک و میلیت نشان می‌دهد. برخلاف بیماری MS، حملات بیمار اغلب با ایجاد سکل همراه است و گرفتاری سیستم عصبی مرکزی خارج از عصب اپتیک و نخاع بخصوص در اوایل بیماری وجود ندارد. رفتار بالینی متمایز بیماری، یافته‌های تصویرنگاری مغز و نخاع و اخیراً یافتن آنتی‌بادی مختص بیماری از نوع IgG برعلیه کانالهای آب Aquaporin – 4 این بیماری را به عنوان یک بیماری مجزا از MS مشخص کرده است. پاسخ حملات بیماری به کورتیکواستروئید ممکن است ناکافی باشد و به پلاسمافرز جهت درمان علامتی بهتر در حملات حاد نیاز باشد. جهت پیشگیری از بروز حملات مجدد، درمان پروفیلاکتیک با ایمون ساپرسیو توصیه می شود در این مقاله دو مورد بیماری NMO با حملات نوریت‌اپتیک و میلیت همراه با یافته‌های تصویر نگاری مشخص گزارش می‌شود که در ابتدا بیماری MS را تقلید کردند و علیرغم عدم پاسخ به پالس متیل‌پرونیزولون، به پلاسمافرز پاسخ خوبی نشان دادند.

واژگان کلیدی: نورومیلیت اپتیکا – ام اس – نوریت اپتیک – میلیت – کورتیکواستروئید – پلاسمافرز

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