LETTERS TO THE EDITOR

A case of Pourfour Du Petit syndrome following parotidectomy

Paralysis of the cervical sympathetic nerves causes an ipsilateral Horner’s syndrome and this is a well recognised phenomenon in clinical neurology. However, the opposite hypersympathetic state following damage to the cervical sympathetic nerves is seldom recognised. We present a case report of such a condition which has been termed the Pourfour Du Petit syndrome.

A 41 year old man had a left parotidectomy for mixed parotid tumour. After the operation he noticed left facial weakness which was of lower motor neuron type. This resolved within three months. He also noticed immediately postoperatively that his left pupil was larger than the right. Initially he had some blurring of vision related to this pupillary dilatation but this completely resolved. However, the pupillary abnormality continued until the time of neurological examination six months later. Examination revealed lid retraction on the left side and a left pupil which when measured in a shaded room was two millimetres larger than the right (see fig). Both pupils responded briskly to light and accommodation; there was full eye movement and no double vision. There was no abnormal sweating response. The rest of the neurological examination was normal.

In conclusion, his signs were of autonomic overactivity to the left eye and this was thought to be due to damage to the sympathetic plexus which ascends around the carotid artery. This could have occurred during the time of surgery. An alternative explanation could be that there was a right-sided abnormality with relative paresis and miosis of the pupil as in Horner’s syndrome. This was thought to be untenable as clearly the surgery was on the left side. Pourfour Du Petit, a French physician, was the first to note signs of increased sympathetic activity in the eyes and upper limb and relate these to cervical sympathetic chain injuries. These were largely caused by sword wounds on the neck inflicted during the Napoleonic war. Further cases have been reported after direct non-penetrating injury to the cervical sympathetic chain and brachial plexus.

We think our case is the first to have been reported following an operation. We suspect that this syndrome is often unrecognised but should be considered as a possible cause of pupillary dilatation and lid retraction.

PAUL BYRNE
CHRISTOPHER CLOUGH
The Brook General Hospital, London, United Kingdom

Hypothermia in a mesodiencephalic haematoma

Hypothermia is defined by rectal temperatures of 35°C or less. The most frequent causes are prolonged exposure to cold especially in the elderly, immersion in cold water, prolonged immobilisation, mountain eering, intoxication (barbiturate, phenothiazine, carbon monoxide, alcohol), hypothyroidism and hypopituitarism, sepsis, ketaedosis and hypoglycaemia, and uraemia.1

This symptom is rarely reported in patients with well defined diseases of the central nervous system. Hypothermia may be continuous, as in various hypothalamic disorders (neoplasms, inflammatory or degenerative lesions) or, unusually, episodic as in “diencephalic autonomic epilepsy”, or agenesis of the corpus callosum (Shapiro’s syndrome).2 Several recent reports emphasise the relative frequency of hypothermia during Wernicke’s encephalopathy and this can be rapidly reversed by thiamine treatment.3

In the case we report, moderate hypothermia with ocular motor disturbances were associated with a haematoma of the mesodiencephalic junction.

A 50 year old man was admitted with a sudden onset of apathy, general malaise and nausea which he experienced when taking a walk. His past history included five years of hypertension, the treatment of which had been interrupted one week earlier. On admission, he was alert and well oriented. Blood pressure was 240/120 mm Hg and pulse rate 75. Respiration was regular. Rectal temperature was 35°C, at repeated intervals. There was moderate motor neglect and French physician, was the first to note signs of increased sympathetic activity in the eyes and upper limb. Visual fields and pupils were normal. Vertical eye movements on command were not possible, both upwards and downwards. During forced elevation, nystagmus retractorius was observed. Vertical oculocephalic reflexes were normal, as were all horizontal eye movements. No other abnormalities were detected.

Despite the absence of evidence of infectious disease, blood cultures were made, but no bacterial growth was observed. Serum sodium was 142 mmol/l, glucose 7-3 mmol/l, and creatinine 92 mol/l. Full blood count was normal.

