

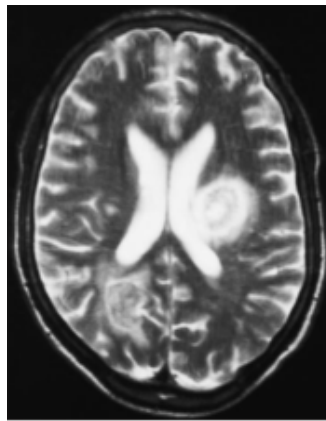
EANS/UEMS European examination in neurosurgery

Variants of questions with answers (compilation - Vyacheslav S. Botev, Department of Neurosurgery, M.Gorky Donetsk National Medical University)

CASE HISTORIES IN HIV/AIDS

Case 1

A 37-year-old HIV positive male presents with a 1-week history of headache and progressive left-sided weakness. On examination he is alert and orientated. He has a left homonymous hemianopia and pyramidal weakness affecting the left face, arm and leg with hyperreflexia and bilateral extensor plantar responses.



1. What is the differential diagnosis of the abnormality on the T2 MRI brain-scan?
2. What blood tests and what other investigations may help in making a diagnosis?
3. How should he be managed?

Case 2

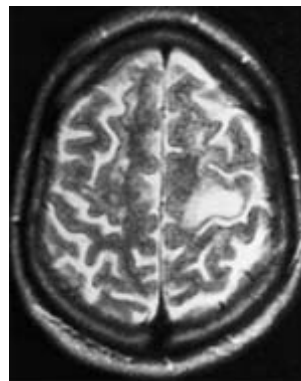
An HIV positive male with a CD4 count of 50 cells/mm³ presented with a 4-day history of back pain, followed by the development of weakness of his legs. By the time of admission he was incontinent of urine. On examination he had a flaccid paralysis of the legs with grade 3 power proximally and grade 0 distally. The knee and ankle reflexes were absent and the plantars unreactive. Pin prick sensation was reduced to the mid thigh on the right and the mid shin on the left. A MRI scan of the thoraco-lumbar spine was normal. CSF examination showed: WBC 60/mm³ (90% neutrophils); protein 1.1 g/L (0.11 g/dl); sugar 3.2 mmol/L (57.7 mg/dl); blood sugar 6.3 mmol/L (113.5 mg/dl).

1. What is the differential diagnosis?

2. What is the most likely cause?
3. What other tests or examination will help confirm the cause?

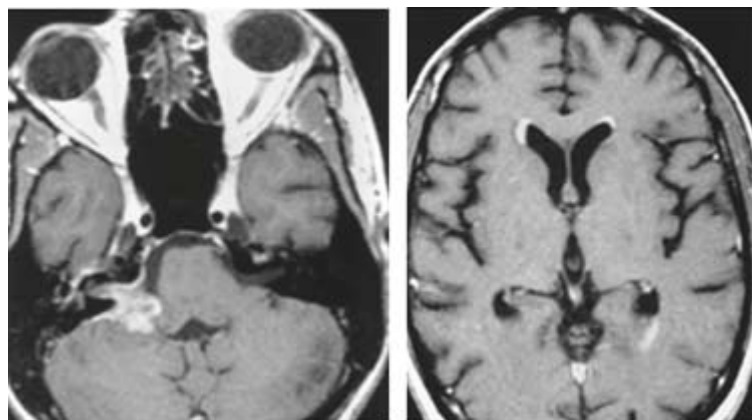
Case 3

A 42-year-old homosexual male presents with a 6-week history of progressive weakness of the right arm and a 2-week history of language disturbance. On examination, he is afebrile and general examination is normal apart from some white mucosal lesions intraorally. Neurologic examination reveals a mild expressive dysphasia and a flaccid right arm with MRC grade 3 power proximally and grade 2 distally. MRI is performed and shown below.



1. What blood tests should be performed?
2. What is the differential diagnosis of the lesions on MRI?
3. What other investigations may be performed to confirm the diagnosis?

Case 4



1. What is the differential diagnosis for the findings identified on imaging?
2. What imaging findings are typical of CNS involvement in patients infected with cytomegalovirus (CMV) in utero?
3. What is the most neoplastic common cause of ependymal enhancement in the

setting of AIDs infection?

4. What were the patient's acute symptoms at clinical presentation?

Answers

Case 1

1. The differential diagnosis in an HIV positive patient with a low CD4 count lies between toxoplasmosis, primary CNS lymphoma, and tuberculoma.

