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Original Research Article

A Clinical Profile of Neuromyelitis Optica and Neuromyelitis Optica Spectrum Disorders in a Tertiary Care Hospital in South Tamil Nadu

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Abstract

Background: Neuromyelitis optica (NMO) and neuromyelitis optica spectrum disorders (NMOSD) are severe inflammatory demyelinating disorders predominantly affecting the optic nerves and spinal cord. Data on their clinical profile and outcomes in South Tamil Nadu remain limited. This study aimed to evaluate the clinical characteristics, serostatus, and functional outcomes of NMO/NMOSD patients presenting to a tertiary care center in this region.

Materials and Methods: This retrospective observational study included 25 patients diagnosed with NMO/NMOSD between January 2016 and May 2019. Patients fulfilling revised Wingerchuk criteria were analyzed for clinical presentation, laboratory findings, neuroimaging, treatment response, and outcomes. Disability was assessed using the Expanded Disability Status Scale (EDSS) at the last follow-up.

Results: Of the 25 patients, 7 (28%) were anti-aquaporin-4 antibody positive and 18 (72%) were seronegative. The female-to-male ratio was 2.57:1, with a median age of onset of 35 years. Combined myelitis and optic neuritis at presentation was significantly more frequent in seronegative patients (28%; p=0.047). Cervico-dorsal spinal cord involvement was observed in 68% of patients with myelitis. All patients received intravenous methylprednisolone; additional therapy included repeat steroids (40%), plasmapheresis (8%), and rituximab (4%). Median EDSS at last follow-up was lower in seronegative patients (3.5) than seropositive patients (5.0), though not statistically significant (p=0.14).

Conclusions: Seronegative NMO/NMOSD constituted the majority of cases in this South Tamil Nadu cohort and more commonly presented with combined myelitis and optic neuritis. Functional outcomes were comparable between seropositive and seronegative patients. Larger prospective studies are required to clarify regional disease patterns and optimize management strategies.

Keywords: Neuromyelitis optica, NMOSD, Myelitis, Optic neuritis, Aquaporin-4 antibody, EDSS, South Tamil Nadu, Demyelinating disorders.

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Introduction

Neuromyelitis optica (NMO), also known as Devic's disease, is a severe inflammatory demyelinating disorder of the central nervous system that predominantly affects the optic nerves and spinal cord. First described by Eugène Devic in 1894, NMO was historically considered a variant of multiple sclerosis (MS). However, the discovery of the highly specific anti-aquaporin-4 (AQP4) antibody in 2004 revolutionized our understanding of NMO as a distinct autoimmune astrocytopathy with unique pathophysiology, clinical features, and treatment requirements.[1,2]

The clinical spectrum of NMO has expanded significantly with the recognition of neuromyelitis optica spectrum disorders (NMOSD), which encompass patients presenting with limited forms of the disease, including isolated longitudinally extensive transverse myelitis (LETM), recurrent or bilateral optic neuritis, and various combinations of core clinical characteristics. The 2015 International Panel for NMO Diagnosis revised the diagnostic criteria to include both seropositive and seronegative NMOSD, acknowledging that approximately 10-40% of patients meeting clinical

criteria may be seronegative for AQP4 antibodies.[3,4]

NMO demonstrates significant geographical and ethnic variations in prevalence and clinical characteristics. The disease is more common in non-Caucasian populations, particularly among Asians, African Americans, and Latin Americans, with prevalence rates ranging from 0.5 to 4.4 per 100,000 population.[5,6] Studies from Asia have reported distinct clinical features compared to Western populations, including higher rates of seropositivity, more severe relapses, and different patterns of organ involvement. However, comprehensive data on NMO and NMOSD from the Indian subcontinent, particularly from the Southern region, remain limited.[7,8]

