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AN INTERESTING CASE OF LOSS OF VISION

Neurology

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ABSTRACT

Neuromyelitis optica is an aggressive inflammatory disorder characterized by recurrent attacks of optic neuritis and myelitis. It is more frequent in women than men, and typically begins in adulthood, but can arise at any age. We are presenting an interesting case of a 38 year old female who presented with optic neuritis and transverse myelitis with renal failure.

KEYWORDS

Neuromyelitis Optica Transverse Myelitis Optic Neuritis Systemic Lupus Erythematosus Lupus Nephritis

INTRODUCTION

Neuromyelitis optica (NMO) was described by Eugène Devic in 1894 and it is known since then as Devic's NMO (Devic's disease). It is an inflammatory demyelinating neuro-immunological disease of the central nervous system (CNS) that can occur idiopathically or in conjunction with other systemic diseases.

NMO diagnostic criteria are characterized by sequential or concomitant attacks of transverse myelitis and optic neuritis, with contiguous spinal cord MRI lesion extending over three or more vertebral segments, and seropositivity for NMO-IgG (anti-aquaporin-4 (anti-AQP4)), which has been recently described as a sensitive and specific marker for NMO.

NMO affects both genders with three to nine times more prevalent in women than in men.

Up to 40% of NMO patients have a systemic autoimmune disorder, such as systemic lupus erythematosus, Sjögren's syndrome, p-ANCA–associated vasculitis, myasthenia gravis, Hashimoto's thyroiditis, or mixed connective tissue disease.

CASE REPORT

A 38 year old female patient presented to the emergency with chief compliants of sudden , progressive loss of vision with pain on movement of the eyes since 1 day. She also complained of mild headache and paresthesias of both lower limbs. Her past history was not significant for any similar episodes.

Imaging (CT and MRI brain) showed no abnormalities.

Patient was planned to give pulse dose of intravenous methyl prednisolone for 3 days.

BUT! One day after hospital admission, patient developed paraplegia which is sudden in onset , gradually progressive with urinary incontinence . She also complained of a tight band like sensation at the umbilicus level which later was felt at subcostal region.

Power in both lower limbs -1/5 compared to upper limbs -4/5
Tone in both lower limbs – hypotonia
Areflexia in both lower limbs.
No objective sensory loss in lower limbs.
Cerebellar , cerebral functions are normal.

MRI Spine revealed intramedullary MRI lesion extending ≥3 contiguous segments (LETM) and axial T2-weighted MRI demonstrated T2 hyperintense involvement of the optic tracts.

Serum Aquaporin-4 antibodies turned out to be positive.
CSF analysis was normal except for mild neutrophilic pleocytosis .

Patient also had abnormal renal function tests and Urine routine examination which revealed elevated serum creatinine (2.8) and blood urea (58) , with 3+ proteinuria on URE.

Ultrasound Abdomen revealed no abnormalities.
Antinuclear antibodies – positive
Anti-double-stranded DNA (anti-dsDNA)- positive
Anticardiolipin antibodies – positive.

Renal biopsy revealed features of rapidly progressive glomerulonephritis. (Lupus nephritis stage 3)

Patient was found to have systemic lupus erythematosus(SLE) With lupus nephritis with autoimmunity triggered NEUROMYELITIS OPTICA.

Patient was continued on intravenous pulse methyl prednisolone therapy for 10 days. Patient showed good response to immunotherapy and planned to maintain the patient on tapering steroid dose.

Nephrologist and rheumatologist consultation was taken and patient was continued on immunosuppressive therapy and advised follow up.

DISCUSSION

SLE is a multisystem autoimmune disorder and its pathophysiology may involve all components of the CNS. The CNS and peripheral nervous systems (PNS) may be involved in SLE. SLE may present both steadily chronic and more episodic neurologic symptoms throughout the life span.

The main pathological findings in patients with SLE are those of inflammation, vasculitis, immune complex deposition, and vasculopathy.

NMO may present a different spectrum of symptoms; generally, it is presented as an acute disease, with an onset in 30–50% of the cases preceded by a virus-like syndrome, with headache, sore throat, fever, and malaise; ascending myelitis resulting in pain, which may be severe; numbness; weakness or acute respiratory and gastrointestinal symptoms. Sometimes patients may experience vision impairment, hearing impairment, olfactory dysfunction, pain, and cognitive dysfunction30–32 and various degrees of paralysis, as well as incontinence.

NMO may co-occur with SLE or other autoimmune diseases such as acute demyelinating encephalomyelitis (ADEM) and Behcet's disease. The first-line therapy with azathioprine or rituximab for severe disease course of NMO calls for prompt initiation of immunosuppressive treatment once the diagnosis of AQP4-Ab-positive NMOSD has been confirmed.

IVIg can be used as the first-line therapy for children or for patients with contraindication to immunosuppressive therapies.

Second-line therapy of NMO: In the case of side effects or poor response, treatment can be switched from azathioprine to rituximab or vice versa, or to mycophenolate mofetil, methotrexate, or mitoxantrone.

Third-line therapy for NMO should be applied if disease progression occurs, and if the above treatments fail, the newer agents such as tocilizumab should be given with combination therapy , combination of immunosuppression plus intermittent plasma exchange (PE); or combination of rituximab with methotrexate or intravenous immunoglobulins.

The mainstay of treatment for any inflammatory life-threatening or organ-threatening manifestations of SLE is systemic glucocorticoids (0.5–1 mg/kg per day PO or 500–1000 mg of methylprednisolone sodium succinate IV daily for 3 days followed by 0.5–1 mg/kg of daily prednisone or equivalent). Survival was significantly better in people with DPGN treated with high-dose daily glucocorticoids (40–60 mg of prednisone daily for 4–6 months) versus lower doses.

Either cyclophosphamide or mycophenolate mofetil is an acceptable choice for induction of improvement in severely ill patients;

In patients whose renal biopsies show ISN grade III or IV disease, early treatment with combinations of glucocorticoids and cyclophosphamide reduces progression to ESRD and death.

CONCLUSION

NMO is an aggressive autoimmune disorder which may be idiopathic or associated with systemic autoimmune disorders , Neoplasms , systemic infections.

Prompt diagnosis and treatment of underlying systemic autoimmune disorders may ameliorate the symptoms and improve quality of life of the patient by decreasing disability and morbidity/ mortality.

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CONFLICT OF INTEREST- NIL
CONSENT – Consent was taken

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