2. If the toxoplasma serology is positive, this means the patient has been exposed to the organism and is vulnerable to reactivation when immunosuppressed. Over 95% of toxoplasmosis in HIV is a reactivation rather than de novo infection. A MRI may be helpful since a single lesion on MR is more likely to be lymphoma. More recently, thallium-201 SPECT scans have been utilized – lymphoma showing increased uptake relative to toxoplasma abscesses.

3. The standard treatment is to treat patients with anti-toxoplasma drugs such as sulphadiazine and pyrimethamine plus folinic acid for at least 2 weeks. If there is a significant response the diagnosis is one of toxoplasmosis. If the patient deteriorates a stereotactic brain biopsy should be considered. The patient continued to deteriorate. A brain biopsy showed primary CNS lymphoma.

Case 2

1. The differential diagnosis of this lumbar polyradiculopathy syndrome in an HIV infected patient is lymphomatous infiltration, syphilitic meningoradiculitis, herpes simplex type 2 and CMV polyradiculopathy.

2. The finding of a neutrophil pleocytosis suggests that CMV is the likely cause. In such cases the MRI scan may be normal or may show thickened nerve roots.

3. PCR for CMV DNA. CSF cytologic examination will help exclude a lymphomatous process but repeated CSF examination may be required. There may be evidence of CMV infection elsewhere and therefore a careful examination of the retina is necessary.

Case 3

1. The white mucosal lesions could be candidiasis which is usually found in patients who are on corticosteroids (oral or inhaled), diabetics, or patients who

are immunosuppressed for any reason such as infection with the HIV. An HIV blood test should be performed and if that is positive then a CD4 count and a viral load should be measured in order to obtain an idea about the degree of immunosuppression.

2. The differential diagnosis of such lesions in an HIV infected person includes PML, CMV encephalitis and HIV encephalopathy. The symptomatic lesion shown on MRI does not show mass effect or enhancement with contrast and therefore is unlikely to be due to toxoplasmosis or primary CNS lymphoma. CMV encephalitis usually presents more acutely; patients usually have evidence of CMV disease elsewhere such as a CMV retinitis, and MRI may show a periventriculitis. HIV encephalopathy presents with a subcortical dementia but no focal neurologic signs. MRI shows changes that are much more diffuse.
3. The diagnosis of PML may be confirmed by the detection of the JC virus using PCR. This has a sensitivity of around 80% and a specificity of 95%. A brain biopsy, as performed in this patient, will show the characteristic features of demyelination in association with inclusion bodies within deformed oligodendrocytes and bizarre looking astrocytes.

Case 4

Cytomegalovirus meningitis and ependymitis in a patient with AIDS.

1. Infection (ventriculitis, ependymitis) and neoplasms (lymphoma or seeding from a systemic or primary brain neoplasm).
2. Bilateral ventricular subependymal calcification, ventricular enlargement, periventricular hypodensity on CT or hyperintensity on T2W MR imaging, atrophy, and migrational anomalies (pachygyria or polymicrogyria).
3. Lymphoma.
4. Sensorineural hearing loss and vertigo.

Comment

Cytomegalovirus is present in the latent form in the majority of the American population. Reactivation usually results in a subclinical or mild flu-like syndrome. In immunocompromised patients, reactivation can result in disseminated infection, usually involving the respiratory and gastrointestinal tracts; however, rarely, it can infect the nervous system. In the CNS, CMV may cause meningoencephalitis and ependymitis. Symptoms may be acute or chronic, developing over months. Patients may have fever, altered mental status, and progressive cognitive decline. Patients may also present with cranial neuropathies (as in this case). CMV polymerase chain reaction in the CSF is

sensitive and specific for the diagnosis of AIDS-related CMV infection of the CNS.

However, conventional CSF findings and neuroimaging may not adequately assess the severity of CNS CMV disease, as demonstrated at autopsy. Magnetic resonance imaging is the diagnostic study of choice in assessing immunocompromised patients suspected of having CNS infection. Imaging may show atrophy; high signal intensity in the periventricular white matter, typically not associated with significant mass effect; and retinitis (frequently seen in the AIDS population) in patients with CMV infection. Although patients with CNS infection may also have ependymal and subependymal involvement, associated imaging findings often are not present. When present, T2W signal abnormality and enhancement along the ependyma are valuable in establishing this diagnosis. Currently, the most common cause of ependymal enhancement in the setting of AIDS is lymphoma.