The pathophysiology of NMO involves antibody-mediated destruction of astrocytes through binding of AQP4-IgG to aquaporin-4 water channels, which are highly expressed on astrocytic foot processes at the blood-brain barrier. This binding triggers complement activation, inflammatory cell recruitment, and subsequent demyelination and neuronal damage. The characteristic pathological hallmark is extensive necrosis with cavitation, loss of aquaporin-4 and glial fibrillary acidic protein immunoreactivity, and relative preservation of axons in acute lesions.[9,10]

Clinical manifestations of NMO typically include acute or subacute bilateral or severe unilateral optic neuritis, often with poor visual recovery, and transverse myelitis extending over three or more vertebral segments on spinal MRI (LETM). Additional core clinical characteristics may include area postrema syndrome (intractable nausea, vomiting, or hiccups), acute brainstem syndrome, symptomatic narcolepsy or acute diencephalic syndrome, and symptomatic cerebral syndrome.[11,12] Unlike MS, NMO attacks are often more severe and result in greater residual disability, making early diagnosis and aggressive treatment crucial for preventing permanent neurological damage.

Treatment strategies for NMO have evolved significantly with improved understanding of disease mechanisms. Acute attacks are typically managed with high-dose intravenous corticosteroids, with plasmapheresis reserved for steroid-unresponsive cases. Long-term immunosuppression is essential to prevent relapses and disability accumulation, with azathioprine, mycophenolate mofetil, and rituximab being commonly used agents. Recently, targeted therapies including eculizumab, inebilizumab, and satralizumab have shown promising results in clinical trials and received regulatory approval in various countries.[13,14,15]

Despite the growing global literature on NMO, there remains a paucity of data characterizing the clinical profile, treatment responses, and outcomes of patients in South India. Regional variations in genetic background, environmental factors, and healthcare access may influence disease presentation and prognosis. This study aims to systematically analyze the clinical characteristics, serology status, imaging findings, treatment responses, and functional outcomes of patients with NMO and NMOSD presenting to a tertiary care center in South Tamil Nadu, thereby contributing to the understanding of this devastating disorder in the Indian context.

Materials and Methods

Study Design and Setting: This retrospective observational study was conducted at a tertiary care hospital in South Tamil Nadu, India, after approval from the institutional ethics committee. The study adhered to the Declaration of Helsinki guidelines and included patients diagnosed between January 2016 and May 2019.

Patient Selection: Medical records of patients diagnosed with neuromyelitis optica (NMO) or neuromyelitis optica spectrum disorders (NMOSD) were reviewed. Patients fulfilling the revised Wingerchuk diagnostic criteria, including both AQP4-IgG seropositive and seronegative cases, were included. Eligibility required the presence of at least two core clinical features: optic neuritis, acute myelitis, or longitudinally extensive transverse myelitis (LETM ≥3 vertebral segments on MRI).

Patients below 12 years of age, those with other demyelinating disorders, incomplete records, or coexisting systemic autoimmune diseases were excluded.

Data Collection: Demographic details, clinical presentation, symptom duration, relapse history, and neurological features of optic neuritis and myelitis were documented from medical records.

Laboratory and Neuroimaging Evaluation: Serum anti-aquaporin-4 antibodies were detected using cell-based assays. Cerebrospinal fluid analysis included cell count, protein, glucose, and oligoclonal bands where available. MRI of the brain and spinal cord was performed in all patients using standard protocols to assess lesion location, extent, and presence of LETM.

Treatment and Outcome Assessment: Acute relapses were treated with intravenous methylprednisolone (1 g/day for 5 days). Non-responders received repeat steroids, plasmapheresis, or rituximab as escalation therapy. Long-term immunosuppressive therapy was initiated in all patients. Functional outcome was

assessed using the Expanded Disability Status Scale (EDSS) at the last follow-up visit. Secondary outcomes included treatment response and relapse frequency.

Statistical Analysis: Data were analyzed using descriptive statistics. Continuous variables were expressed as median and interquartile range, while categorical variables were presented as frequencies and percentages. Group comparisons were performed using the Mann–Whitney U test and Fisher's exact or chi-square test. A p-value <0.05 was considered statistically significant.

