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Young female presenting as Acute Disseminated Encephalomyelitis

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Authors' contribution

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Abstract- Acute disseminated encephalomyelitis (ADEM) is an acute inflammatory immune-mediated demyelinating disorder of the central nervous system usually occurring after a post infectious or post vaccineal event.[(1)] It is usually monophasic but can be polyphasic or be recurrent making the diagnosis difficult[(2)]. Commonly affects the children. There is activation of t cell clones causing widespread demyelination involving the white matter of the brain and spinal cord. As it can affect any part of the neuraxis the clinical features are variable and polysymptomatic.

Key Words- acute disseminated encephalitis, demyelinating, inflammatory

Case report:

21 year old female apparently alright 1 day back came to the casualty with the complaints of inability to pass urine since the past 24 hours. Bladder sensation was present but was unable to initiate urine. She was catheterized and about 1 litre of urine was collected. Within the next six hours she started complaining of inability to lift legs or move legs on bed. Over the course of next 3-4 hours, she started complaining of inability to get up from supine position to sitting position in bed or turn from side to side in bed. Over a period of the next eight to ten hours, patient started complaining of weekness in the upper limbs. In the next two to three hours patient was unable unable to speak and chew food or communicate. During the same time, patient did not complain of breathlessness, chest pain, palpitations. The progression of the disease occurred over 24-30

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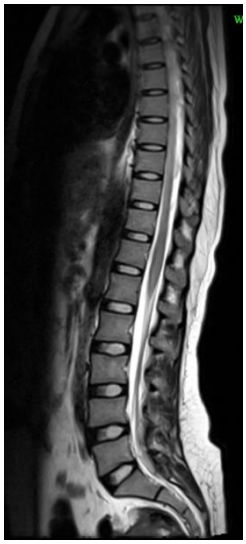
hours. She was able to understand the words and was able to communicate by blinking of eyes. Patient did not state any sensory complaints. There were no involuntary movements of hands and feet. There were no complaints of blurring of vision or retro-orbital pain.

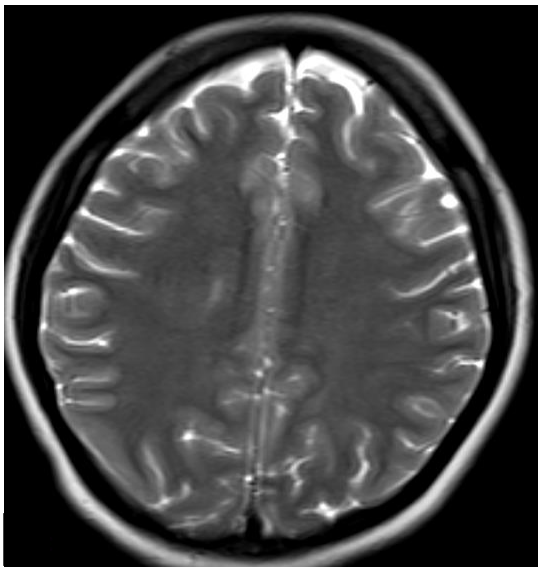
On examination on presentation power in the both lower limbs was 2/5. Bilateral plantars showed extension. Ankle reflex and knee reflex were decreased. There were both one plus. However. The upper limb showed a power of 5 by 5 and the reflexes of stupid of triceps, biceps and supinator were 2 plus. However, over the courts of the next 24 hours, the power in the upper limb decrease from 5/5 to 2/5, and that in the lower limb decrease to 1/5. The reflexes of knee, ankle. Biceps, triceps and supinator all became 1+. On examination, there was bilateral facial nerve palsy. Along with the involvement of the lower cranial nerves. Provisional diagnosis of ?GPS with bilateral facial nerve palsy was made. Patient was shifted to the ICU in view of above complaints, findings and treatment was started.

Past history: patient had a history of viral infection 7 days back for which she took treatment from a local hospital on OPD basis and was resolved in 3 days.

Investigations- Hb-12.9 , TLC-8700, platelets- 3.13 lakhs, sodium- 143, potassium- 3.8, serum procalcitonin- 0.2. LFT, RFT and rest all labs were within normal limits. Testing for the antibodies aquaporin IgG4 and NMO-MOG (neuromyelitis optica-myelin oligodendrocyte glycoprotein) was unsuccessful. ANA by IF was sent which came back negative. CSF:- TLC- 30 (neutrophils- 0, lymphocytes- 100%) CSF- proteins- 34.1 , CSF glucose- 103 (corresponding glucose- 116). CSF culture was sent for bacterial and viral panel, both showed no growth MRI brain Angiography plus venography with whole spine screening was performed and was suggestive of acute disseminated encephalomyelitis.



MRI Spine – hyperintense signal extending from C1 to L1



MRI Brain- hyperintensity present

Treatment- started on inj methylprednisolone 1gm IV OD for 3 days, Inj Pantoprazole 40 mg IV OD, Inj Ondansetron 4mg IV TDS, Inj B-Complex in 100 ML IV OD, and RT feeds were started for the patient. No response to treatment observed after 3 days.

Patient then shifted from Methylprednisolone to Plasmapheresis. 8 cycles of plasmapheresis were done over a period of 17 days. 20% of human serum albumin was used as replacement fluid during each session of plasma exchange

Patient showed significant improvement. Initially it was noticed in the form that patient was able to speak after the first two cycles and was able to chew food. Later over the course of a 15 days the power in upper limbs increased from 2/5 to 4/5 and that in the lower limbs increased from 1/5 to 3/5. The reflexes increased to 2+ of b/l biceps,triceps and supinator. However that of lower limb remained 1+. Patient started having bladder sensations and foleys was removed. Patient was then discharged on t prednisolone 60mg OD with tapering dose and a close follow up was kept on the patient.

Patient has shown significant improvement in the past 6 months and is currently not on any medications. The power in the upper limbs in 4+/5 and in lower limbs has come upto 4/5. Patient is able to speak and chew food. No complaints of chest pain breathlessness, postural hypotension, sweating, bladder incontinence.

Discussion- our patient was a young female who presented with symmetrical ascending flaccid paralysis with lower motor neuron lesion type of bladder.

Anytime there is a close temporal relationship between an infection or a vaccination and the subacute, polysymptomatic start of neurological impairments attributed to the CNS, the diagnosis ADEM should be quickly considered[3]. The most common investigation is MRI which in case of ADEM shows widespread, multifocal, or extensive white matter lesions (lesion load >50% of total white matter volume).[4] In ADEM, deep grey matter lesions have also been observed. These include thalamic or basal ganglia regions, which frequently occur bilaterally and are situated at the confluence of the white and grey matter[5,6]. Our patient showed extensive hyperintensities in the cerebral hemisphere and hyperintensities in the spinal cord extending from C1 to conus medullaris at L1 involving central column.

High-dose intravenous corticosteroids are frequently recommended as the initial course of treatment[7]. The recommended treatment for steroid- and IVIG-resistant conditions is TPE[8,9]. A course consisting of minimum 5 cycles is recommended in case of steroid failure [10]. In our patient high dose corticosteroids were given initially but no response was seen and hence patient was given plasmapheresis. In ADEM instances similar to our investigation, Borras-Novell et al. discovered clinical improvement in all of their patients following plasma exchanges[12]. It is a second-line therapy option that can be administered independently or in concert with immunomodulatory treatments[11].

Conclusion: In patients diagnosed as steroid resistant ADEM cases should be started on Plasmapheresis at the earliest. Doing so is associated with good prognosis.

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