



A case of atypical acute disseminated encephalomyelitis mimicking primary angiitis of the CNS and intravascular lymphomatosis

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INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is a monophasic neurological disease triggered by infectious illnesses or vaccines. It is characterized by encephalopathy ranging from lethargy to coma, and by focal and multifocal neurological signs like hemiparesis, cranial nerve palsies and paraparesis. Additional features include meningismus, ataxia, seizures and varied movement disorders. Systemic symptoms (fever, malaise, myalgias, headache, nausea and vomiting) often precede the neurological symptoms¹. Given the heterogeneous clinical, cerebrospinal fluid (CSF) and radiological findings of the disease, differential diagnosis of ADEM include various conditions such as multiple sclerosis, vasculitis, reversible posterior leucoencephalopathy, neurosarcoidosis, progressive multifocal leucoencephalopathy, HIV infection, toxic and mitochondrial encephalopathies and osmotic myelinolysis. Nonetheless, some rare entities like intravascular lymphomatosis can mimic ADEM as well, both clinically and radiologically.²

CASE REPORT

In May 2014 a 40-year-old man reported a two-week history of flu-like syndrome, associated to diarrhea, headache, diffuse myalgias and arthralgias, followed by sudden onset of gait disturbance, diplopia, confusion and drowsiness. Brain computed tomography (CT) was normal. When admitted to the Neurology Unit, CSF examination showed a mild increase in protein levels (57 mg/dl) with lymphocytic pleocytosis (60 cell/mm³) and normal glucose levels; oligoclonal bands were detected at isoelectrofocusing (IEF).

CSF virological and bacteriological tests were negative; CSF cytology revealed an inflammatory pattern, with a predominance of CD4+ lymphocytes.

Complete serum virological assessment (EBV, VZV, HSV, CMV, HHV6, HHV8, Adenovirus) was negative, except for a positive anti HSV1/2 IgM title, suggestive of a recent infection.

Patient was first treated with I.V. Acyclovir and Ampicilline in the hypothesis of infective leucoencephalopathy, without clinical improvement.

Brain magnetic resonance imaging (MRI) showed diffuse punctate T2 hyperintense perivascular lesions involving basal ganglia and corona radiata bilaterally, some of them with contrast enhancement. Diffuse perivascular white matter alterations in brainstem and cerebellum were detected as well. (Fig. 1 and 2)

