LETTERS TO
THE EDITOR

Episodic paroxysmal hemianasia responsive to calcium channel blockers

Based on the paucity of published reports,1-6 a recent paper on episodic paroxysmal hemianasia (EPH) emphasises the rarity of this headache syndrome and its particular sensitivity to indomethacin.1 Our experience with this syndrome is that it may be more common than previously stated. In fact, throughout the last year we were able to identify six cases in two different general hospitals, which represents 1-5% of the total number of patients with vascular type headaches seen. These two hospitals were located in two different environments, suggesting that this high prevalence was not related to geographical, economical, or social factors.

The table illustrates the clinical features of our patients. All of them fulfilled the clinical criteria of EPH.7 A further interesting feature of our patients was the dramatic response to calcium channel blockers (CCB) (case 1 to nicardipine and cases 2-6 to flunarizine). After a follow up period of six months all patients noticed complete relief of their complaints. Two of them were initially treated with indomethacin with good results, but this drug had to be withdrawn because of disabling gastrointestinal symptoms. In contrast, CCB were well tolerated by all patients and they showed similar efficacy to indomethacin. Therefore, CCB have become the first choice treatment in our outpatient clinics.

The responsiveness of our cases of EPH to CCB suggest that this syndrome may be related pathophysiologically to episodic cluster headaches and to other vascular type headaches which also respond to these drugs.7,8

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Table Clinical features of patients with EPH (scheme adapted from Blau and Engel for comparison)

<table>
<thead>
<tr>
<th>Patient number</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>F</td>
<td>F</td>
<td>F</td>
<td>M</td>
<td>F</td>
<td>M</td>
</tr>
<tr>
<td>Age (years)</td>
<td>48</td>
<td>23</td>
<td>35</td>
<td>37</td>
<td>25</td>
<td>42</td>
</tr>
<tr>
<td>Age at onset (years)</td>
<td>44</td>
<td>19</td>
<td>33</td>
<td>25</td>
<td>23</td>
<td>20</td>
</tr>
<tr>
<td>Hemicrania</td>
<td>L</td>
<td>R</td>
<td>L</td>
<td>R</td>
<td>L</td>
<td>L</td>
</tr>
<tr>
<td>Horner syndrome</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Continuous injection</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Episphora</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Rinoesthesia</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Duration of episodes (weeks)</td>
<td>3</td>
<td>1</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Duration of remission (weeks)</td>
<td>3</td>
<td>2</td>
<td>12</td>
<td>2</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>Minimum frequency (attacks/day)</td>
<td>5</td>
<td>5</td>
<td>15</td>
<td>6</td>
<td>15</td>
<td>10</td>
</tr>
<tr>
<td>Maximum frequency (attacks/day)</td>
<td>10</td>
<td>10</td>
<td>15</td>
<td>6</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>Duration of attacks (min)</td>
<td>15</td>
<td>15</td>
<td>15</td>
<td>5</td>
<td>15</td>
<td>15</td>
</tr>
<tr>
<td>Efficiency of indomethacin</td>
<td>+++</td>
<td>+</td>
<td>++</td>
<td>++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Efficiency of CCB</td>
<td>+++</td>
<td>+</td>
<td>+++</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

Nu = not used.

Efficacy of cyclophosphamide in sarcoid radiculoneuritis

Paraplegia due to neurosarcoidosis is unusual and its treatment not well defined. We report a case of a sarcoid radiculoneuritis which benefited from immunosuppressive therapy with cyclophosphamide.

A 46 year old man was admitted in 1981 for hand tremor, bilateral cranial nerve VIIIth dysfunction and progressive weakness of the legs. Neurological examination showed depressed reflexes in the legs, plantar responses were extensor. Motor nerve conduction velocities were normal. CSF examination revealed an aseptic meningitis with 100% mononuclear cells (table). A chest x ray photograph showed bilateral hilar adenopathy, and a histological diagnosis of sarcoidosis was made after transbronchial biopsy. Treatment of 1500 mg methyl-prednisolone was given daily, for three days. Substantial recovery of strength was obtained in the legs. The patient remained stable for the next four years while treated with oral corticosteroids (prednisone, 10-40 mg/day).

In 1985 paraparesis with sphincter disturbance recurred. CSF examination showed a persistent meningitis (table). CT scan was normal. Treatment with methyl-prednisolone was repeated and stability was achieved again.

Two months later he developed complete flaccid paraplegia with areflexia, bilateral extensor plantar responses, and total incontinence. Vibration sense and pain sensitivity were impaired below the T10 level. There was proximal weakness in the arms with bilateral areflexia. The patient declined a magnetic resonance scan. Methyl-prednisolone infusions and four intrathecal steroid injections failed to improve his clinical state. A single infusion of 1000 mg of cyclophosphamide was administered five weeks after the paraplegia developed. Within two weeks the weakness in the arms had recovered, all the reflexes were present except those in the ankles, but he had recovered some movement in the lower leg. Two further cyclophosphamide infusions at three week intervals led to complete sensory and incomplete sphincter recovery. During the next six months three more cyclophosphamide infusions were administered and continuing neurological improvement was noted: walking with an aid became possible and the patient complained only of mild sphincter disturbance. CSF analysis two and six months after starting cyclophosphamide are shown in the table. Over the next three years the patient received small doses of oral corticosteroids (10-20 mg/day) and remained neurologically stable.