Computed tomography of the brain without contrast revealed a right haematoma in the posterior mesodiencephalon, with a discrete mass effect on median structures, and traces of blood in the adjacent lateral ventricles. Magnetic resonance imaging (MRI) gave more precise localisation of the lesion (fig). The upper edge of the haematoma reached the pulvinar, while its lower limits only skimmed the red nucleus. Anteriorly, it did not cross the rostro-caudal axis of the brainstem, and was clearly posterior to the hypothalamus.

Hypothermia remained at 35°C for three days. Body temperature then rose slowly to 37°C. 10 days after admission. At that time, recording of eye movements showed full recovery of downward saccades and smooth pursuit, whereas upward saccade velocity remained extremely slow. The patient, when seen at follow up two months later, had completely recovered.

The physiology of thermal regulation by the central nervous system is not well understood. Experimental data from animals and pathological observations of human brain show that the hypothalamus plays a crucial role.1 Two types of neurons are found in the anterior hypothalamic nuclei and in the preoptic area: some respond to increases in blood temperature by releasing noradrenaline, while others respond to decreases in temperature by releasing serotonin.1

The former are more numerous, indicating that the anterior hypothalamus is particularly involved in heat dissipation. Anterior nuclei are connected by efferent pathways to the caudal part of the hypothalamus that is insensitive to temperature variations but essential for heat maintenance by regulating thermogenesis.2 The hypothalamus is connected to the thermosensitive areas of the brainstem, especially the lateral parts of the medulla oblongata, by several pathways. Fibres sensitive to cold would seem to be distinct from those sensitive to heat, the latter being redundant.3

Anatomical correlations are difficult to establish in human pathology since the lesions are usually large compared to the hypothalamic nuclei. Furthermore, one cannot exclude a remote mass effect. In our patient, the posterior hypothalamic region seemed intact on the MRI. Indeed, the mammillary bodies, located under the posterior
hypothalamus, are clearly anterior to the limits of the haematoa (fig). Moreover, the absence of weakness suggests that the internal capsule, located between the haematoa and the hypothalamus, is unaffected. Hypothermia might therefore be ascribed to a damage to the efferent pathways from the hypothalamus to the brainstem.

Bilateral lesions of the hypothalamus seem necessary to produce thermic dysregulation, whereas unilateral damage to the medulla oblongata appears to be sufficient.1 In our case, the initial downward gaze palsy indicates a bilateral involvement of the upper mesencephalon.1 This can be compared to Wernicke’s encephalopathy where hypothermia may occur, and where the mesodiencephalic lesions, although more extended rostro-caudally, are bilateral.8

To our knowledge, no previous observation of hypothermia related to a mesodiencephalic lesion has previously been reported. Its rarity might be explained by the need for bilateral lesions, and the fact that hypothermia may go unnoticed.


de expected to induce sufficient hypothermia to permit the use of traditional hyperosmotic solutions. Treatment with hypothermia must be rapid and reliable following subcutaneous injection. We have sought alternative, more convenient routes of delivery for the drug. Effective mucosal absorption has been reported1 and we now describe the use of apomorphine delivered intranasally.

Eight patients with Parkinson’s disease were studied. Their mean age was 58–1 years (48–70), duration of disease 12–9 years (5–22) and length of treatment with levodopa 11–8 years (4–19). All had severe on-off fluctuations which had already been shown to respond to intermittent subcutaneous injections of apomorphine.

Patients were assessed in an off period following withdrawal of usual medication using a modified Webster rating scale,2 timed walking over a 12 metre course and unilateral hand-tapping for 30s on digital counters mounted 20 centimetres apart. Apomorphine solution (10mg/ml) was administered, intranasally, using a metered-dose device and the above assessments were repeated at regular intervals until motor performance returned to baseline.

