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Case Report

Diagnosis of neuromyelitis optica with bilateral vision loss as initial symptom secondary to acute middle encephalitis: a report of three cases

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Abstract: Neuromyelitis optica (NMO) is an inflammatory demyelinating syndrome primarily characterized by optic neuritis and longitudinally extensive myelitis. However, the heterogeneity of neuromyelitis optica spectrum disorders (NMOSD) has expanded beyond the classical definition, revealing increased phenotypic diversity. Around 20-40% of NMOSD patients present with brainstem or hypothalamic involvement as their initial symptom. Case studies describing NMOSD with midbrain syndrome as the primary presentation are rare. This report details three cases where acute bilateral vision loss was the first symptom, followed by midbrain dysfunction, with a final diagnosis of NMOSD confirmed by serum AQP4 antibody testing or characteristic imaging findings.

Keywords: Neuromyelitis optica, bilateral vision loss, acute middle encephalitis, case report

Introduction

Since Devic first described neuromyelitis optica (NMO) in 1894, the disease has been defined as a specific inflammatory demyelinating syndrome involving optic neuritis and longitudinally extensive myelitis [1-3]. Clinical manifestations typically include optic neuritis and myelitis, which often result in poor or no recovery [4]. Traditionally, diagnosis has relied on the combined involvement of the optic nerve and spinal cord, treating other central nervous system lesions as exclusion criteria. However, with the advent of serum AQP4 antibody testing and the accumulation of clinical cases, the phenotypic heterogeneity of neuromyelitis optica spectrum disorders (NMOSD) has expanded beyond the classical framework, revealing a more complex pathological landscape.

Recent prospective cohort studies have shown that 20-40% of NMOSD patients present with brainstem or hypothalamic syndrome as their initial symptoms, a proportion that may be higher in Asian populations [5]. Atypical presentations, including acute dizziness, ophthalmoplegia, somnolence, behavioral abnormalities, and thermoregulatory disorders, are often misdiagnosed as stroke, encephalitis, or tumor-related lesions. The pathogenesis is thought to involve AQP4 antibody-mediated attacks on the astrocytic processes of perivascular organs, which are rich in AQP4 aquaporins and have relatively weak blood-brain barriers, making them prone to characteristic "pancake-like" imaging findings (low signal in the center with surrounding edema) [6, 7]. Some patients exhibit isolated midbrain or brainstem involvement in the early stages of the disease, lacking typical optic nerve or spinal cord lesions. In response, the international NMOSD diagnostic consensus has been updated, incorporating AQP4 antibody positivity or characteristic imaging findings into core diagnostic criteria, enabling earlier identification of atypical cases. Clinically, patients presenting with acute midbrain syndrome should be closely monitored, with timely serological testing and cranial MRI screening to avoid delays in immunomodulatory treatment.

In AQP4 antibody-positive NMOSD patients, optic neuritis or myelitis is more common than

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Table 1. General information and follow up results of the cases

Case	Gender	Age (years old)	Clinical symptoms	Diagnosis	Treatment	Follow up
1	Female	17	one-week history of progressive bilateral vision loss (0.1 in the right eye; 0.5 in the left eye) and dizziness	neuromyelitis optica spectrum disorders	intravenous methylprednisolone (1 g/d, for three consecutive days), followed by oral tapering; Rituximab for maintenance therapy (every six months)	Vision improved to 0.8 (both eyes) within a week, and hypersomnia resolved; no recurrence at six-month
2	Female	58	repeated vision loss and limb numbness and weakness for 7 years, and vision loss for 1 month	mesencephalic syndrome, vitamin B12 deficiency, severe osteoporosis	1 cycle of immunoglobulin IV followed by rituximab every 6 months	a slight improvement in limb numbness but continued vision loss
3	Female	9	vision decline accompanied by somnolence	neuromyelitis optica spectrum disorders	intravenous methylprednisolone (1 g/d for three consecutive days), followed by gradual oral tapering	vision improved to 0.5 (both eyes) within one week, correction without improvement

other symptoms, as demonstrated by a large international cohort study [8]. Due to the historical underrecognition of atypical NMOSD manifestations, such as midbrain or brainstem syndromes, traditional diagnostic paradigms have relied heavily on the combined involvement of the optic nerve and spinal cord as the core diagnostic basis. Large cohort studies have shown that AQP4 antibody positivity can predict atypical NMOSD, even in patients presenting with isolated vertigo, cognitive impairment, or thermoregulatory disorders. In such cases, immunomodulatory therapy should be prioritized over symptomatic treatment [1, 5, 9]. This strategy overcomes the limitations of traditional classification systems. Some centers still classify patients as "brainstem encephalitis" or "myelitis" based solely on clinical presentation, leading to delayed critical treatment.

