Matters arising

In the case which I described there is a striking resemblance to CPH in the pre-CPH stage but there has been no transition from episodic to chronic stage. It is possible that this transition may yet occur but as the attacks have been exclusively episodic for 34 years, I labelled the case "indomethacin-responsive episodic cluster headache" rather than "pre-chronic paroxysmal hemicrania". In this way I hoped to draw attention to the possibility that cases of cluster headache of an episodic variety, in addition to cases of CPH, may be dramatically responsive to indomethacin if the headaches are very frequent and of short duration.

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Spontaneous ear pain as the initial present-ing manifestation of giant cell arteritis

Sir: I read with interest the article by Friedman and associates "Neglected conditions producing preauricular and referred pain".¹ It is an excellent review. No such review can expect to be exhaustive. However, I believe one important differential diagnosis was omitted and I use the following case to exemplify it.

An 82-year-old woman complained of pain in front of her left ear. That morning while reading in bed she "felt a pop" and then a severe pain in the left preauricular area. Severe pain and some lightheadedness persisted for over an hour. She reported having had intermittent episodes of lightheadedness and intermittent similar pains the previous week. They were unassociated with chewing or with talking. Between the severe attacks of pain there persisted a light ache in and in front of the left ear. Her roommate reported that she was becoming listless and apathetic over the past few weeks and had lost most of her interest in her favorite activities including cooking. She had lost weight over the previous month which she attributed to pain on swallowing.

A Westgren erythrocyte sedimentation rate was 53 mm/hr. A left temporal artery biopsy was positive for giant cell arteritis. Treatment with systemic corticosteroids

resulted in prompt and complete remission of the pain. At no time before or during treatment was there tenderness or swelling in the ear or preauricular area.

This patient demonstrates that ear and preauricular pain can be an important initial complaint of patients with giant cell arteritis.² Early recognition of this symptom is important in order that early diagnosis may prevent blindness. Such patients frequently present to the dentist or neurologist.³ The aetiology of the pain is variable. In some such patients we have studied the pain has been related to prolonged masticatory activity thereby implicating muscle ischaemia. However this patient had ear pain without mastication. The pain may have been brainstem in origin. The lateral medullary syndrome is the most common central nervous system syndrome in patients with giant cell arteritis.³ Incomplete forms of the syndrome are common and may present purely as facial pain and in some cases simply as ear pain.

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Behaviour disturbances during recovery from herpes simplex encephalitis

Sir: The treatment team of The Kemsley Unit (a unit for the treatment and rehabilitation of individuals with severe behaviour disorder resulting from brain injury) were particularly interested in the paper of Greenwood *et al* (*J Neurol Neurosurg Psychiatry* 46, 809-17) since they immediately recognised two of the individuals from their case reports as patients who had been referred to the unit.

Neither patient had in fact been treated in the unit, because funding was unavailable. However, shortly after the paper appeared (but unconnected with this event) the first patient (Case 1, JBR) was re-referred with a view to a one month stay, partly to give his parents "holiday

relief" and partly for assessment of potential for rehabilitation. It may be of interest, therefore, to record some of the follow-up information, and also to mention the results of the brief application of behaviour modification methods to the behaviour disorder.

In the last paragraph of the case report, mention is made of his "present state", perhaps the two most impressive aspects being his ability to go out cycling and follow instructions home if lost, and the fact that "attempts at rehabilitating him further have been frustrated by uncontrollable outbursts of rage and aggression". It transpires that it is in fact some two years since he went out cycling, the reason being the continuation and, indeed, extension of explosive outbursts, appearing mainly on "frustration" but also apparently spontaneously. Indeed, he had a period of admission to a psychiatric unit under the Mental Health Act, earlier this year, because of his aggression. His memory disturbance has continued to be severe, but he has gradually learned a wide array of "factual information" (for example, recurring events in his weekly programme), but shows no sign of episodic memory (for example, he never remembers actual occurrences of these recurring events).

He has remained grossly obsessively ritualistic. He has had no further major fits for some 2½ years but continued to have occasional partial seizures (altered consciousness, stereotyped movements and utterances, lasting some hours at a time) until carbamazepine was added to sodium valproate in June, 1983. Because of the behavioural difficulties, he has had haloperidol and for the past six months, chlorpromazine 100 mg, qds, though there was no definite evidence on whether or not this gave any benefit.

Physical and neurological examinations on admission were normal except for a left homonymous field defect in which the upper temporal quadrant was less affected than the lower, neither being fully lost. Neuropsychologically, he showed evidence of mild but varying disturbances of language, principally nominal dysphasia with specific dysphasias (colours, parts of body and so on), and a subtle comprehension deficit, together with marked dyslexia, dysgraphia and dysorthographia. Memory showed severe limitation in all areas, but he performed distinctly more poorly in formal test settings than in day-to-day settings (for example Digit Span was five forwards, three backwards; but he scored a consistent good average of nine on Simon).

During the course of a one month stay, he learnt and used spontaneously the names of two staff members, and learnt about half of the geography of the unit. His behaviour was frequently disrupted by three things: his memory disorder, his obsessional rituals (dominated by his need to record in his "diary" every little event "before I forget"), and explosive outbursts, mostly but not exclusively in response to frustration.

On admission, chlorpromazine was stopped, carbamazepine was increased to 400 mg bd, and epilim stopped. He was also given clobazam 20 mg on. During his first week, his behaviour was actively uncooperative, aggressive and "depressed" (as was his wont at home, he frequently asked for "suicide pills"). All episodes of outburst behaviour led contingently to five-minute periods of "time out" (in a Time Out Room). Interruption of sessions by ritual was prevented. All inappropriate utterances ("suicide pill", and also "All I need is a driving licence") were timed on the spot (in other words, ignored).

During the remaining three weeks, all of his behaviours improved, and his apparent mood became one of cheerfulness for the most part, with evidence of his enjoying many activities. He could be argumentative at times, but only briefly, and responded to prompting. He was generally very active, and his physically perfect state was exemplified by his extreme skill in swimming. To be more specific on the behavioural side, explosive outbursts occurred with the following frequencies during the four weeks:

37, 4, 1, 0.

(These weekly figures obscure an initial increase over the first few days, and a brief upsurge of three in one day at the end of the second week, and the daily graph is very typical for an extinction process.) Although actual counts were not made, there was not one single request for "suicide pills" during the second two weeks, and only one mention of "driving licence" (on the direct question "What problems do you see yourself having?")

Certainly, this patient has shown solid evidence of good response to behaviour modification. A one-month period is extremely unlikely to be sufficient for these behaviour changes to endure. Nevertheless, the exercise makes it virtually certain that sufficiently prolonged treatment would lead to lasting improvements in behaviour control. The fact that the changes took place in a setting in which he was under

very considerable pressure to co-operate in rehabilitative therapies demonstrates, we believe, that "attempts at rehabilitating him further *need not be* frustrated by uncontrollable outbursts of rage and aggression".

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Greenwood replies:

Dr Eames' comments regarding a good response to behaviour modification in our case 1 (JBR) are well taken. If these techniques are as successful as is claimed there lack of general availability to patients with more or less diffuse brain injury is unfortunate.

Notice

World Society for Stereotactic and Functional Neurosurgery

The Ninth meeting will be held 4-7 July, 1985 in Toronto, Canada. Further information may be received from:

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