BILATERAL OPTIC NEURITIS: A RARE MANIFESTATION OF ACUTE DISSEMINATED ENCEPHALOMYELITIS

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Summary

Bilateral Optic Neuritis (ON) is an alarming feature for the patient and not commonly faced by clinicians. Acute Disseminated Encephalomyelitis (ADEM) is a demyelinating disease that commonly underdiagnosed due to features sharing that of stroke. But progressive clinical course, behavioral change, features of bilateral Upper Motor Neuron (UMN) lesion, convulsion, bilateral optic neuritis (Sometimes unilateral) can help clinician to think quickly that ADEM could be a possibility. Among its clinical presentation optic neuritis is least common (6%) whereas behavioral change is universal (100%). Acute disseminated encephalomyelitis is suspected in this adult male patient because of progressive painful visual impairment, urinary retention, limb weakness which were associated with behavioral change. After clinical evaluation and relevant investigations he was diagnosed as a case of ADEM, & treated with intravenous methylprednisolone followed by oral prednisolone. During follow up (After 9 weeks) his symptoms and signs improved about 80% whereas MRI lesions regressed about 50%.

Key Words

Acute Disseminated Encephalomyelitis (ADEM); Neuromyelitis Optica (NMO); Bilateral Optic Neuritis (ON).

Introduction

ADEM is an inflammatory disease of central nervous system [1]. In ADEM multifocal demyelinating lesions affect gray & white matter of brain & spinal cord [2]. Prevalence of ADEM is 8/10,00000. ADEM is more common following a viral illness or vaccination and most often seen in children as a monophasic event. ADEM is extensively studied in children but there is paucity of data in adult [3].

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Diagnostic difference between ADEM, Multiple Sclerosis (MS) and other demyelinating disease, like- neuromyelitis optica can be difficult but got paramount importance. According to the new consensus definition of ADEM optic neuritis is one of the least common presenting feature approximately 6% of ADEM patients [2]. Bilateral optic neuritis is uncommon in MS but well recognized in NMO and ADEM [4]. ADEM patients can be presented with bilateral ON. Besides history, clinical examination findings, MRI of brain and spiral cord, consistent Cerebrospinal Fluid (CSF) findings will help to diagnose ADEM, NMO and MS.

Case Report

A 42 years old male hypertensive patient used to take antihypertensive regularly, nondiabetic presented with sudden onset urinary hesitancy urgency followed one day later with retention. Three days later he noticed diplegia of limbs (Spastic all 4 limb weakness more marked in lower limbs) decreased visual acuity with bilateral painful normal ocular movement without any diplopia. He got slurring of speech and features of forgetfulness of both recent and remote events earlier to onset of this episode. He had a history of viral fever persisted for five days about one week before onset of above mentioned clinical features, but no history of headache,neck pain, Joint pain, rash, convulsion, low grade evening rise of temperature, weight loss. His positive clinical findings BP: 130/80mmHg Temperature: 98.4^0F, mildly confused patient (GCS=E4+v4+M4 =12/15). Except evidence of bilateral optic neuritis (Afferent pupillary detect, visual acuity 6/12 in both eyes, red colour desaturation, optic disc swelling) all Cranial Nerves (CN) were intact. Regarding diplegia features of Upper Motor Neuron (UMN) lesion in all 4 limbs and extensor plantar response on both sides with ataxic gait but without any truncal sensory level, any modalities of sensory impairment and cerebellar sign.

MRI of brain and dorsal spine with whole spine survey without contrast revealed multiple, asymmetrical lesions of variable sizes diffusely involving different parts of both cerebral hemispheres and right cerebellum which didn't follow any particular arterial territory, hypointense in T1 but hyperintense in T2 and FLAIR images. Same type of small one segment lesion at D7 level associated with segmental mild cord swelling also noted. CSF examination revealed normal pressure, cell count and sugar level but raised protein (110mg/dl). Oligoclonal Band (OCB) in CSF and NMO antibody couldn't advised because these facilities are not available in the city.

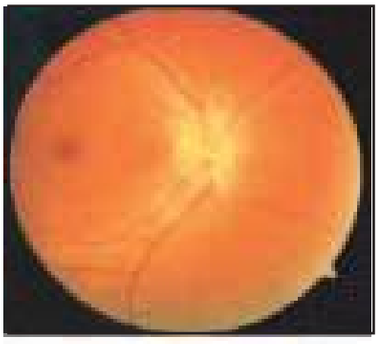

Fig 1 : Fundal Photograph of the patient showing features of optic neuritis

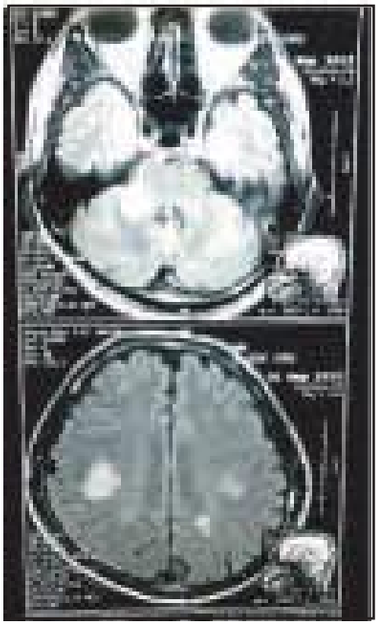

Fig 2: FLAIR image of MRI brain of the patient showing demyelinating lesions involving different areas

Discussion

ADEM is an emerging neurological disease in our perspective. In ADEM behavioral change ranging from mild irritability to coma found in 100% cases whereas ON is least common. Regarding causes of ON demyelinating CNS disorders are important and MS, ADEM, NMO are commonly seen in clinical practice. NMO is also associated with predominant spinal cord related clinical features without behavioral abnormality and long (Usually>3) spinal segment lesions. ADEM is unique with behavioral change, diffuse cerebral, cerebellar, brainstem and occasionally short segment or diffuse spinal cord lesion. Different studies confirmed the overall good outcome of ADEM [5]. In conclusion it is to be noted that in presence of bilateral optic neuritis if features of behavioral change are not inquired diagnosis of ADEM could be missed. Presence of optic neuritis facilitates the specialist to find out any behavioral change and presence of bilateral optic neuritis with behavioral change almost exclude NMO and help consultant early refer the patient to neurologist because management and long term prognosis are different in this two conditions.

Disclosure

Both the authors declared no competing interest.

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