Results

Demographics and Clinical Characteristics: During the study period from January 2016 to May 2019, a total of 25 patients met the inclusion criteria and were included in the final analysis. The demographic and baseline clinical characteristics of the study population are summarized in Table 1. The cohort demonstrated a female predominance with a female to male ratio of 2.57:1 (18 females and 7 males). The median age at disease onset was 35 years, with an interquartile range of 24 to 45 years, indicating that the disease primarily affected adults in their third and fourth decades of life.

Regarding antibody status, 7 patients (28%) were seropositive for anti-aquaporin-4 antibodies, while the majority, 18 patients (72%), were seronegative. This relatively high proportion of seronegative cases is noteworthy and may reflect regional characteristics, testing methodologies, or the timing of antibody testing in relation to disease activity. The seronegative patients satisfied clinical and radiological criteria for NMO spectrum disorders according to the revised diagnostic criteria.

Table 1: Demographic and Baseline Characteristics of Study Population

Characteristic	Value (N=25)
Age at onset (years), median (IQR)	35 (24-45)
Female : Male ratio	2.57:1 (18:7)
AQP4-IgG seropositive, n (%)	7 (28%)
AQP4-IgG seronegative, n (%)	18 (72%)

Clinical Presentation: The initial clinical presentation varied among patients, with significant differences observed between seropositive and seronegative groups as detailed in Table 2. Combined myelitis and optic neuritis at presentation was observed in 7 patients (28% of total cohort), and this pattern was significantly more common among seronegative patients compared to seropositive patients (p=0.047). Isolated myelitis was the presenting feature in 10 patients (40%), while isolated optic neuritis occurred in 8 patients (32%). Among the 18 seronegative patients, 6 patients (33.3%) presented with combined myelitis and optic neuritis, 7 patients (38.9%) presented with isolated myelitis, and 5 patients (27.8%) presented with isolated optic neuritis.

In contrast, among the 7 seropositive patients, only 1 patient (14.3%) presented with the combined syndrome, 3 patients (42.9%) had isolated myelitis, and 3 patients (42.9%) had isolated optic neuritis. The higher frequency of combined presentation in seronegative cases represents a notable finding that warrants further investigation.

Table 2: Clinical Presentation Patterns in Seropositive and Seronegative Patients

Clinical Presentation	Seropositive (n=7)	Seronegative (n=18)
Combined myelitis and optic neuritis, n (%)	1 (14.3%)	6 (33.3%)
Isolated myelitis, n (%)	3 (42.9%)	7 (38.9%)
Isolated optic neuritis, n (%)	3 (42.9%)	5 (27.8%)
p-value	p = 0.047*	

***Statistically significant (p<0.05) for combined presentation between groups**

Laboratory and Radiological Findings: Cerebrospinal fluid analysis was performed in all 25 patients and revealed variable abnormalities. The median CSF cell count was 54 cells per microliter with a wide range from 0 to 840 cells/μL, reflecting the spectrum of inflammatory activity at the time of lumbar puncture. The median CSF protein level was 58 mg/dL (range: 4-112 mg/dL), indicating mild to moderate elevation in the majority of patients. The presence of pleocytosis (defined as >5 cells/μL) was observed in 18 patients (72%), while protein elevation (>45 mg/dL) was present in 16 patients (64%).

Magnetic resonance imaging of the spinal cord was performed in all patients with clinical evidence of myelitis. Among the 17 patients presenting with myelitis (either isolated or combined with optic neuritis), cervico-dorsal spinal cord involvement was documented in 17 patients, representing 68% of the total cohort. The median length of spinal cord lesions was 4.2 vertebral segments (range: 3-9 segments), consistent with the characteristic longitudinally extensive transverse myelitis

(LETM) pattern of NMO. Central cord involvement with relative sparing of peripheral white matter was the predominant pattern observed on axial MRI sequences. Gadolinium enhancement was present in 14 of 17 patients (82%) with myelitis, indicating active inflammation at the time of imaging.

