



# A child with acute onset of neurological signs

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An eight year old boy presented with a short history of headache, vomiting, drowsiness and increasing left sided weakness, following an episode of a flu-like illness one week earlier. On examination he was drowsy, with neck stiffness, a left hemiparesis, and mild right sided weakness. His initial CT scan is shown (Figure 1 a, b). A lumbar puncture revealed a mildly increased protein level and a normal cell count. MRI was subsequently performed (Figure 2 a, b, c). He was commenced on steroids and showed rapid improvement of neurological signs.

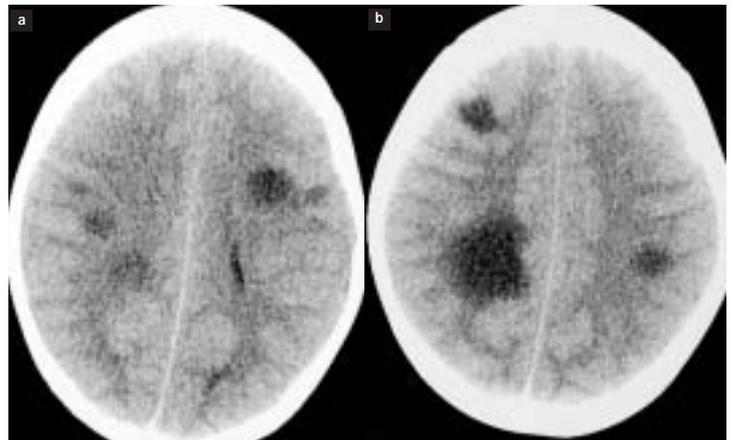


Figure 1 a) b). CT scan showing multiple lesions



Figure 2 a) b) c). MRI showing multiple lesion

## Question 1

The imaging findings shown in Figures 1 and 2 are:

- A. multiple lesions with little mass effect involving white matter only
- B. multiple lesions with no mass effect involving white matter and deep grey matter
- C. multiple periventricular lesions
- D. multiple mass lesions.

## Question 2

What is the most likely diagnosis?

- A. acute viral meningitis
- B. acute disseminated encephalomyelitis (ADEM)
- C. acute viral encephalitis
- D. gliomatosis cerebri or multifocal gliomas.

## Question 3

The differential diagnosis of the imaging findings includes:

- A. cerebral vasculitis eg. systemic lupus erythematosus (SLE)
- B. toxin exposure
- C. progressive multifocal leukoencephalitis (PML)

- D. multiple sclerosis
- E. all of the above.

## Question 4

Which of the following statements are true?

- A. ADEM does not occur in adults
- B. The presence of contrast enhancement would indicate that tumour is more likely than demyelination
- C. The diagnosis of ADEM can usually be made by a combination of typical clinical presentation and imaging findings
- D. MRI is more sensitive than CT for cerebral lesions of ADEM
- E. Cerebral abnormalities in ADEM are due to demyelination and usually resolve.

## Question 5

ADEM may follow:

- A. exanthematous viral illnesses
- B. vaccination
- C. mycoplasma infection
- D. no preceding illness
- E. all of the above.

## Answers

### 1. Answer B

Both CT and MRI images show multiple abnormalities. Several large lesions are seen in the deep cerebral white matter, but there is also bilateral involvement of the thalami. There is no significant mass effect associated with these abnormalities.

### 2. Answer B

The combination of an acute neurological illness following a viral infection, with asymmetrical bilateral lesions in white matter, deep grey matter and occasional brain stem and spinal cord involvement is typical of acute disseminated encephalomyelitis (ADEM). Uncomplicated viral meningitis typically has no focal neurological signs and normal MRI. Viral encephalitis eg. herpes simplex type 1 (HSV-1) is an important diagnosis to consider and treat early. It typically produces temporal and inferior frontal grey and white matter abnormality on imaging, but in equivocal cases, antiviral therapy is often instituted before other results are available. Multifocal gliomas could cause white matter and thalamic infiltration, and could present as an acute illness, but tumours often have more mass effect and patchy irregular contrast enhancement.

### 3. Answer E

The imaging features of ADEM are nonspecific, and require clinical correlation. The differential diagnosis is very long, and includes:

- CNS infections eg. HSV-1 encephalitis, PML (caused by papovavirus in immunocompromised patients), human immunodeficiency viral (HIV) encephalopathy, subacute sclerosing panencephalitis
- noninfectious inflammatory conditions eg. multiple sclerosis (MS), cerebral vasculitis (eg. SLE) and other collagen vascular disorders
- cerebral infarction
- toxin exposure
- metabolic leukoencephalopathies and mitochondrial disorders
- trauma
- tumour
- radiation or chemotherapy injury.

In older adults, patchy white matter abnormality related to hypertension and aging may have a similar appearance.

### 4. Answer

A. *False*

ADEM usually occurs in children or young adults, but can be seen in much older patients.

B. *False*

While enhancement following IV contrast and mass effect are typical features of tumours, both these features are sometimes seen in regions of acute demyelination (in ADEM and MS), due to inflammatory alteration of the blood–brain barrier and oedema.

C. *True*

The diagnosis of ADEM is generally made on the basis of a monophasic illness following viral infection or vaccination, CSF analysis, imaging studies, and the absence of other definable conditions. In some cases it may be difficult to distinguish ADEM from MS, leukodystrophy, or mitochondrial disorders.

D. *True*

CT may show abnormalities as in this case (Figure 1), but is relatively insensitive. False negatives occur, and CT is unlikely to show the full extent of the abnormalities.

E. *True*

The prognosis of ADEM is usually good, with the highest morbidity (around 10%) in the first week of the illness. The MRI lesions usually resolve within a few weeks, responding rapidly with steroid therapy. Some lesions resolve more gradually over 6–18 months after the onset of symptoms, and organise into glial scars. Approximately 10–30% of patients have some permanent neurological deficit. If the number of the lesions increases on long term follow up, or if the patient develops new neurological symptoms, then MS is the more likely diagnosis.

### 5. Answer E

ADEM is an immune mediated disorder causing perivenous inflammation and demyelination. It often occurs days to weeks after a systemic viral infection (measles, rubella, varicella, Epstein-Barr virus, mumps, pertussis, influenza, herpes simplex, herpes zoster) or

postimmunisation. It is rarely seen after mycoplasma or other bacterial infection, and in some cases no precipitating cause is identified.

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