In seven cases there was a prompt response to 6mg (0-6ml) intranasal apomorphine. No adverse effects were noted and the nasal spray caused no local irritation. In these seven patients the mean Webster scores improved from 21–8 to 10–9; walking times from 17–3 to 9–0 seconds; and tapping counts from 29 to 50. The onset of the motor response occurred after a mean interval of 8–9 minutes (6–15) and persisted for a mean duration of 44 minutes (36–55). Similar times were reported by the patients for subcutaneous use and equivalent improvements in the measures of Parkinsonism were also seen with both methods of administration. The onset and duration of response appeared to correlate with blood levels of apomorphine measured in one case (fig). The intranasal dose was between 1–5 and twice the subcutaneous one. The patient who did not respond to 6mg did so following a larger dose of 8mg. She had a long history of nasal congestion, perhaps hindering the mucosal absorption of apomorphine.

This preliminary study suggests that intranasal delivery may offer an effective alternative to subcutaneous injection of apomorphine. The benefits of the latter, including the speed and quality of motor response, appear to be retained in most cases with this simpler technique, prompting further evaluation of its long-term use.

Intranasal apomorphine: a new treatment in Parkinson’s disease

Apomorphine, a directly acting dopamine agonist, has recently been used in the treatment of Parkinson’s disease complicated by motor fluctuations. Benefit is seen more rapidly and reliably following subcutaneous injection. We have sought alternative, more convenient routes of delivery for the drug. Effective mucosal absorption has been reported1 and we now describe the use of apomorphine delivered intranasally.

Eight patients with Parkinson’s disease were studied. Their mean age was 58–1 years (48–70), duration of disease 12–9 years (5–22) and length of treatment with levodopa 11–8

Figure The motor response and plasma concentration following single 6mg intranasal administration of apomorphine.

Somatostatin in cerebrospinal fluid after generalised convulsions or cerebral infarction in humans

A role for somatostatin in the generation of epileptic seizures is discussed as increased concentrations of this peptide in epileptic foci have been reported.6 In the cerebrospinal fluid (CSF) of rats increased levels of somatostatin-like immunoreactivity (SIR) were found following ethylbenzotetrazol-induced convulsions.7 The same group of investigators, however, was unable to demonstrate a change of CSF SIR levels in nine epileptic patients presenting with generalised convulsions.8

We have measured SIR by specific radioimmunoassay9 in the CSF of 16 patients with epilepsy (table). Of these, eight patients were treated with phenytoin, two with phenytoin and phenobarbital, two with carbamazepine and primidone, one with bromazepam. The patients received no anti-convulsant drugs. There were no significant differences or trends in SIR levels apparent when patients were grouped according to their drug treatment.

In six patients generalised convulsions preceded the lumbar puncture by periods of several hours to three days. Their levels were compared with those of epileptic patients without recent seizures, of control patients without proven neurological diseases and of patients with cardiovascular disease (CVD) and cerebral infarction. The mean level of the epileptic patients in CVD and cerebral infarction was not significantly different from that of the control patient group and that of the group of patients with

Table Clinical data and SIR levels

<table>
<thead>
<tr>
<th>Group</th>
<th>n</th>
<th>Age mean (SD)</th>
<th>Sex (female/male)</th>
<th>SIR mean (SD) pmol/l</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control patients</td>
<td>27</td>
<td>35 (3)</td>
<td>10/17</td>
<td>113 (9) (6-3)</td>
</tr>
<tr>
<td>Epileptic patients</td>
<td>16</td>
<td>46 (8)</td>
<td>6/10</td>
<td>113 (5) (6-3)</td>
</tr>
<tr>
<td>Subgroup with recent convulsion 6</td>
<td>52 (8)</td>
<td>2/4</td>
<td></td>
<td>166-12 (12)*</td>
</tr>
<tr>
<td>Cerebrovascular disease</td>
<td>20</td>
<td>51 (8)</td>
<td>5/15</td>
<td>125 (5) (11-3)</td>
</tr>
<tr>
<td>Subgroup with recent infarction 5</td>
<td>54 (6)</td>
<td>1/4</td>
<td></td>
<td>164-21 (21)**</td>
</tr>
</tbody>
</table>

*significantly different from epileptic patients (p < 0.01, Wilcoxon test)
**significantly different from cerebrovascular disease (p < 0.01)