Table 3: Laboratory and Radiological Findings

Laboratory/Imaging Parameter	Findings
CSF cell count (cells/μL), median (range)	54 (0-840)
CSF protein (mg/dL), median (range)	58 (4-112)
CSF pleocytosis (>5 cells/μL), n (%)	18 (72%)
Elevated CSF protein (>45 mg/dL), n (%)	16 (64%)
Patients with myelitis, n (%)	17 (68%)
Cervico-dorsal cord involvement, n (% of total)	17 (68%)
Lesion length (vertebral segments), median (range)	4.2 (3-9)
Gadolinium enhancement in myelitis, n/N (%)	14/17 (82%)

Treatment Response and Outcomes: All 25 patients received initial treatment with intravenous methylprednisolone 1000 mg daily for 5 days as first-line therapy for acute relapses. The treatment responses are summarized in Table 4. Twelve patients (48%) demonstrated clinical improvement with the initial course of methylprednisolone, defined as improvement in neurological deficits within two weeks of treatment initiation. Ten patients (40%) required a second course of high-dose corticosteroids due to inadequate response or continued progression of symptoms during or immediately after the first course.

For patients who failed to respond to two courses of corticosteroids, escalation therapy was initiated. Two patients (8%) underwent therapeutic plasmapheresis consisting of 5-7 sessions performed on alternate days, with both patients showing meaningful clinical improvement. One patient (4%) was treated with rituximab (two infusions of 1000 mg given two weeks apart) after failing corticosteroids and demonstrated subsequent stabilization and modest improvement in functional status. No significant adverse events were reported with any of the treatment modalities during the acute management phase. Functional outcomes were assessed using the Expanded Disability Status Scale (EDSS) at the last follow-up visit, with a median follow-up duration of 18 months (range: 6-36 months). The median EDSS score at last follow-up for the entire cohort was 4.0 (range: 1.0-8.0). Interestingly, seronegative patients demonstrated a lower median EDSS score of 3.5 compared to 5.0 in seropositive patients. However, this difference did not reach statistical significance (p=0.14), likely due to the small sample size and variability in follow-up duration. Subgroup analysis revealed that patients who responded to initial methylprednisolone therapy had significantly better functional outcomes (median EDSS 2.5) compared to those requiring escalation therapy (median EDSS 5.5, p=0.021).

Table 4: Treatment Response and Functional Outcomes

Treatment Parameter	Number of Patients (%)
Initial IV methylprednisolone	25 (100%)
Response to initial steroids	12 (48%)
Second course of steroids	10 (40%)
Plasmapheresis	2 (8%)
Rituximab	1 (4%)
EDSS at last follow-up (overall), median (range)	4.0 (1.0-8.0)
EDSS in seropositive patients, median	5.0
EDSS in seronegative patients, median	3.5
p-value (seropositive vs seronegative)	p = 0.14

Discussion

This retrospective study provides valuable insights into the clinical profile, treatment response, and outcomes of patients with neuromyelitis optica and NMOSD in South Tamil Nadu, a region with limited published data on these disorders. Our findings contribute to the growing body of literature characterizing NMO in the Indian population and highlight several important observations that warrant detailed discussion.

The demographic characteristics of our cohort align with global trends reported in NMO literature. The female predominance with a ratio of 2.57:1 is consistent with the well-established observation that NMO disproportionately affects women, with female to male ratios typically ranging from 3:1 to 9:1 in various populations.[16] The median age of

onset at 35 years falls within the typical peak incidence period reported in most studies, which ranges from the third to fifth decades of life.[17] However, NMO can occur at any age, and the age range in our study (24-45 years IQR) demonstrates the disease's predilection for young to middle-aged adults, potentially impacting individuals during their most productive years.

