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CASE REPORT

First reported case of Acute Disseminated Encephalomyelitis in pregnancy in Sri Lanka

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Abstract

Acute Disseminated Encephalomyelitis (ADEM) complicating pregnancy is a rare occurrence, posing challenges to the management due to maternal and fetal risks including fetal loss, fetal anomalies, intrauterine fetal growth restriction (FGR) and preterm labour. We report a case of a 21-year-old primiparous woman who was diagnosed with ADEM during the second trimester. She was treated with high dose intravenous (IV) steroids as well as IV immunoglobulin. She made complete neurological recovery within two months, but there were fetal complications including oligohydramnios, intrauterine growth restriction and low birth weight. The baby was delivered via elective caesarean section due to suspicious cardiotocograph and abnormal fetal doppler studies indicative of compromised fetal circulation. The baby was given special baby care unit (SBCU) care. However, both the mother and baby were safely discharged following six days of special neonatal care. This is the first reported case of ADEM in pregnancy in Sri Lanka, and highlights the challenges in management.

KEYWORDS

Acute disseminated encephalomyelitis (ADEM), pregnancy, IV Immunoglobulin, IV corticosteroids, fetal risk

INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is an autoimmune inflammatory demyelinating disease of the central nervous system (CNS). It manifests as an acute-onset rapidly progressive encephalopathy with multifocal neurologic deficits. Common identified triggers are viral infections and immunization. ADEM is uncommon in adults and occurs more frequently in children. However, adult-onset ADEM, appears to have similar outcome and a typically favourable prognosis.[1] It is a monophasic illness, which may rarely have relapses.[2]

Earliest manifestations of ADEM include sudden onset encephalopathy with unexplained behavioral changes and fever. Descending white matter motor tracts, optic nerves, and spinal cord are commonly involved while focal or generalized seizures also can occur in some cases. Complex immune changes in pregnancy may affect the progression and prognosis of the disease.[3] Only few cases of ADEM complicating pregnancy have been reported so far, with no previously published cases in Sri Lanka. This case report highlights the caveats of management of ADEM in pregnancy.

CASE REPORT

A 21- year- old previously well primiparous woman who was at a period of gestation of 19 weeks presented with speech difficulty and right sided face, arm, and leg weakness for one day. There was no preceding headache, visual disturbances, fever, or head injury. She had no history of seizures, or symptoms of systemic infection. She had been immunized with tetanus toxoid in pregnancy one month back.

On admission her Glasgow Coma Scale was 10/15 (E4M5V1). She was globally aphasic, afebrile and there was no neck stiffness. Pupils were 2 mm bilaterally, equally reacting to light.

