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RESEARCH ARTICLE

ACUTE NEUROMYELITIS IN PREGNANCY: CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Neuromyelitis Optica spectrum disorders (NMOSD) are a group of rare autoimmune diseases affecting mainly the central nervous system. The optic nerve and spinal cord are commonly involved in the disease process. Prevalence is higher in females of reproductive age group than males at a ratio of 3–9:1. [1-5] Diagnosis and management of NMOSD cases with pregnancy is challenging and needs a multidisciplinary approach for management. We are presenting the case of a primigravida with sudden onset of weakness of both lower limbs at 20 weeks of gestation, diagnosed as a case of Neuromyelitis Optica Spectrum Disorder; in her third trimester, pregnancy was complicated with oligohydramnios and fetal growth restriction.

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INTRODUCTION

Neuromyelitis Optica (NMO) is an autoimmune demyelinating disease mainly affecting the optic nerve and spinal cord and associated with poor recovery, progressive disease course, and relapses. In cases of pregnancy with NMOS disease course is not the same as in nonpregnant females and has an increased risk of pregnancy loss and various maternal and fetal complications. The disease is characterized by the formation of autoantibodies targeting astrocyte water channel protein aquaporin-4 (AQP4) which is an important biomarker of the disease. (6) These autoantibodies damage astrocytes and cause secondary demyelination which leads to astrocyte dysfunction.

CASE PRESENTATION

We are presenting a case of a 24-year-old female of Asian origin belonging to the Indian subcontinent. She was primigravida at 20 weeks period of gestation and presented to hospital emergency with complaints of sudden onset weakness and inability to move both the lower limbs with loss of sensation for 2-3 days.

The patient had a history of weakness in bilateral upper limbs 3 days back which recovered spontaneously. The symptom was associated with urinary and bowel incontinence. No history of breathing difficulties, fever, loose stools, vomiting, or blurring of vision. She can appreciate sensations below the level of the umbilicus. The patient was evaluated by the general medicine, neurology and obstetric team. On admission, the patient was conscious and oriented, GCS 15/15, was vitally stable, SPO2 99% on room air, power in bilateral lower limbs was 0/5, deep tendon reflex absent, sensory loss present, power in bilateral upper limb 5/5, DTR 2+, no sensory loss. Her uterus was 20 weeks in size, relaxed, and FHS was localized by USG. The Patient was admitted under neurology for further workup and management. MRI whole spine was done that showed: a longitudinally extensive heterogeneous patchy segmented T2 hyperintense signal noted involving predominantly dorsal and central and dorsal aspects of the spinal cord extending from the cervical-medullary junction to the D11 vertebra. Mild expansion of the cord is noted in the cervical region from the C2-C7 level. Mild T2 hyperintense signals are also noted in the lower aspect of the medulla.



Fig. 1. Pressure ulcer at gluteal region of 15 cm * 11 cm * 6cm, grade 4 with skin discoloration

Anterior subarachnoid space was prominent at the lumbar level, however no evidence of any clumping of nerves was noted. Features suggestive of non-compressive myelopathy. Longitudinally extensive transverse myelitis. The patient was eventually started on Injection of Methylprednisolone 1gm OD for 5 days after explaining the due risks to the fetus. There was a gradual improvement in the power of both lower limbs of 1/5 and DTR 1+. Anti NMO (neuromyelitis optica) antibody/AQP4 that was positive with titers of 1:10 and was diagnosed as a case of neuromyelitis Optica spectrum disorder. The patient was eventually shifted to oral steroids and discharged from the hospital after explaining rehabilitation measures. The patient was lost to follow-up and reported to the hospital at 34 weeks with an outside ultrasound report suggestive of oligohydramnios, fetal growth restriction, and breech presentation of the fetus. At the time of admission, the power in bilateral lower limbs was 1/5, and also notice was made of a large decubitus ulcer at the sacral region of size 8x8 cm with the exposed tail of the sacrum (grade 4), the base of the ulcer was covered with slough. Daily dressing of the bedsore with pregnancy care and fetal monitoring were done. The patient underwent elective cesarean section in view of breech with FGR with oligohydramnios at 36 weeks and delivered a live male baby of weight 1.72 kg. The baby needed NICU care for low birth weight. Adequate post-op care was given. The patient was discharged on post-op day 16 after suture removal on oral methylprednisolone with follow-up advice.

DISCUSSION

During pregnancy, there is an increase in the formation of antibody-producing B cells and Th2-mediated immune response which, may be responsible for increased disease activity during pregnancy and more severe disability in the postpartum period.⁽⁷⁾ According to some studies, these AQP4 antigens are also expressed by placenta, which leads to autoantibody-mediated damage of the placenta and subsequently placental ischemia, infarction, and placental dysfunction. It possibly explains the association of these cases with hypertensive disorder of pregnancy, eclampsia, oligohydramnios, FGR, and IUD. So, increased risk of maternal and fetal complications should be explained to patients.

Multidisciplinary approaches are required for optimized and personalized medical care in NMOSD patients with pregnancy. Intravenous or oral methylprednisolone, plasma exchange, or immunoadsorption are various treatment options available during pregnancy. Azathioprine (AZA) and rituximab (RTX) are other treatment options for non-pregnant NMOSD patients. (8,9,10)

Rohan D Souza et al, in a systematic review of 22 case reports and seven case series in pregnant patients with NMOSD noted a considerable increase in NMOSD-related disability and relapses during pregnancy and especially in the immediate postpartum period, more in those not on immunosuppressive treatment at the time of conception. Of the postpartum relapses described in the literature, most occurred within the first 3 months postpartum. The most commonly reported maternal neurologic signs and symptoms reported during pregnancy included sensory abnormalities, motor weakness, visual symptoms, bladder and/or bowel incontinence, and spasticity. Hypertensive disorders of pregnancy including preeclampsia were the most common obstetric complication. There were three reported cases of fetal growth restriction. Most (60%) of them had vaginal births. Initial attacks and relapses of NMOSD were successfully managed with oral or intravenous corticosteroids and immunosuppressants, and refractory cases with immunoglobulin, plasma exchange, and immunoadsorption. Neuropathic pain was most commonly managed with gabapentin, amitriptyline and clonazepam, and painful spasticity with baclofen (11). Wuebbolt et al in their 13 case series study of NMOSD cases most cases, neurologic symptoms either worsened or developed for the first time during pregnancy or the postpartum period, and often responded to a combination of steroids, immunosuppressant medications, plasma exchange, and intravenous immunoglobulin. The 13 pregnancies resulted in two miscarriages, three preterm and eight-term births (12)

CONCLUSION

Pregnancy with NMOSD cases needs a multidisciplinary approach for diagnosis and management. Patients should be educated regarding the increased risk of maternal and fetal complications. Immunosuppressants should be started to mitigate the risk of relapse both during pregnancy and the postpartum period.

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