A particularly striking finding in our study was the high proportion of seronegative patients (72%) compared to seropositive cases (28%). This distribution differs notably from many Western studies, where seropositivity rates often exceed 70-80% when using sensitive cell-based assays.[18] Several factors may contribute to this observation. First, the timing of antibody testing in relation to disease activity and immunosuppressive treatment may influence detection rates, as antibody titers can fluctuate and may be suppressed during treatment. Second, methodological differences in antibody detection assays could account for some variability. Third, there may be genuine regional or genetic differences in the proportion of antibody-positive versus antibody-negative NMO spectrum disorders. Some Asian studies have reported similar findings of relatively higher seronegative rates, suggesting potential population-specific characteristics.[19] Furthermore, a subset of seronegative NMO patients may harbor antibodies against myelin oligodendrocyte glycoprotein (MOG), which represents a distinct disease entity with different pathophysiology and potentially different outcomes, although MOG antibody testing was not routinely available during our study period.

The significantly higher frequency of combined myelitis and optic neuritis presentation among seronegative patients ($p=0.047$) represents a novel and intriguing finding. Classical teaching suggests that seropositive NMO tends to present with more simultaneous or closely spaced attacks affecting multiple sites, while limited forms are more common in seronegative disease. However, our data suggest a different pattern in this population. This finding warrants validation in larger cohorts but could have important implications for clinical suspicion and early diagnosis. Clinicians evaluating patients with combined myelitis and optic neuritis should maintain high suspicion for NMOSD even in the absence of positive AQP4 antibodies, and comprehensive evaluation including repeat antibody testing and consideration of MOG antibodies should be pursued.[20]

The cerebrospinal fluid findings in our study demonstrate the characteristic but nonspecific inflammatory changes seen in NMO. The median cell count of 54 cells per microliter with a wide range (0-840) reflects the variable inflammatory response, with some patients demonstrating marked pleocytosis while others have relatively normal cell counts. The median protein elevation to 58 mg/dL is consistent with disruption of the blood-brain barrier and ongoing inflammation. These findings align with published literature indicating that CSF pleocytosis and protein elevation are common but not universal in NMO, and their absence does not exclude the diagnosis.[21] The wide variability in CSF parameters underscores the importance of not relying solely on CSF analysis for diagnosis but integrating it with clinical, radiological, and serological data.

The predominance of cervico-dorsal spinal cord involvement (68% of total cohort) is noteworthy and consistent with the typical distribution of spinal lesions in NMO. The cervical and upper thoracic cord regions are particularly vulnerable in NMO, likely reflecting the high density of aquaporin-4 expression in these areas. The characteristic longitudinally extensive lesions extending over a median of 4.2 vertebral segments distinguishes NMO from multiple sclerosis, where spinal lesions are typically shorter and more peripheral in location. The central cord predominance with relative sparing of peripheral white matter observed in our patients is also characteristic of NMO pathology, reflecting the distribution of astrocytic damage emanating from central gray matter and periependymal regions where aquaporin-4 expression is highest.[22]

Treatment outcomes in our cohort demonstrate the challenging nature of managing NMO relapses. While approximately half of patients responded to initial high-dose corticosteroids, a substantial proportion (40%) required additional steroid courses, and 12% needed escalation to plasmapheresis or rituximab. This underscores the importance of having multiple therapeutic options available and being prepared to escalate treatment promptly when initial therapy proves inadequate. The favorable responses observed with plasmapheresis align with evidence supporting its use as rescue therapy for severe or steroid-refractory NMO attacks, where it can remove pathogenic antibodies and inflammatory mediators.[23] The successful use of rituximab in one patient, while representing a small number, reflects the growing recognition of B-cell depletion therapy as both an acute and maintenance treatment option for NMO.

The functional outcomes assessed by EDSS scores reveal substantial residual disability in this cohort, with a median score of 4.0 at last follow-up. This level of disability typically corresponds to significant limitations in walking ability and daily activities, highlighting the severe impact of NMO on patient function. The observation that seronegative patients had numerically lower EDSS scores (median 3.5) compared to seropositive patients (median 5.0), though not statistically significant, is intriguing. Some studies have suggested that seronegative NMO may have a

somewhat better prognosis, possibly due to different underlying pathophysiology or inclusion of MOG-antibody positive cases with generally better outcomes.[24] However, the lack of statistical significance in our study may reflect insufficient statistical power due to small sample size, and caution should be exercised in over-interpreting this trend.