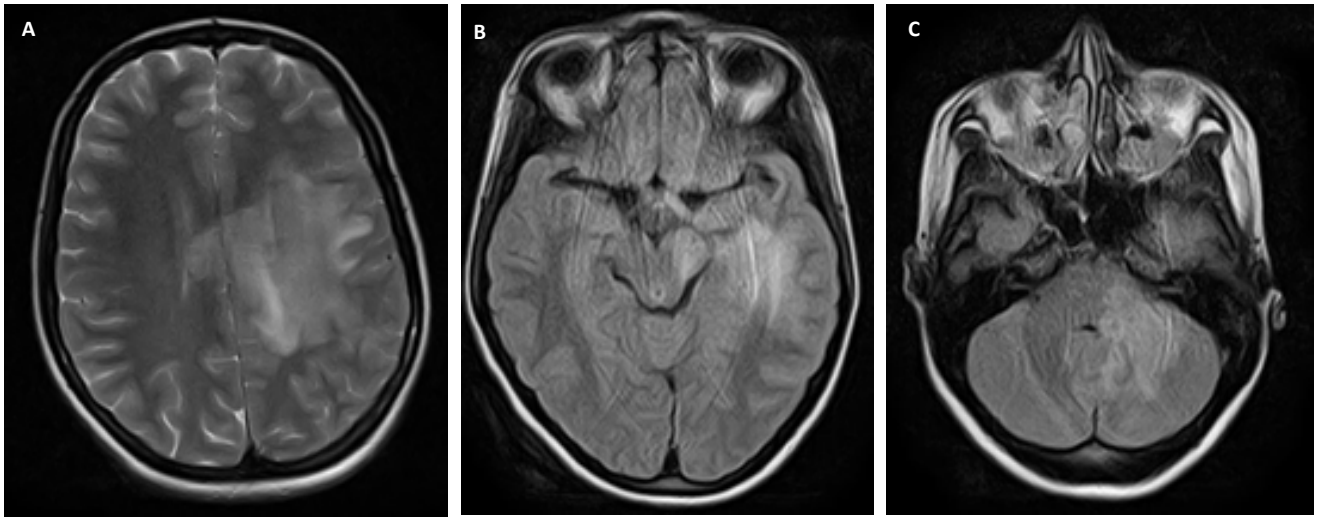

FIGURE 1 **Pre-treatment MRI images: A: T2W image of subcortical hyperintensities, B and C: FLAIR images of brainstem and cerebellum**

She did not have gaze palsy. There was right sided upper motor neuron type facial palsy but no other cranial nerve palsy. A right sided hemiparesis was seen with diminished deep tendon reflexes. Her vital parameters were normal.

The initial MRI brain scan, done prior to commencement of treatment revealed hyperintense lesions involving the left side cerebral subcortical and deep white matter extending to the left side midbrain and cerebellum crossing the midline (Figure 1 A,B,C). There were no areas of diffusion restriction or microhaemorrhage.

The basic hematological and biochemistry investigations were normal, while microbiological cultures revealed no growth. Cerebrospinal fluid (CSF) analysis showed no cellular response. Protein and sugar were within normal limits. The CSF was negative for herpes simplex virus polymerase chain reaction (PCR). HIV and VDRL tests were negative. Anti-MOG antibody testing was not done due to unavailability. Serum beta hCG was within normal limits of pregnancy and ultrasound scan of the abdomen revealed a single live fetus with no sonological evidence of choriocarcinoma.

Immediate treatment with intravenous immunoglobulin (IVIG) 0.4g/kg/day for five days was initiated with empiric antibiotic and antiviral cover along with prophylactic antiepileptics. Later she was started on a dose of IV methylprednisolone 1000 mg daily for five days followed by oral prednisolone at a dose of 1mg/kg daily. Oral steroids were gradually tapered off over the next 4-6 weeks. Within 2 weeks of starting the treatment, the aphasia completely improved while the hemiparesis showed gradual improvement. There was complete recovery at two months from the onset of the illness. Interval resolution of demyelinating lesions with marked reduction in size were seen in the repeat MRI brain scan done two months after treatment (Figure 2).

