An Interesting Case Of Acute Disseminated Encephalomyelitis

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Abstract:
Acute disseminated encephalomyelitis (ADEM) is an uncommon inflammatory demyelinating disease of the central nervous system. The disease typically occurs after infections or vaccinations. However, in many patients with ADEM, no evidence of prior infection or vaccination can be found. We are reporting a patient who developed clinical and radiological features of acute disseminated encephalomyelitis after trauma of repeated attempts at lumbar puncture for spinal anesthesia.

Key Words: Acute disseminated encephalomyelitis (ADEM), Monophasic, Exanthematous fevers
Introduction:
Acute disseminated encephalomyelitis (ADEM) is an unusual monophasic or multiphasic demyelinating disorder of the CNS. Although the pathophysiology of ADEM is largely unknown, an autoimmune response to myelin basic protein triggered by infection or immunization is strongly suspected to be the main etiological factor. The disease typically occurs after infections or vaccinations. However, in many patients with ADEM, no evidence of prior infection or vaccination can be found. Some investigators have reported that ADEM can follow treatment with certain medications, can occur on the heels of trauma, and can develop with absolutely no precipitating cause (idiopathic ADEM). We report a case of acute disseminated encephalomyelitis presenting after trauma of repeated attempts at lumbar puncture.

Case Report:
A 56 year old male government employee was referred to our institution with complaint of inability to move both lower limbs for last five days. History revealed that patient had been posted for surgery for direct inguinal hernia of the right side under spinal anesthesia. Patient was taken to operation theater but surgery had to be deferred because repeated attempts at lumbar puncture failed and spinal anesthesia could not be administered. The CSF was not aspirated during any of the attempts and the anesthetist did not inject any anesthesia. Patient reported loss of sensations in both lower limbs and inability to move both lower limbs from the time he was shifted back from operation theater. There was also history of bowel and bladder incontinence from that day. Patient underwent surgery next day under local anesthesia and was subsequently referred to our institution. There was no history of recent febrile illness, symptoms of upper respiratory tract illness or any immunization. On examination patient was conscious and his general physical examination was within normal limits. Nervous system examination showed that higher mental functions, speech and cranial nerves were within normal limits. Sensory system examination showed symmetrical pan-sensory loss below nipples. Motor examination showed power was reduced in both lower limbs and power of various groups of muscles ranged from grade 1 to grade 4. Power in upper limbs was normal. Knee jerks were exaggerated bilaterally, rest of the deep tendon reflexes were normal. Superficial reflexes were bilaterally absent and plantars were bilaterally mute. Abdominal examination showed a healthy scar of right inguinal hernia surgery. Rest of the systemic examination was within normal limits. Investigations including blood urea, serum creatinine, serum bilirubin, AST, ALT, Alkaline phosphatase, serum electrolytes, complete blood count, chest x ray were normal. MRI brain and spinal cord showed focal hyperintensities in upper cervical spinal cord opposite 3rd to 7th cervical vertebra (Fig.1) and white matter hyperintensities in Rt. Cerebellar hemisphere, medulla, Pons, B/L Basal Ganglia and B/L Temporal lobes (Fig.2). Radiological diagnosis of acute disseminated encephalomyelitis was kept. Patient was treated with intravenous methylprednisolone 1 gram o.d for 3 days followed by an oral taper. Patient showed steady improvement during his stay in the hospital and was able to walk and perform his daily chores with minimal support and had regained complete bowel and bladder control within one week of admission.
Discussion:
Acute disseminated encephalomyelitis (ADEM) is an uncommon inflammatory demyelinating disease of the central nervous system. The true incidence of the disease in India is undetermined and is likely to be more frequent than reported, as the common antecedent events e.g. exanthematous fevers which predispose to ADEM, are still prevalent. Differentiation of ADEM from the first attack of multiple sclerosis (MS) is important from prognostic as well as therapeutic point of view. However, in the absence of biological markers, at times differentiation of ADEM from the initial presentation of MS may not be possible even by combination of clinical, CSF analysis, and MRI. This differentiation is more relevant to India where the incidence of MS is low.

Trauma rarely has been reported as a preceding event in pathogenesis of acute disseminated encephalomyelitis. However in the patient discussed here causal relation ship with the trauma of preceding repeated lumbar puncture is probable as there are no other apparent probable precipitating events. The MRI findings and the fact that the dural space was not reached during attempts of lumbar puncture indicate that the deficit was not due to the complications of lumbar puncture itself or of any anesthetic agent. It is still difficult to differentiate between acute disseminated encephalomyelitis and the first manifestation of multiple sclerosis. The criteria for the diagnosis of multiple sclerosis are not met because of the monophasic, acute course of the disease. In addition, the age of onset (56 years) is uncommon for a first presentation of multiple sclerosis. Though there was definite temporal relation ship of trauma and ensuing neurological deficit in our patient, it is difficult to say that the trauma of attempted lumbar puncture had led to the development of acute disseminated encephalomyelitis because ADEM usually occurs several days after trauma or vaccination. It may just be a coincidence of idiopathic ADEM and trauma of attempted lumbar puncture.

References: