Case Report

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Acute Disseminated Encephalomyelitis Presenting as Stiff-Person Syndrome in Middle-Aged Adult: Case Report

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Abstract

Acute Disseminated Encephalomyelitis (ADEM) is a monophasic, inflammatory demyelinating disorder of the white matter often preceded by viral infection or recent vaccination. It is commonly seen in children and young adults, where prognosis is favorable. To date, very few cases of ADEM has been reported in middle-aged and elderly patients. Approximately 25% of ADEM cases in adults will develop Multiple Sclerosis (MS) within five years of initial presentation but majority of individuals do not progress beyond three months. Stiff-Person Syndrome (SPS) is a rare neurologic disorder with wide variety of symptoms that mimic neuro-inflammatory and demyelinating disease which causes a delay in the diagnosis. Here we present a case of ADEM in a middle-aged adult masquerading as SPS on initial presentation.

Keywords: Stiff-Person Syndrome (SPS); Acute Disseminated Encephalomyelitis (ADEM); Cerebrospinal fluid; Hyperreflexia

Introduction

Acute Disseminated Encephalomyelitis (ADEM) is a monophasic, inflammatory demyelinating disorder of the white matter often preceded by viral infection or recent vaccination. It is commonly seen in children and young adults, where prognosis is favorable. To date, very few cases of ADEM has been reported in middle-aged and elderly patients [1]. Approximately 25% of ADEM cases in adults will develop Multiple Sclerosis (MS) within five years of initial presentation but majority of individuals do not progress beyond three months [2]. Stiff-Person Syndrome (SPS) is a rare neurologic disorder with wide variety of symptoms that mimic neuro-inflammatory and demyelinating disease which causes a delay in the diagnosis [3].

Case Study

Here we present a case of ADEM in a middle-aged adult masquerading as SPS on initial presentation. A 44-year old, right handed, female, Overseas Filipino Worker (OFW) from Cabanatuan City presented with a 6-month history of progressive stiffness and painful spasms of her upper and lower extremities. Two months prior to admission, her condition worsened resulting to an inability to stand and ambulate. She had no known co-morbidities and had unremarkable family history. On admission, her limbs were rigid and movements were severely limited and painful. The strength could not be assessed because of rigidity and spasms. There was an increased tone on both extremities and hyperreflexia. Babinski sign was present bilaterally. Sensory examination was normal. She had pseudobulbar affect and her masseter and temporalis muscles were spastic resulting to restriction in opening her jaw leading to poor nutritional intake. Her intellect was normal and she had no other neurological abnormalities.

Results and Discussion

Results of the routine laboratory tests which include complete blood count, electrolytes, glucose, lipid profile and liver enzymes were normal except for hypoalbuminemia (Albumin 2.75 g/dl). Chest roentogram showed bilateral pleural effusion whereas abdominal ultrasound revealed uterine myoma. Cerebrospinal Fluid (CSF) examination was normal with no atypical cells found on cytopathological analysis. CSF cultures were also negative. Cranial CT scan done on admission was likewise unremarkable. MR imaging of the brain revealed patchy T2 hyperintensities in the frontoparietal periventricular subcortices consistent for a demyelinating disease **(Figure 1)**.

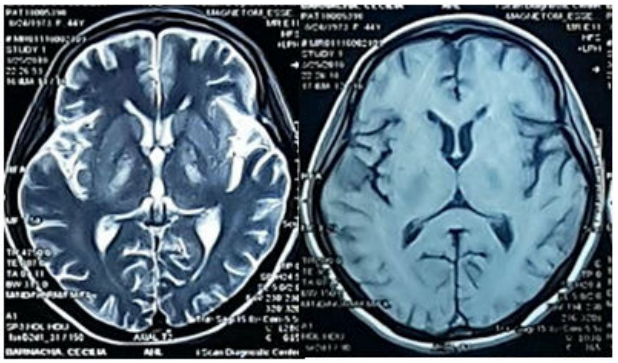

Figure 1: Cranial MRI shows patchy T2 hyperintensity foci in the fronto-parietal area periventricular subcortices, these appears as isointensities on T1.

There is also diffuse slowing of the background activity seen on electroencephalogram suggestive of encephalopathy. She was started on pulse therapy with Methylprednisolone (1 gram/day for 5 days) as well as with Baclofen 30 mg/day and Diazepam 20 mg/day which reduced the spasticity. Her neurological examination on discharge showed marked improvement of limb rigidity and absence of pseudobulbar affect. Her masseter and temporalis muscles were no longer spastic and she was able to eat well. Out Patient follow up done at 90 days' post-discharge, revealed normal muscle tone and reflexes. She was also able to ambulate without assistance and was independent on her activities of daily living with a Modified Rankin Score (MRS) of 0.

The patient presented with an insidious onset and progressive symptoms. The stiffness and painful spasms became so severe that her movements became limited. Her ability to eat was also impaired. The presence of pyramidal signs like hyperreflexia and Babinski likely involves a problem in the corticospinal tract particularly the upper motor neuron thus, other diseases which might present with rigidity and spasms like Stiff-Person Syndrome (SPS) and Neuromyotonia were ruled out as possible diagnosis. Several diagnostic modalities, including imaging, was done to search for evidence of an infectious, vascular and malignant etiology but results were pertinent only for T 2 hyperintensities suggestive of a demyelinating disease hence, ADEM and MS were considered as possible differentials.

