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A case report: Acute neuromyelitis in pregnancy

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Abstract

Neuromyelitis optica spectrum disorder (NMOSD) is a demyelinating disease of the central nervous system. NMOSD is characterized by severe relapse of optic neuritis, longitudinal extensive myelitis and the presence of anti aquaporin-4 IgG (AQP4-IgG) in serum. Female factors like genetic, epi-genetic, or hormonal, may play an important role in pathogenesis. Female gonadal hormones, estrogen and progesterone, rise significantly during pregnancy, and decrease during the postpartum period. We are reporting a case of Gravida 2 Para 1 Living 1 Abortion 0 with full term with known case of acute neuromyelitis (on treatment for 3 years). Patient delivered vaginally, post natal period was uneventful.

Keywords: Neuromyelitis optica spectrum disorder (NMOSD), pregnancy, hormones, aquaporin-4 IgG (AQP4-IgG), Devic's syndrome

Introduction

Neuromyelitis optica (NMO), also known as Devic's syndrome/disease is an autoimmune syndrome largely characterised by optic neuritis and myelitis with poor recovery and a progressive course^[1]. NMOSD is approximately two to nine times more common in women as in men and mainly develops during reproductive age^[2]. Diagnosis and management will be challenging in pregnancy with NMO. Patient may present with increased chances of pregnancy loss, eclampsia, PIH, IUGR, IUD. NMO is an inflammatory diseases of the CNS resulting in astrocyte dysfunction and secondary demyelination. A unique circulating immunoglobulin G(I_gG) autoantibody targeting the astrocyte water channel protein aquaporin-4(AQP4) is an important biomarker of the disease and plays a key role in NMO pathogenesis^[3].

Case Report

A 33 year old female gravida 2, para 1, living 1, with term gestation with previous normal delivery presented to our hospital with diagnosed NMO since 3 years on treatment with tab Azathioprine 50mg bd. She had bilateral extensive myelitis and diagnosed by laboratory investigation being I_gG positive and she was on tab Azathioprine 50mg BD continued throughout antenatal period and her antenatal check ups are normal and uneventful. She delivered a single live male baby of weight 3.0 kg vaginally. Her post partum period was uneventful and is advised to continue tab Azathioprine 50mg BD and as per paediatrician's advice neonate's CBC was monitored. Patient was discharged on 5th postnatal day.

Discussion

Neuromyelitis optica is an immune mediated chronic inflammatory diseases of the CNS^[4, 5, 6]. Clinically it presents with optic neuritis (ON) and myelitis, often characterised by increased chances of pregnancy loss, eclampsia, IUGR, PIH, IUD. Posited mechanisms include antibody-mediated placental damage and a heightened risk of eclampsia-associated PRES. Differential diagnosis include Multiple Sclerosis, Myasthenia Gravis, SLE, Sjogrens Syndrome, Celiac disease and Sarcoidosis^[7]. Histopathologically, NMO is characterised by astrocyte damage, demyelination, neuronal loss, and often pronounced necrosis and the presence of specific autoantibodies (Aquaporin 4 antibodies AQP4-Ab; also termed NMO-IgG) in the serum^[8]. Diagnostics criteria proposed by Wingerchuk *et al.* in 2006 include (a) Contiguous spinal cord MRI lesion extending over three or more vertebral segments. (b) Brain MRI not meeting Patys diagnostic criteria for MS at disease onset. (c) NMO IgG seropositive status^[9]. Clinical evaluation when NMO is suspected includes a detailed medical history and physical examination. Special attention should be paid to brainstem symptoms, neuropathic pain and painful tonic spasm.

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Laboratory investigations include Blood count, coagulation profile, Serum chemistry, blood sedimentation, blood glucose, Vitamin B12, Folic acid antibodies, associated with connective disorders (ANA, anti-ds-DNA antibodies, lupus anticoagulant, antiphospholipid antibodies, ANCA etc. Urine analysis and sediment, Treponema Pallidum haemagglutination assay. Detection of AQP4 ab are detected in 60-90% of patients. The specificity of these assays varies between ~90 and 100%. Others include CSF analysis, Electrophysiology, MRI, Optic Coherence Tomography (OCT). The main treatment goals are (1) Remission and improvement of relapse-associated symptoms. (2) Long-term stabilization of disease course by means of relapse prevention. (3) Symptomatic therapy of residual symptoms.

Early initiation of long term immunosuppressive therapy to delay a second relapse is recommended. Treatment of acute attacks on oral steroid tapering should be considered and Therapeutic plasma exchange can be performed. Data on the long term treatment of NMO (>5YEARS) concern mainly Azathioprine (AZA) and Rituximab (RX).

Azathioprine: Several studies review patient with NMO /NMOSD shown Azathioprine to reduce relapse rate and ameliorate neurological disability in NMO [11]. A dosage regimen of 2.5-3mg/kg body weight/day orally with monitoring of hematologic parameters and liver enzymes is recommended. As the treatment may only take full effect after 3-6months, it should initially combine with oral steroids therapy (1mg/kg body weight/day), as oral steroids shown to suppress disease activity

in NMO. Common side effects include black tarry stools or blood in urine or stools, cough or hoarseness, dizziness, fever or chills, nausea and vomiting painful or difficult urination, pinpoint red spots on skin, new marks or change in marks on your skin.

Rituximab: Rituximab is drug of choice for patients who have not responded to Azathioprine. B cell depletion with Rituximab has been demonstrated as effective in the treatment of NMO [12]. Rituximab treatment can be initiated using one of two regimens: either two 1gm infusions of RX at an interval of 2 weeks or four weekly 375mg/m² body surface area applications. To prevent infusions related side effects, premedication (1gm paracetamol, 100mg prednisolone, 4mg dimethindene maleate i.v) should be dispensed. Common side effects include: headache, fever, chills, stomach pain, nausea, diarrhea, heartburn, night sweats, flushing.

Other drugs include which can be used in treatment of NMO are Mycophenolatemofetil, Immunoglobulins Mitoxantrone, Cyclophosphamide, methotrexate, Interferon-beta/glatiramer acetate, natalizumab, Fingolimod, Anti-IL-6 Therapy.

Conclusion

Vigilant antepartum monitoring is necessary with active disease, it includes education about the early signs and symptoms of preeclampsia eclampsia and postpartum relapse. It is better to deliver the patient in an institution with a team of doctors available which include gynaecologist, anaesthetist, neurologist & paediatrician.



Fig 1: Bilateral swollen optic nerves with a T2 hyperintensity and contrast enhancement

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