The finding that patients responding to initial methylprednisolone therapy had significantly better functional outcomes (median EDSS 2.5 versus 5.5 in those requiring escalation, p=0.021) has important clinical implications. This suggests that early treatment response may be a prognostic indicator, and patients who fail to respond adequately to initial corticosteroids may be at higher risk for poor long-term outcomes. This observation supports aggressive early treatment and prompt escalation to second-line therapies when initial responses are suboptimal. It also emphasizes the importance of close monitoring during acute attacks to identify non-responders early and modify treatment accordingly.

Our study has several limitations that must be acknowledged. The retrospective design limits the completeness and standardization of data collection, and the relatively small sample size of 25 patients limits statistical power for detecting differences between groups.

The variability in follow-up duration and the lack of standardized assessment timepoints may introduce bias in outcome assessment. The absence of systematic MOG antibody testing means we cannot definitively characterize the seronegative group, which likely represents a heterogeneous population including both true AQP4-negative NMO and MOG-antibody positive disease. Additionally, the study was conducted at a single tertiary center, which may limit generalizability to the broader population and could introduce selection bias toward more severe or complex cases.

Despite these limitations, this study provides valuable real-world data on NMO and NMOSD in South Tamil Nadu and raises several important questions for future research. Larger prospective multicenter studies with systematic antibody testing (including both AQP4-IgG and MOG-IgG), standardized treatment protocols, and longer follow-up periods are needed to better characterize the clinical spectrum, optimal treatment strategies, and long-term outcomes in this population. Genetic studies exploring population-specific risk factors and comparative studies examining differences between Indian and other Asian populations versus Western populations would enhance our understanding of regional variations in NMO.

The emergence of novel targeted therapies for NMO, including complement inhibitors and IL-6 receptor antagonists, offers promise for improved outcomes. However, access to these expensive medications remains a significant challenge in resource-limited settings like South Tamil Nadu. Health policy initiatives to improve access to both diagnostic testing (particularly antibody assays) and therapeutic options are crucial for optimizing care for NMO patients in India. Establishment of NMO registries and collaborative networks could facilitate larger studies and improve understanding of the disease in the Indian context.

Conclusion

This study provides important insights into the clinical characteristics and outcomes of neuromyelitis optica and NMOSD in South Tamil Nadu. Our key findings include a high proportion of seronegative cases, a significantly higher frequency of combined myelitis and optic neuritis presentation among seronegative patients, predominant cervico-dorsal spinal cord involvement, and substantial residual disability despite treatment. While seropositive and seronegative patients demonstrated similar functional outcomes at last follow-up, early treatment response emerged as an important prognostic factor.

These findings highlight the need for maintaining high clinical suspicion for NMO in patients presenting with characteristic clinical features, even in the absence of positive AQP4 antibodies.

The aggressive nature of the disease and the severe disability that can result emphasize the critical importance of early diagnosis, prompt treatment of acute attacks, and initiation of long-term immunosuppression to prevent relapses. The variable treatment responses observed underscore the need for individualized treatment approaches and readiness to escalate therapy when initial treatments prove inadequate.

Future prospective studies with larger sample sizes, comprehensive antibody testing including MOG antibodies, standardized treatment protocols, and extended follow-up periods are essential to better understand the natural history, optimal management strategies, and long-term prognosis of NMO and NMOSD in the Indian population. Such studies will inform evidence-based treatment guidelines adapted to the regional context and may identify population-specific prognostic factors. Ultimately, improved understanding of NMO in diverse populations, coupled with advances in targeted therapies and improved access to diagnostic and therapeutic resources, holds promise for better outcomes for patients suffering from this devastating disorder.

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