Although the peripheral neuropathy or radiculoneuropathy could have explained the flaccid presentation of the paraplegia, the sphincter disturbance, the extensor plantar responses, and the T10 sensory level clearly indicated spinal cord involvement. Signs in the arms suggested an associated radiculopathy. Spinal cord sarcoidosis has been described in fewer than two cases of neurosarcoidosis,5,6 but its exact incidence is difficult to assess as spinal involvement may remain clinically silent.4 Various lesions have been described including extra- and intradural granulomas developing as compressive masses and infiltrative granulomatous nodules usually associated with arachnoiditis.10 To ascertain spinal cord involvement in sarcoidosis has proved to be difficult.4,5 Myelography has shown the spinal cord to be swollen, thinned, or normal.4 The value of MRI remains to be shown. Despite the absence of controlled trials, corticosteroids have been widely recommended in neurosarcoidosis with considerable results.7 Alternative treatments are few. Radiotherapy has been occasionally proposed and some improvement noted in intracranial compressive granulomas or in

Table CSF data from patient with sarcoid radiculoneuritis

<table>
<thead>
<tr>
<th>Time of CSF examination</th>
<th>Call count (WBC/mm³)</th>
<th>Protein content (mg/l)</th>
</tr>
</thead>
<tbody>
<tr>
<td>At first presentation (1981)</td>
<td>23</td>
<td>1080</td>
</tr>
<tr>
<td>At recurrence of symptoms (1985)</td>
<td>37</td>
<td>2500</td>
</tr>
<tr>
<td>Two months after start of cyclophosphamide treatment</td>
<td>11</td>
<td>2280</td>
</tr>
<tr>
<td>Six months after start of cyclophosphamide treatment</td>
<td>1</td>
<td>1400</td>
</tr>
</tbody>
</table>

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sarcoid meningitis with cranial nerve involvement.\(^4\) Transient benefit from chlorambucil in a case of sarcoid meningoencephalitis has been reported.\(^4\) In our case, high dose intravenous cyclophosphamide, at doses energetic enough in multiple sclerosis,\(^1\) quickly and dramatically improved the clinical picture. Although spontaneous remission cannot be excluded, it seems reasonable to assume a causal relation between introduction of treatment and the clinical improvement. We suggest that cyclophosphamide should be considered in cases of severe neurosarcoidosis when steroids are unsuitable or ineffective.

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Unsuspected meningioma presenting as a subdural haematoma

Symptomatic haemorrhages associated with meningiomas are rare and most are subarach- noid in location.\(^1\) Subdural haemorrhages are seldom caused by a meningioma.\(^2\) We report a case of a subdural haematoma in a patient with a long-term and intractable treatment for chronic atrial fibrillation. A menin- gioma was unsuspected preoperatively and intraoperatively. Only after pathological examination of the blood clot was the tumour discovered.

A 79 year old, right handed white man who had been receiving Coumadin anticoagula- tion therapy for chronic atrial fibrillation for two years was admitted to this medical centre. He complained of headaches over the left side of his head for the previous three days. He had difficulties with word finding but no focal weakness. He had a personal and family history of a recent injury. The patient had suffered a brain stem cerebrovascular accident 20 years previously and had residual mild numbness of his right side and weakness in his left leg. Examination showed the patient to be awake, alert, and fully oriented. His pupils were equal, round, and reactive to light and there was no papilloedema. His cranial nerve system was intact apart from a decrease in visual acuity in the right eye. Muscle power was normal except for a mild weakness in the proximal part of his left leg. Sensory examination showed a slight decrease to pinprick sensation over the right side of his body.

Computed tomography (CT) showed a large left sided subdural haematoma with edemous and hypodense components. Coagulation studies showed a pro-time of 13-8 seconds (control 11-5 s) and a prothrombin time of 42-8 seconds (control 26-4 s). He was given fresh frozen plasma to correct this coagulopathy and abnormality. Postoperatively, he underwent a left frontal temporal parietal craniotomy with evacuation of the subdural haematoma. The haematoma was large, extending from the temporal floor to the falx cerebri and from the lateral to the frontal pole. It appeared to be a typical subdural haematoma. There were no apparent soft tissue components, nor was the haematoma attached to the leptomeninges. After complete removal of the clot, the cerebrospinal fluid was exposed subdural space was examined. There was no mass on the undersurface of the dura nor on the cortical surface of the brain, and a source of the haematoma could not be identified. Postoperatively, the patient was lethargic and had severe expressive dys- phasia. He obeyed commands well with equal strength in all four limbs. Further CT did not show reaccumulation of the subdural haematoma or evidence of a cerebrovascular accident. The patient’s neurological condi- tion gradually improved, and two weeks after operation he was discharged home. MRI scans, with and without gadolinium, were performed at six months and one year later but did not indicate any residual mass lesion. All surgical specimens are routinely sub- mitted for pathological analysis at this centre. Gross pathological examination of formalin- fixed clot specimens showed clotted blood with several pale foci but no distinct tumour nodules. Microscopic examination showed relatively fresh clotted blood interspersed with fragments of tumour. Some areas of the tissue clearly showed the pattern of a menin- gioma, with whorls of meningothelial cells and meninomann bodies. Occasionally, mitotic figures were identified, and there was extensive necrosis. Some of the necrotic tissue was basophilic and contained psammoma bodies; this was likely to be necrotic tumour. In other areas, the cells were spindle shaped, and the tissue contained numerous small, vascular channels. Some of the vascular areas were clearly in tumour while others were composed of dilated channels.

Benign tumours such as meningiomas are rarely associated with massive intracranial bleeds. Helle and Conley found only 43 cases of meningioma associated with haemorrhage. Of these, only four haemorrhages were strictly in the subdural space, while five were
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*J Neural Neurosurg Psychiatry* 1992 55: 166-167
doi: 10.1136/jnnp.55.2.166-a

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