Since ADEM and MS are both disseminated disorders of the central nervous system, broad range of neurological signs-pyramidal, cerebellar and brainstem are possible. However, encephalopathy with depressed consciousness and altered sensorium is more common in ADEM than in MS [4]. An essential part in the investigation of ADEM is the cranial MRI. A number of studies have reported differences in the imaging features between ADEM and MS. MS lesions have well-defined plaque-like margins whereas ADEM lesions often have poorly defined margins. The majority of lesions in ADEM tend to be in the deeper white matter with periventricular sparing. Only 29%-60% of patients with ADEM have periventricular lesions demonstrated in the MRI [5]. Since our patient presented with disseminated white matter lesions with poorly defined margins, the diagnosis of ADEM is favored.

ADEM is an immune-mediated inflammatory demyelinating condition that affects the white matter of the brain and the spinal cord. It is more commonly seen in children than in adults following a viral infection [6]. The pathogenesis of ADEM although incompletely understood appears to be an autoimmune disorder of the central nervous system that is triggered by an environmental stimulus in genetically susceptible individuals. One proposed mechanism is that myelin autoantigens share antigenic determinants with those of infecting pathogens. This anti-viral antibodies or cell-mediated response to the pathogen cross-react with the myelin pathogens thus, resulting in ADEM [7].

ADEM may also be due to an increased vascular permeability and congestion in the central nervous system triggered perhaps by a circulating immune complexes or other humoral factors that develop after exposure to a foreign antigen introduced by infection or vaccination [8]. This process leads to infiltration in the vessel walls by mononuclear cells, followed by perivenous edema and occasionally hemorrhages. Within days, microglia, lymphocytes, and phagocytes appear, ultimately causing demyelination and possible gliosis and necrosis. The extent of demyelination and the subsequent glial and neuronal changes account for the variation in clinical features and disease prognosis [9].

There have been no controlled trials to determine the efficacy of immunomodulatory treatments in ADEM. However, most patients with suspected inflammatory demyelinating CNS diseases are treated with steroids. It is common practice to use 10-30 mg/kg/day intravenous methylprednisolone (maximum 1 g/day) for three days with the clinical improvement seen mostly in patients after therapy [10].

Our patient presented with the clinical history, neurologic exam, CSF results, imaging and electroencephalogram findings suggestive of ADEM. It is important to make it clear in this case that it does not represent as a first demyelinating event of MS given its sub-acute presentation of encephalopathy confirmed by the results of the EEG and the presence of multifocal ill-defined lesions on the white matter showing inflammation. In contrast, MS patients presents with distinct episodes of focal neurological deficits with confluent demyelinating lesions affecting the white more than the gray matter [11].

Conclusion

The disseminated involvement of the white matter in Acute Disseminated Encephalomyelitis accounts for its varied clinical presentation that initially may manifest as other clinical syndromes like SPS. However, pyramidal signs and imaging favors ADEM as the diagnosis. Differentiating the diagnosis of ADEM from MS is quite difficult since the differences are quite subtle and the possibility of an ADEM to develop into MS is likely. Nevertheless, imaging findings of poorly defined white matter regions and clinical findings suggestive of a demyelinating disease as well as the absence of symptoms recurrence 90 days' post-discharge leads to ADEM as the most likely diagnosis than MS for this patient.

References

1. MR Rahmlow, O Kantarci (2013) Fulminant demyelinating diseases. Neurohospitalist 3: 81-91.
2. Nicole M, Peter AA, Mark C, Barak B (2015) A case of acute disseminated encephalomyelitis in a middle-aged adult. Case Rep Neurol Med 2015.
3. Hajjioui A, Benbouazza K, Faris M, Missaoui A (2010) Stiff limb syndrome: A case report. Cases J 3: 60.
4. Dale RC, Branson JA (2005) Acute disseminated encephalomyelitis or multiple sclerosis: Can the initial presentation help in establishing a correct diagnosis? Arch Dis Child 90: 636-639.

5. Ketelslegers IA, Neuteboom RF, Boon M, Catsman-Berrevoets CE, Hintzen RQ (2010) A comparison of MRI criteria for diagnosing pediatric ADEM and MS. Neurol 74: 1412-1415.

6. S Schwarz, A Mohr, M Knauth, B Wildemann, B Storch-Hagenloche (2001) Acute disseminated encephalomyelitis: A follow-up study of 40 adult patients. Neurol 56: 1313-1318.

7. Reik L (1980) Disseminated vasculomyelinopathy: An immune complex disease. Ann Neurol 7: 291-296.

8. Young NP, Weinshenker BG, Parisi JE (2010) Perivenous demyelination: Association with clinically denied acute disseminated encephalomyelitis and comparison with pathologically con irmed multiple sclerosis. Brain 133: 333.

9. Rudick RA, Eskin TA (1981) Enhanced vascular permeability in acute disseminated vasculomyelinopathy. Annals Neurol. 9: 621-621.

10. Dale RC, de Sousa C, Chong WK (2000) Acute disseminated encephalomyelitis, multiphasic disseminated encephalomyelitis and multiple sclerosis in children. Brain 123: 2407-2422.

11. Hynson JL, Kornberg AJ, Coleman LT (2001) Clinical and neuroradiologic features of acute disseminated encephalomyelitis in children. Neurol 56: 1308-1312.