Additionally, some regions still follow a "two-step" diagnostic approach: initially classifying based on symptom distribution (e.g., midbrain involvement), followed by immunological testing and genetic analysis. However, this approach may overlook overlapping syndromes in patients who are AQP4 antibody-negative but MOG antibody-positive. Few case studies address NMOSD patients presenting primarily with midbrain syndrome. Thus, this paper not only provides a more detailed description of the clinical features and diagnostic process for midbrain syndrome but also identifies it as one of the six core clinical symptoms of NMOSD. Furthermore, we analyze the diagnosis and treatment process for this group of patients and offer additional treatment options for reference.

Here, we report three cases of acute bilateral vision loss as initial symptoms, followed by midbrain dysfunction, and ultimately diagnosed as NMOSD through serum AQP4 antibody testing or characteristic imaging findings (**Table 1**). Our focus is to summarize the clinical phenotype of this condition. This study was approved by the ethics committee of each participating center, and all patients involved in the study provided informed consent.

Case report

Case 1

A 17-year-old female patient presented to the ophthalmology department at Children's Hospital of Fudan University (Xiamen Hospital) with a one-week history of progressive bilateral vision loss (0.1 in the right eye, 0.5 in the left eye) and dizziness. She denied any pain, fever, or exposure to toxins. Initial ophthalmic evaluation revealed a flattened fundus retina, pale optic disc with clear margins, and normal visual evoked potentials (VEPs), suggesting the need for neurological consultation. The patient was admitted in August 2023. On August 25, cranial MRI with T2-weighted and FLAIR sequences showed bilateral optic tract damage, symmetrical hyperintense lesions around the third ventricle, and "pancake-like" lesions in the pineal region (central T1 low signal, peripherally FLAIR high signal) (**Figure 1A**, **1B**). Further tests were conducted, with blood routine indicators within normal limits. The total bilirubin level was 2.6 μmol/L (5.1-17.1 μmol/L); triglycerides were 2.28 mmol/L (0.56-1.7 mmol/L); high-density lipoprotein was 0.91 mmol/L (1.1-1.8 mmol/L); low-density lipopro-

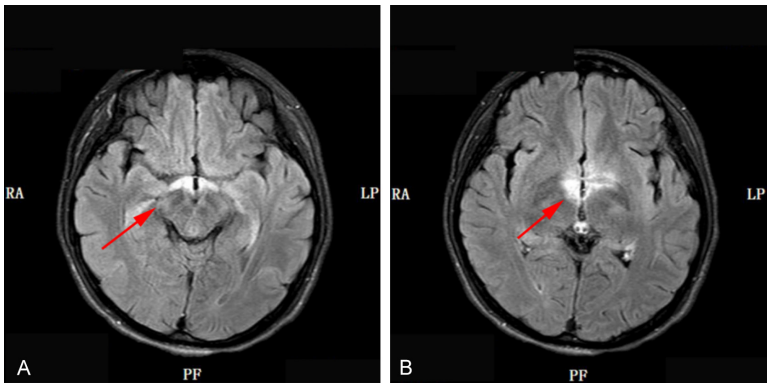

Figure 1. Typical MRI image cases. A, B: T2-weighted and FLAIR sequences of the brain MRI show bilateral optic tract damage with symmetrical hyperintense lesions around the third ventricle. Red arrows indicated hyperintense lesion area.

