**Introduction**

The one-and-a-half (OAAH) syndrome is characterized by lateral gaze palsy in one direction (“one”) and internuclear ophthalmoplegia (INO) in the other (“and a half”) [1]. This rare syndrome was described 30 years ago by Miller-Fisher [2] and is due to a unilateral lesion of the dorsal pontine tegmentum, involving the ipsilateral paramedian pontine reticular formation, internuclear fibres of the ipsilateral medial longitudinal fasciculus and, usually, the abducens nucleus. The main causes of this rare syndrome are stroke and multiple sclerosis. Few cases have been reported since the introduction of MRI. Our aim was to examine clinicoradiological correlations in six patients with a one-and-a-half syndrome due to a stroke. Ophthalmological symptoms were diplopia, oscillopsia or blurred vision. Four patients had an associated facial nerve palsy, three a hemiparesis and one a unilateral hemihypoesthesia. MRI revealed an infarct in the pons in all patients. The cause of the infarct was a basilar artery dissection in one patient, bilateral vertebral artery dissection in a second and unknown in the other four. All patients recovered within 2 days to 8 weeks. This study showed a good correlation between the site of the lesion (superior, inferior or extensive pontine ischaemia) and clinical deficits.

**Abstract**

The one-and-a-half syndrome is characterised by a lateral gaze palsy in one direction and internuclear ophthalmoplegia in the other. It is due to a unilateral lesion of the dorsal pontine tegmentum, involving the ipsilateral paramedian pontine reticular formation, internuclear fibres of the ipsilateral medical longitudinal fasciculus and, usually, the abducens nucleus. The main causes of this rare syndrome are stroke and multiple sclerosis. Few cases have been reported since the introduction of MRI. Our aim was to examine clinicoradiological correlations in six patients with a one-and-a-half syndrome due to a stroke. Ophthalmological symptoms were diplopia, oscillopsia or blurred vision. Four patients had an associated facial nerve palsy, three a hemiparesis and one a unilateral hemihypoesthesia. MRI revealed an infarct in the pons in all patients. The cause of the infarct was a basilar artery dissection in one patient, bilateral vertebral artery dissection in a second and unknown in the other four. All patients recovered within 2 days to 8 weeks. This study showed a good correlation between the site of the lesion (superior, inferior or extensive pontine ischaemia) and clinical deficits.

**Key words**

One-and-a-half syndrome · Infarcts pontine · Magnetic resonance imaging

**Patients and methods**

Over a 2-year period six patients, three men and three women with a mean age of 57 years (range 39–82 years) were admitted to our stroke unit for sudden diplopia caused by AAH syndrome. Vascular risk factors, association with other neurological deficits (motor or sensory deficits, cranial nerve palsies), ocular symptoms at the onset, side of gaze palsy and INO, presence of nystagmus, extroopia, convergence and horizontal oculocephalic reflex (OCR) preservation were recorded for all patients.

MRI was performed at 1.5 T with T1- and T2-weighted 5-mm sections in sagittal, coronal and axial planes. The site of the infarct was determined using Tatu’s templates [11]. All patients underwent complete diagnostic investigation with biological tests, ECG and Holter testing, ultrasound (cervical artery duplex Doppler sonography plus transthoracic or transoesophageal echocardiography), and conventional angiography was performed in four [12].
Table 1 Ocular motor deficits and clinical data

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Ocular symptoms</th>
<th>Oculo-cephalic reflex</th>
<th>Nystagmus</th>
<th>Convergence</th>
<th>Exotropia</th>
<th>Gaze palsy</th>
<th>Inter-nuclear ophthalmoplegia</th>
<th>Nerve palsy</th>
<th>Motor deficit</th>
<th>Sensory deficit</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>62</td>
<td>F</td>
<td>Diplopia, oscillopsia</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>Left</td>
<td>None</td>
<td>Right</td>
<td>VII</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>82</td>
<td>M</td>
<td>Diplopia</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>Right</td>
<td>–</td>
<td>Left</td>
<td>–</td>
<td>Left</td>
</tr>
<tr>
<td>3</td>
<td>70</td>
<td>M</td>
<td>Diplopia</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Left</td>
<td>Right</td>
<td>Right</td>
<td>None</td>
<td>Right</td>
</tr>
<tr>
<td>4</td>
<td>46</td>
<td>F</td>
<td>Blurred vision, diplopia</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Left</td>
<td>Right</td>
<td>Right</td>
<td>Right</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>39</td>
<td>M</td>
<td>Diplopia</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Right</td>
<td>–</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>6</td>
<td>41</td>
<td>F</td>
<td>Diplopia</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Right</td>
<td>Left</td>
<td>Left</td>
<td>V, VII</td>
<td>None</td>
</tr>
</tbody>
</table>
Results

The main features are shown in Table 1. In five patients, MRI revealed low signal on T1- and high signal on T2-weighted images (Figs. 1–5) suggesting an infarct. In patient 6, CT revealed a dense brain-stem lesion, with high signal on T2-weighted images, suggesting haemorrhagic infarct (Fig. 6). Angiography revealed bilateral vertebral artery dissection.

