Progressive multifocal leukoencephalopathy limited to the posterior fossa

A 41 year old ethnic Chinese male with past history of Hepatitis B and liver cirrhosis of 5 years, presented with progressive ataxic gait, motor incoordination, dysarthria and tremor for 2 months. Clinical examination revealed dysarthria, horizontal nystagmus, bilateral dysdiodokynesis and dysmetria, with normal muscle power and bilateral flexor plantar response. There was marked gait ataxia with patient confined to bed. He was diagnosed to have HIV/AIDS with a CD4 count of 29 copies/uL and viral load of 106 copies/mL.

The initial MRI Brain (Figure 1) revealed lesions in the right cerebellar hemisphere extending to the right cerebellar peduncle and pons. These lesions were hypointense on the T1WI and hyperintense on the T2W/FLAIR images. There was no associated mass effect or enhancement. Highly active antiretroviral therapy (HAART) comprising of tenofovir 300 mg od, Combivir (lamivudine/zidovudine 150mg/30mg) one tablet bd and efavirenz 600 mg od was given.

Despite being on the antiretroviral therapy for five weeks, his condition worsened with...
progressive right sided cerebellar symptoms. Repeated MRI showed extension of the right cerebellar lesions with additional lesions seen in the right side of the medulla and the left cerebellar hemisphere (Figures 2 and 3).

Stereotactic guided biopsy of the right cerebellar lesion was performed. The paraffin embedded formalin fixed tissue sample showed scattered bizarre astrocytes in a background of foamy macrophages, lymphocytes, some enlarged
oligodendrocytes with intranuclear inclusions (Figure 4). The immunohistochemistry for the JCV capsid protein VP-1 was positive in the nuclei of the infected oligodendrocytes. JC virus was subsequently identified in the CSF using polymerase chain reaction (PCR).

The patient deteriorated further, became totally dependent and was only able to produce incomprehensible sound. He later succumbed to recurrent chest infections.

**DISCUSSION**

The diagnosis of PML in our patient with underlying HIV/AIDS is confirmed by the histology and presence of JC virus PCR in CSF. It is impossible to exclude coexistent primary CNS lymphoma or infiltrative astrocytoma. However, previous study has shown that primary brain tumors rarely co-exists with PML.3

MRI is a sensitive modality of early lesion detection, determination of the extent and character of the abnormalities. The most frequently affected regions are the cerebral hemispheres at the periventricular and/or subcortical white matter, followed by the cerebellum and brain stem.4 The frontal and parieto-occipital lobes are classically affected. The lesions are typically multifocal, diffuse and asymmetric, involving the subcortical U-fibres giving rise to the scalloped borders along the cortex. These lesions usually demonstrate high signal intensity on T2-weighted/FLAIR sequence.

**Table 1: Clinical and MRI features of PML limited to posterior fossa reported in literature.**

<table>
<thead>
<tr>
<th>Author/year</th>
<th>Age/sex</th>
<th>Location of lesions on MRI</th>
<th>Underlying illness</th>
<th>Clinical manifestations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kastrup, 20025</td>
<td>31/M</td>
<td>Brainstem</td>
<td>HIV</td>
<td>Progressive leg weakness, visual loss, dysarthria</td>
</tr>
<tr>
<td>Matthew, 20046</td>
<td>44/F</td>
<td>Bilateral medulla</td>
<td>HIV</td>
<td>Ataxia, hyperreflexia, limb dysmetria</td>
</tr>
<tr>
<td>Kastrup, 20057</td>
<td>42/M</td>
<td>Cerebellar / brainstem</td>
<td>HIV</td>
<td>Dysarthria, limb dysmetria, gait instability</td>
</tr>
<tr>
<td>Svensen, 20088</td>
<td>51/M</td>
<td>Cerebellar / brainstem</td>
<td>SLE</td>
<td>Hemiparesis, dysarthria, diplopia, head tremor</td>
</tr>
<tr>
<td>Rahmat, 2010</td>
<td>41/M</td>
<td>Cerebellar / brainstem</td>
<td>HIV</td>
<td>Ataxia, motor incoordination, dysarthria</td>
</tr>
</tbody>
</table>
and hypointense on T1-weighted images. Contrast enhancement is usually not observed and mass effect is uncommon.4

Our patient shows unusual white matter abnormality limited to the posterior fossa, with no clinical symptom elsewhere in the brain. This unusual selective infratentorial involvement has been previously reported in a small number of cases, shown in Table 1.5–8

Our patient thus further confirms that PML may present with isolated posterior fossa lesions.

REFERENCES