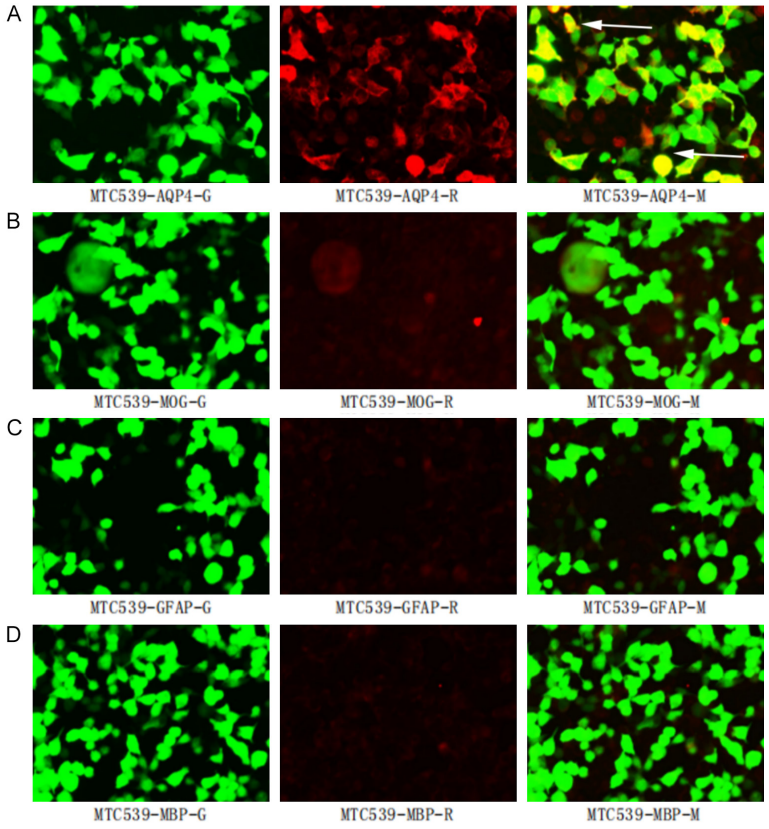

Figure 2. Protein expression detection using CBA method. A: Serum AQP4 antibody (+); B: Serum MOG antibody (-); C: Anti-glial fibrillary acidic protein (GFAP) antibody (-); D: Anti-myelin basic protein antibody (-). White arrows indicated positive results.

tein was 1.58 mmol/L (2.07-3.1 mmol/L); and the erythrocyte sedimentation rate was 5 mm/hr (0-15 mm/hr). Cerebrospinal fluid (CSF) analysis showed mild pleocytosis. Serum testing was negative for MOG antibodies, GFAP antibodies, and anti-myelin basic protein antibodies, but positive for AQP4 antibodies on cellular basis testing (CBA) (**Figure 2**), confirming the diagnosis of NMOSD. The patient was treated with intravenous methylprednisolone (1 g/d for three consecutive days), followed by oral tapering. Vision improved to 0.8 (both eyes) within a week, and hypersomnia resolved. Rituximab was initiated for maintenance therapy (administered every six months), with no recurrence at the six-month follow-up.

Case 2

A 58-year-old female patient presented with a 7-year history of recurrent vision loss, limb numbness, and weakness, with recent vision loss lasting for one month. The patient had a history of lumbar compression fractures. In 2017, she experienced neck and lower back pain, followed by bilateral vision loss. Upon presentation at another hospital, CSF testing revealed positive AQP4 antibodies, and the patient was diagnosed with NMOSD. She received steroid pulse therapy and intravenous immunoglobulin infusion, which improved her vision. After discharge, the patient continued oral steroid therapy. Despite recurrent episodes of vision loss and gait instability, the condition continued to progress. During her most recent hospitalization in March 2023, the patient experienced further vision loss, inability to walk, and incoherent speech. Imaging, including visual MRI, susceptibility-weighted imaging, and spinal MRIs, revealed multiple intracranial lesions and spinal cord lesions from T3-T8 lev-

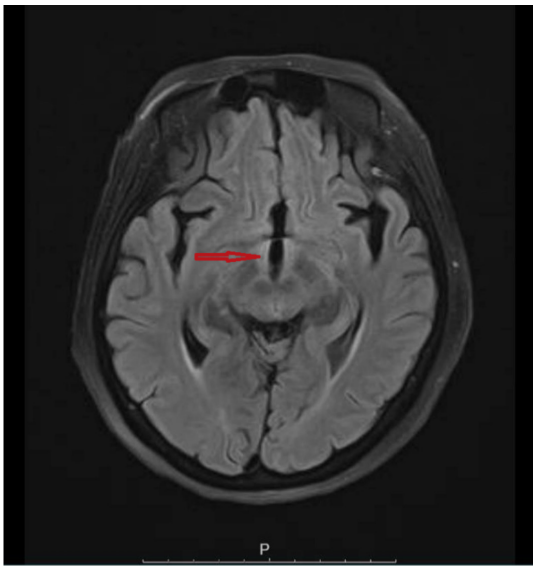

Figure 3. Typical case of thalamus and aqueduct lesion. T2-weighted and FLAIR sequences of brain MRI showed multiple lesions in the thalamus and aqueduct; Red arrows indicated hyperintense lesion area.