In patients 2 and 5, the infarct was limited to the ventral and tegmental pons. The lesions probably involved only the anterosuperior part of the anteromedial group of arteries arising from the basilar artery, and the arteries of the interpeduncular fossa. Lesions were more prominent in the anterior part of the pons, leading to corticospinal impairment with contralateral weakness. The abducens nucleus was probably normal. In patient 3, we observed an infarct in the lower part of the pons (Fig. 3), involving the abducens and facial nerve nuclei. We presume to have arise from occlusion of the artery of the foramen caecum, a branch of the basilar artery. In the remaining patients the lesions were larger and involved the whole pons (Figs. 1, 4, 6).

An OAAH syndrome was associated with other neurological deficits in all patients: contralateral motor in three, and sensory in one, a facial nerve palsy in four and facial numbness in one. Ophthalmological symptoms consisted of diplopia in all patients, oscillopsia in two and blurred vision in one. There was no exotropia; nystagmus was present in four patients, but vertical eye movements were preserved in all cases. Convergence and horizontal OCR were impaired in four patients and preserved in the other two, OCR contralateral to the lesion. Four patients had vascular risk factors (cases 1, 2, 3 and 5) but the cause of stroke was determined in only two: basilar artery dissection in patient 4 (Fig. 4a) and bilateral vertebral artery dissection in patient 6. The patients with dissection were treated by heparin then warfarin, and their arteries were recanalised 3 months later. Aspirin (300 mg/day) was started in the other four patients. The outcome was good in all cases, with total recovery of the visual symptoms within 2 months or less.

Discussion

The lesion associated with the OAAH syndrome was a pontine infarct in all patients. Since 1983, a few cases with MRI study have been reported [4–10]. OAAH syndrome occurs in only 8% of paramedian pontine infarcts [13]. Pontine lesions may lead to four types of lateral eye movement disturbance [6]: INO, pontine reticular formation syndrome, abducens nucleus syndrome and, in combination with the first two or three of these, the so-called one-and-a-half syndrome. The causative infarcts are usually due to occlusion of paramedian perforating branches of the basilar artery [14]. Correlation between MRI, clinical findings and arterial territories, confirmed that when infarcts are limited to the upper ventral and tegmental pons, the abducens nuclei are preserved. In these cases, as in patients 2 and 5, OAAH is due to the association of INO and pontine reticular formation impairment. OCR were spared be-

Fig. 5 Case 5. T2-weighted axial image plane high signal in the antero-medial part of the pons

Fig. 6a, b Case 6. An axial and sagittal T2-weighted images shows widespread high signal in the posterior part of the pons
cause of the preservation of the abducens nuclei and the inferior nerve fibers [3, 15]. OCR are spared when the infarct is limited to the pontine reticular formation and abolished when the final neural pathway (abducens nuclei and medial longitudinal fasciculus) is concerned [11]. However, even when limited, infarcts in the inferior or part of the pons induce abducens lesions. In patients with an inferior (case 3) or extensive pontine infarcts (case 1, 4 and 6), OCR and convergence were abolished, probably because of a lesion of the abducens nuclei and medial longitudinal fasciculus [15].

In previous studies [3], visual symptoms also consisted of horizontal diplopia (60%), blurred vision (40%) and oscillopsia (20%) [3]. Isolated OAAH syndromes are rare, having been also reported with small haemorrhages and with vascular malformations [10, 16]. The main causes of OAAH are brain-stem infarcts and multiple sclerosis [3], in which the syndrome is usually associated with other neurologic deficits [3]: cranial nerve palsy (facial in 75%), hemiplegia (30%) or hemihypeaesthesia (35%) [3]. An association with a cheiro-orbital syndrome was also recently reported [17]. Our six cases are similar to those in the literature as regards the visual and associated deficits. Vascular risk factors are frequent, as in four of our patients, but the direct cause of ischaemia is rarely found [3]. To our knowledge, basilar artery dissection has not previously been reported with an OAAH syndrome. The outcome of the OAAH syndrome is usually good, with recovery in a few days or weeks, without sequelae. The site of the infarct, at the border between the reticular formation and medial longitudinal fasciculus might explain the good, early recovery [11]. Oculopalatal myoclonus sometimes occurs, up to 3 years after the OAAH syndrome, especially when there was a facial nerve palsy [18].

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References