LETTERS TO THE EDITOR

Episodic paroxysmal hemiancia responsive to calcium channel blockers

Based on the paucity of published reports, a recent paper on episodic paroxysmal hemiancia (EPH) emphasises the rarity of this headache syndrome and its particular sensitivity to indomethacin. Our experience with a similar 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. 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 Two of them were initially treated with indomethacin with good results, but this drug had to be withdrawn because of disabling gastric symptoms.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 clinic.

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. 1

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Nu = not used.

Efficacy of cyclophosphamide in sarcoid radiculomyelitis

Paraplegia due to neurosarcoidosis is unusual and its treatment not well defined. We report a case of a sarcoid radiculomyelitis which benefited from immunosuppressive therapy with cyclophosphamide. A 46 year old man was admitted in 1981 for hand tremor, bilateral cranial nerve VI, VII, VIII 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 spincter 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 sensibilty were impaired below the T10 level. There was proximal weakness in the arms with spastic reflexes. 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 corticosteroid (prednisone, 10-20 mg/day) and remained neurologically stable.

Although the peripheral neuropathy or radiculoneuropathy could have explained the flaccid presentation of the paraplegia, the spinher 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 20 cases of neurosarcoidosis, but its exact incidence is difficult to assess as spinal involvement may remain clinically silent. Various lesions have been described including extra- and intradural granulomas developing as compressive masses and infiltrative granulomatous nodules usually associated with arachnoiditis. To ascertain spinal cord involvement in sarcoidosis has proved to be difficult. Myelography has shown the spinal cord to be swollen, thinned, or normal. The value of MRI remains to be shown. Despite the absence of controlled trials, corticosteroids have been widely recommended in neurosarcoidosis with considerable results. Alternative treatments are few. Radiotherapy has been occasionally proposed and some improvement noted in intracranial compressive granulomas or in...
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