els, along with compression fractures at T8 and L5 vertebrae, mild disc protrusions at L3-4, L4-5, and L5-S1, and degenerative changes in the thoracolumbar spine. A diagnosis of mesencephalic syndrome, vitamin B12 deficiency, and severe osteoporosis was made. Blood tests showed normal results. Total protein was 56.77 g/L (↓), alanine aminotransferase was 12.5 U/L (↓), aspartate aminotransferase and was 13.5 U/L, and estimated glomerular filtration rate was 108.04 mL/min. CSF analysis revealed a white blood cell count of 0.6×10^6/L and total protein of 389.20 mg/L, with glucose at 3.96 mmol/L (↑), chloride at 129.00 mmol/L, lactate dehydrogenase at 18.5 U/L, and adenosine deaminase at 1.4 U/L. Serum testing showed positive AQP4 antibodies and negative MOG antibodies, confirming NMOSD. The patient was treated with one cycle of intravenous immunoglobulin followed by rituximab every six months. Limb numbness improved slightly, but vision loss continued. The latest follow-up (January 2025) included cervical spinal cord and cranial MRIs, which showed stable lesions at the C2-T6 level and intracranial lesions similar to previous findings. Abnormal signals in the cervical spinal cord raised concern for subacute combined degeneration (SCD). Cervical spondylosis was also noted at C3-4, C4-5, C5-6, and C6-7.

Case 3

A 9-year-old female patient presented with sudden vision decline and somnolence for one week, and was admitted in April 2025. On April 23, MRI revealed symmetrical hyperintense lesions around the third ventricle (**Figure 3**). Blood routine tests were normal. Total protein was 69.6 g/L, alanine aminotransferase was 10.6 U/L, and aspartate aminotransferase was 23.4 U/L. Serum testing was positive for both MOG and AQP4 antibodies. The patient received intravenous methylprednisolone (1 g/d for three consecutive days), followed by gradual oral tapering. Vision improved to 0.5 (both eyes) within one week, although correction was not achieved. Follow-up after two months showed stable results.

Discussion

NMO is a rare disease that predominantly affects adults, especially females, with a sex ratio of up to 9:14 [10]. Approximately 5% to 10% of NMOSD cases begin before the age of eighteen [11]. Adolescent patients are more prone to brain lesions (such as involvement of the diencephalon and brainstem) and hyponatremia, with a higher recurrence rate and greater risk of disability. Adolescent NMOSD often presents with atypical symptoms, such as persistent nausea and vomiting (extreme posterior syndrome), somnolence, or brainstem dysfunction, which can be misdiagnosed as gastrointestinal disorders or psychiatric conditions. MRI typically shows a higher proportion of long-segment lesions around the third ventricle, medulla, or spinal cord, with AQP4 antibody positivity comparable to that in adults (approximately 80%) [12, 13]. Younger age at onset in adolescents correlates with a higher risk of cognitive impairment, with only 17% responding to hormone therapy, significantly lower than that in adults. Traditional immunosuppressants (such as mycophenolate mofetil and azathioprine) have a high failure rate (59% to 75%), making early initiation of targeted biologics (such as siltuximab) crucial to reduce the risk of recurrence. A multicenter retrospective analysis of childhood-onset NMOSD (n=67 cases, age<18 years) over four years found that 86.6% of patients experienced relapse during follow-up, 47.8% developed visual impairment, and 25.4% suffered cognitive impairment.

Among those not treated with biologics, 40% had an EDSS score ≥7 (severe disability) within six years of onset. After switching to siltuximab, the annual relapse rate (ARR) decreased from 1.5 to 0.08, and immune function indicators (such as B cell ratio and AQP4 antibody titers) significantly improved [14-16].

The 2021 edition of the Chinese NMOSD Diagnosis and Treatment Guidelines recommends biological agents (e.g., sartilizumab) for adolescent patients, as these agents have demonstrated efficacy and safety in patients over 12 years of age. Due to the unique nature of adolescent NMOSD, early and precise intervention is critical [17-19]. Adolescents are more prone to Th17/Treg imbalance and overactivation of the IL-6 signaling pathway, suggesting that targeting IL-6R (e.g., sintilimab) may achieve long-term remission by modulating immune homeostasis. However, large-scale epidemiological studies on NMOSD in adolescents remain limited, and more prospective cohort data are needed to optimize treatment strategies.

A systematic search of the PubMed database (keywords: "NMOSD AND diencephalon/thalamus") identified 17 case reports in the past five years. All patients were positive for AQP4 antibodies, and MRI prominently showed the "pancake sign" in the midbrain area (low signal at the lesion center with surrounding edema), with 76% of cases also involving the spinal cord or optic nerve [14, 20]. Notably, the patients in these cases were all young, suggesting that NMOSD in children may have a distinct clinical phenotype. The mechanism of midbrain involvement in NMOSD has not been fully elucidated, but it may be related to AQP4 antibodies disrupting the blood-brain barrier and triggering local inflammatory responses. The high expression of AQP4 in structures surrounding the third ventricle (such as the pineal gland and hypothalamus) makes them potential targets for attack. Clinicians should be vigilant in patients presenting with acute vision loss accompanied by midbrain symptoms (such as sleep cycle disturbances and abnormal body temperature regulation), promptly conducting AQP4 antibody and MRI examinations to avoid misdiagnosis.

This study illustrates the diversity of atypical phenotypes of NMOSD: 1. Diverse clinical manifestations: NMOSD patients may present with atypical symptoms, such as decreased vision, dizziness, and somnolence, rather than typical optic neuritis or myelitis.

2. Diverse MRI features: MRI manifestations of NMOSD may involve multiple regions, including the hypothalamus, midbrain, and spinal cord, with varying lesion morphology and signal characteristics.

3. Diverse serological tests: Although AQP4 antibody positivity is a critical diagnostic criterion, some patients may have other autoimmune diseases or antibodies, such as MOG antibody positivity.

These cases expand the phenotypic spectrum of NMOSD, emphasizing that hypothalamic syndrome can be an initial manifestation, particularly in pediatric patients. Future studies should aim to establish early screening procedures for atypical NMOSD and explore the value of immunoadsorption therapy in improving long-term outcomes.

This study highlights that NMOSD can present as atypical midbrain involvement and isolated visual impairment. Key diagnostic clues include periventricular MRI lesions and AQP4-IgG seropositivity. Normal VEPs may reflect transient functional deficits or detection limitations in uncooperative patients. Early steroid therapy and immunosuppression are essential to prevent relapse. However, this case report has limitations due to the low level of evidence in the literature, as there are few similar cases for reference. A cohort of 16 NMOSD cases from Senegal [21] showed that the average patient age was 30 years, with 10 cases presenting the optic-spinal variant, and the remaining cases were isolated transverse myelitis. Ten cases tested positive for AQP4 antibodies. After three months of systemic corticosteroids and azathioprine treatment, clinical outcomes were favorable. A report from East Africa [22] documented 11 NMOSD cases with an average onset age of 30 years. Eight patients had the optic-spinal variant, while the rest presented with acute brainstem syndrome. Seven cases were AQP4 antibody-positive. All patients received systemic corticosteroids and azathioprine/mycophenolate mofetil therapy. Four patients required plasma exchange, and all had few relapses and disabilities at the end of follow-up.

In our cases, patients presented with atypical midbrain involvement and isolated visual im-

pairment. The effects of steroid therapy and immunosuppression vary among individuals and may depend on disease duration. Older patients diagnosed early but untreated may experience limited treatment efficacy upon recurrence. The study lacks long-term follow-up data after treatment, relying primarily on clinical manifestations. The last patient was diagnosed shortly before treatment, requiring further investigation.

Conclusion

For patients with unexplained visual impairment, even in the absence of typical optic neuritis, acute midbrain syndrome should be considered. This atypical NMOSD is often misdiagnosed as encephalitis, multiple sclerosis, or paraneoplastic syndromes, emphasizing the importance of clinical vigilance. Combined AQP4/MOG/GFAP antibody testing and cranial MRI can improve early diagnosis rates. Aggressive immunotherapy can improve the prognosis of NMOSD patients.

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Disclosure of conflict of interest

None.

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