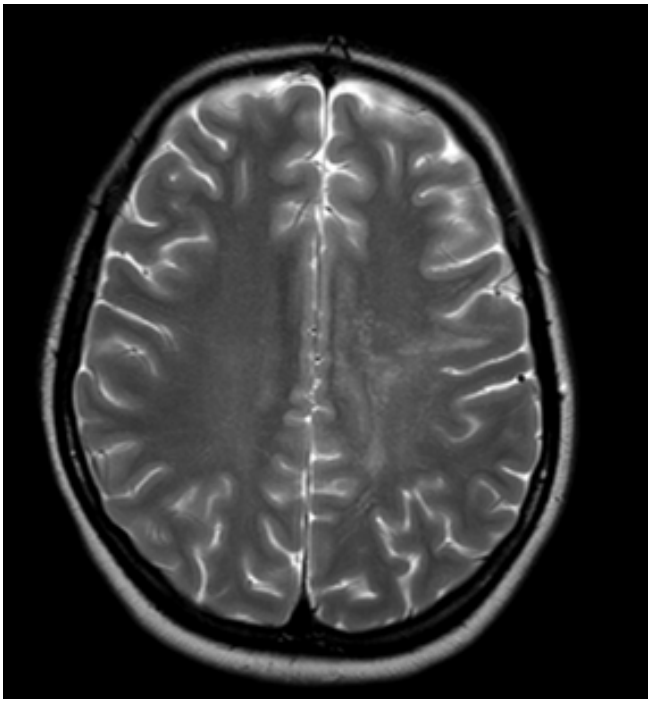

FIGURE 2 **T2W MRI image 2 at two months post-treatment showing improvement of subcortical hyperintense lesions.**

Her serial cervical length assessment at antenatal clinic revealed no risk of pre-term birth and she was euglycaemic throughout. However, she had oligohydramnios with fetal growth restriction. She was admitted at 37 weeks of gestation for the operative delivery. The cardiotocograph was concerning with reduced baseline variability. She underwent a category III caesarean section following administration of a stress dose of IV hydrocortisone as she was on long-term high dose steroids. She delivered a baby with birth weight of 1510 g with no congenital abnormalities. The neonate was observed in the Special Care Baby Unit for 5 days due to low birthweight and

mild respiratory distress. Both the mother and baby were discharged from the hospital on the 6th day post-partum.

DISCUSSION

The most widely accepted hypotheses of pathogenesis of ADEM include molecular mimicry and self-sensitisation secondary to CNS infection, where T helper cells (type 1 and 2), cytokines and autoreactive T cells play a key role.[3,4] Pregnancy-associated changes in the immune status, may worsen the symptoms of ADEM and affect the disease progression and prognosis.[3] Risk of relapse and increased severity may be seen with advancing pregnancy trimesters, posing difficulty in treatment of the disease due to maternal and fetal risks.

Some common differential diagnoses include stroke, infective meningitis and encephalitis, autoimmune encephalitis, cerebral venous sinus thrombosis, multiple sclerosis, CNS complications of HIV, primary CNS lymphoma, cerebral lupus, and neurosyphilis. Neuroimaging plays a main role in distinguishing between ADEM and other conditions. The ADEM diagnosis is confirmed by T2-weighted and fluid attenuated inversion recovery (FLAIR) images where characteristic asymmetric and widespread hyperintense lesions are seen.[5] The American College of Obstetricians and Gynaecologists (ACOG) recommends that the use of gadolinium contrast should be limited during pregnancy, and it should only be used if the diagnosis will significantly improve and its use is expected to improve fetal or maternal outcome. The inability to use contrast media in imaging during pregnancy due to the potential risk to the fetus posed a challenge in the diagnosis of this case. Other supportive investigations in ADEM include CSF analysis, immune profile, and specific antibody tests. Occasionally, brain biopsy which shows typical perivenular demyelinating changes with axonal sparing may be needed to distinguish ADEM from other conditions.[5]

Immune suppression is the mainstay of treatment for ADEM, initially with high dose intravenous corticosteroids.[6] For the non-responders, the treatment options include intravenous immunoglobulin (IVIG) 0.4g/kg/day for five days[7] or plasma exchange.[8] Severe ADEM may be treated with alternative immunosuppressants such as cyclophosphamide,[8] but not during pregnancy due to its teratogenicity. Another challenge faced during the management of this case was the need for treatment with high dose steroids which may increase the risk of preterm delivery, low birth weight, gestational diabetes, neonatal hypoadrenalism and increased incidence of cleft palate.[9] Therefore the risks versus benefits for both mother and fetus need to be carefully considered. Although most patients improve with treatment, complete recovery is seen only in 10-46% of adult patients.[10]

CONCLUSION

Treatment of ADEM in pregnancy is challenging due to maternal and fetal risks associated with immunosuppression. However, prompt treatment with immunosuppression is needed to minimise serious maternal neurological sequalae.

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