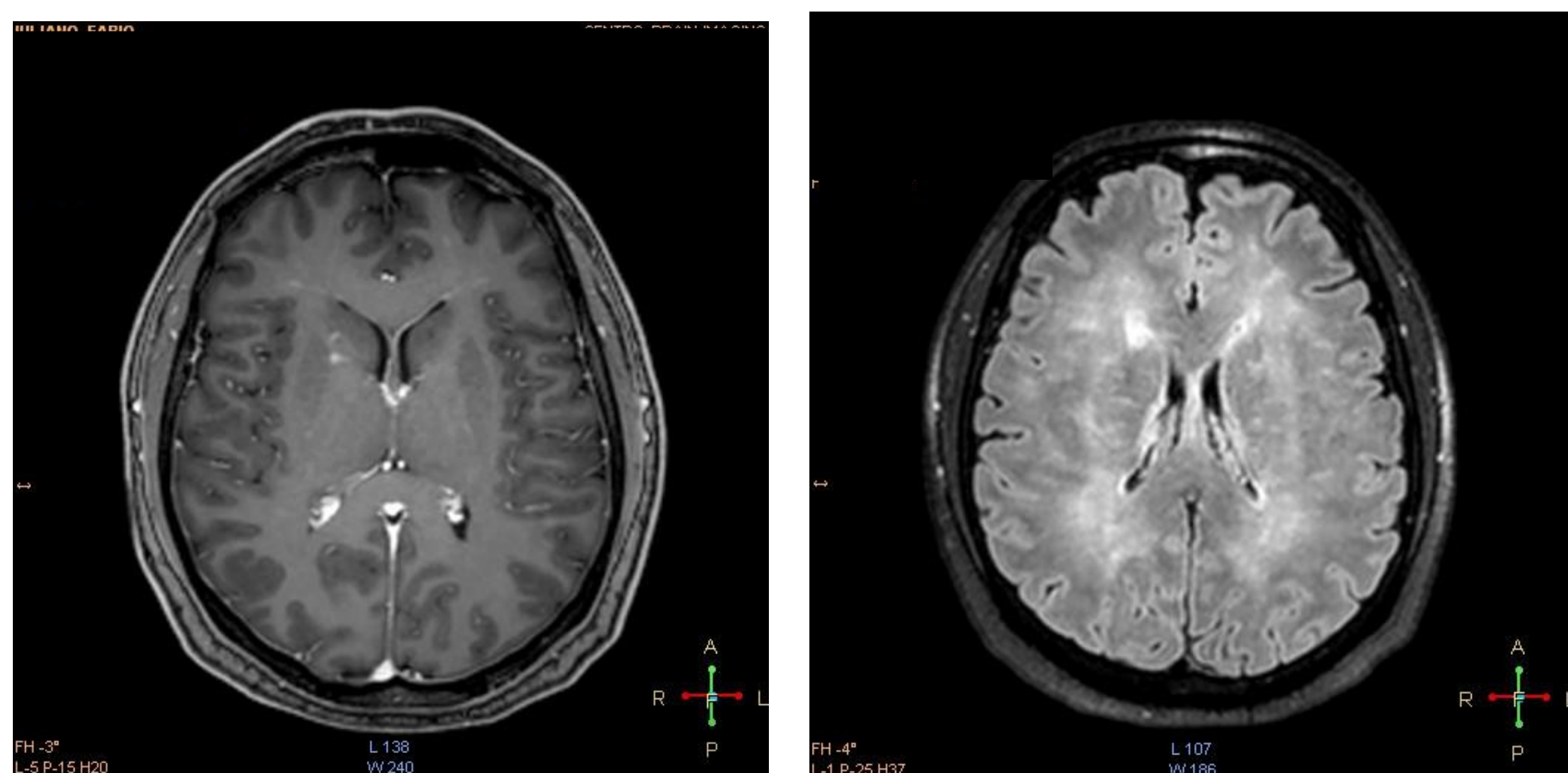


Fig 1 and 2. Brain MRI showing diffuse punctate T2 hyperintense perivascular lesions, some of them contrast-enhancing.

The radiological findings were considered atypical for ADEM; a diagnosis of possible CNS vasculitis was suggested. The hypothesis of an intravascular lymphomatosis was considered as well. Several instrumental and laboratory examinations were performed, including immunorheumatological screening; chest/abdomen CT; retinal fluorangiography; esophagogastroduodenoscopy with duodenal biopsy. They all tested negative. Since MRI alterations were adjacent to small vessels, cerebral angiography was not performed. Patient was treated with high dose I.V. corticosteroids (methylprednisolone 1 g/day) for two weeks, followed by oral tapering, with prompt clinical improvement. Cerebral biopsy was non performed given the rapid clinical recovery. A diagnosis of atypical ADEM was performed.

One-year clinical and neuroradiological follow-up, in the absence of chronic steroid therapy, demonstrates almost complete regression of brain MRI alterations, without recurrence of neurological impairment or long-term disability.

DISCUSSION

In our case report, MRI findings were different from the ones of classical ADEM, which usually shows patchy T2 hyperintense lesions involving mainly white matter and possibly grey matter as well (in particular basal ganglia, thalamus and brainstem).

The first disease considered in the differential diagnosis was vasculitis, namely primary angiitis of the CNS (PACNS), which can show diffuse white matter lesions on MRI, suggestive for microangiopathy, with ischemic infarctions or haemorrhagic lesions.³ An increased cell count on CSF examination (with occasional presence of oligoclonal bands) is a feature of both PACNS and ADEM.

Intravascular lymphomatosis is a rare but clinically important differential diagnosis of encephalopathy and encephalitis². Diagnosis is based on histological finding of a neoplastic proliferation of large, atypic, lymphoid cells (mainly B lymphocytes) in lumina of capillaries and small arteries; prognosis of the disease is very poor in most cases. MRI findings include non-specific white matter lesions, multiple mass lesions, progressive infarct-like alterations or dural and arachnoidal enhancement. CSF often reveals mild increase of cell count without cytological signs of malignancy.

Both in CNS vasculitis and intravascular lymphomatosis, a definite diagnosis can be established only by biopsy of clinically affected organs.

In our case biopsy was not performed given the rapid and sustained clinical and neuroradiological response after steroid therapy.

CONCLUSIONS

In the case of a meningoencephalitic syndrome, especially if with heterogeneous brain MRI findings, multiple neurological and systemic evaluations are often required to exclude some rare but life-threatening diseases.

In our case report, the negativity of CSF citofluorimetry and of instrumental and laboratory systemic evaluations, the complete and persistent clinical recovery even after corticosteroid withdrawal, make a diagnosis of ADEM plausible, even if with atypical MRI findings. However, a close clinical and neuroradiological follow-up is needed in order to exclude a possible inflammatory reactivation, which may lead to a re-consideration of final diagnosis.

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