Plasmocytoma masquerading as a pituitary tumour

Sir: Brain and orbital involvement due to infiltration of the base of the skull are not uncommon complications of myelomatosis. Cranial nerve palsies, especially of the VI nerve are commonly seen. The skull is occasionally the site of a solitary myeloma or plasmocytoma, which may mimic paracellar or orbital tumours. Even when myelomatosis is found, it may be difficult to diagnose an intracranial plasmocytoma. We have recently studied a patient with clinical and radiological findings indicating a pituitary tumour, but further laboratory tests indicated a plasmocytoma, which was confirmed by biopsy. The patient was not operated upon, but received radiation treatment and chemotherapy with initial good effect. After 1 1/2 years remission the patient died.

A 57-year-old male worker was admitted in July 1980 because of headache and diplopia of two months duration. Neurological examination showed a left sided VI nerve palsy accompanied by diplopia. The visual acuity and visual fields were normal. Pattern reversal visual evoked responses (VER) showed borderline latencies for the P2 peak for both eyes. No involvement of the cranial nerves was found. A plain radiograph of the skull showed destruction of the dorsum sellae and floor of the sella turcica (fig la). A CT scan showed enlargement of the pituitary fossa with an intrasellar mass extending into the basal part of the suprasellar cistern (fig 1b). A complete radiological skeletal survey as well as scintigraphy revealed no osteolytic bone lesions. Laboratory tests of pituitary function revealed normal values of prolactin, TSH, triiodothyroxin, thyroxin, LH, FSH, growth hormone, morning and afternoon serum cortisol level as well as urinary excretion of 17-KS and 17-OH KS. Routine blood examination, urinalysis, liver and renal tests were normal. However, electrophoresis of the serum protein showed increased gammaglobulin (24 g/l) and immune electrophoresis revealed high levels of monoclonal IgG (19-7 g/l). Examination of the bone marrow demonstrated normal cellularity, but an increase of plasma cells (12%). Cerebrospinal fluid examination showed no increase of cells and normal total protein, but agarose electrophoresis revealed marked monoclonal components in the cathode part of the gamma region. Bence-Jones protein was not present in the urine. It was concluded that the pituitary mass represented a solitary intracranial plasmocytoma. A transphenoidal needle biopsy was performed via the nasal cavity. Light microscopy showed plasmocytoma with abundance of plasma cells, partly atypical. No further operative procedure was performed. Radiation therapy of the patient’s skull with a total dose of 30 Gy was given, followed by treatment with melphalan, prednisone and vincristine. The patient responded well to this treatment. After the chemotherapy was completed, no neurological deficit was found. A CT scan 1 year later showed a normal sella. In January 1982, 1 1/2 years after the initial admission, the patient suffered a sudden relapse and died of renal failure. At autopsy no myelomatous involvement of the brain was found. However, multiple deposits in ribs and vertebra as well as in the spleen were found.

The neurological and radiological findings in this patient were highly suggestive of a pituitary tumour with erosion of the sella. However, a plasmocytoma was suspected because of the high IgG levels of the serum and the bone marrow biopsy. This was confirmed by a needle biopsy from the tumour. Although the skull is occasionally the site of an apparently solitary plasmocytoma, this is usually the initial manifestation of myelomatosis. In agreement with other authors we emphasise that a plasmocytoma must be considered in patients with intracranial lesions eroding the base of the skull, even in the absence of other concomitant characteristics.
We have subsequently analysed a further 137 cases reported between 1977 and April 1983, although in a number of these the drug had been administered considerably before 1977. In these further cases, a relationship to the administration of hydroxyquinolines was considered probable in 27, possible in 28 and unlikely in 17. Insufficient information was available in 31, and 14 were excluded from evaluation because the symptoms were not neurological or because documentation of hydroxyquinoline intake was not presented.

Combining the assessments from the two series yields a total of 359 reported cases with the following attributability distribution: probable 69, possible 97, unlikely 59, no relationship 30, insufficient information 104. As was emphasized in our previous report, it is striking that the number of cases reported from outside Japan is of quite a different order from the large numbers encountered in that country before clioquinol sales were stopped in 1970. The reason for this disparity remains uncertain, but the greater consumption of clioquinol in Japan is likely to have been the most important factor.

In our assessment of the cases reported since 1977, the aetiological categorisation again included cases of acute fully reversible toxic encephalopathy with amnesia as a prominent feature. This usually occurred following the intake of a large amount of the product over a short period. Also included were cases of isolated optic atrophy of subacute or insidious onset, most commonly in children. Thirdly there were cases of myelopathy, usually of subacute onset, either in isolation or accompanied by optic neuropathy. It is of interest that no cases of peripheral neuropathy were identified in the probable category, either in isolation or associated with optic neuropathy or myelopathy. It is now clear that the neurotoxic effects of the halogenated hydroxyquinolines are substantially confined to the central nervous system. The term subacute myeloptic neuropathy (SMON) is therefore probably a misnomer.

Isolated ictal autonomic symptoms in complex partial seizures

Sir: A variety of autonomic changes may occur at the onset of complex partial seizures. Autonomic disturbances may frequently comprise the seizure aura, and may occur independently after the remission of generalised seizures.

A 46-year-old right-handed man was hospitalised for transurethral prostate resection. Eighteen years earlier he had fallen from a vehicle travelling at 50 miles per hour, and had sustained a right frontotemporal extradural haematoma which was surgically evacuated. He recovered uneventfully from the prostate operation, but three days later was found seated in a chair with intermittent difficulty in comprehending what was said to him, although he remained alert and conversant. For periods of one to three minutes, the patient became flushed and had left-sided piroerection over the arm, leg, and trunk, with enlargement of the left pupil from 3 to 5 mm. The right pupil enlarged occasionally, but right-sided piroerection was not present. These episodes were separated by periods of five to six minutes during which his skin colour was normal, piroerection was absent, and pupillary size and reaction were normal and symmetric. On examination he was able to speak and comprehend, and speech and mentation did not change; flushing, pupillary dilatation, and piroerection subsided. He was afebrile and had no change from a normal pulse and blood pressure during the episodes. He had a mild left hemiparesis with increased tone, which did not change between episodes, and focal clonic jerking was not present. Deep tendon reflexes were increased on the left and left Babinski sign was present. Pathological reflexes and frontal release signs were otherwise absent. After approximately one hour of episodic autonomic changes, he developed focal clonic jerking of the left arm, followed by a generalised convulsion with incontinence and postictal confusion. He was given intravenous phenytoin and had no further seizures. An electroencephalogram several hours later showed polymorphic delta slowing in the right temporoparietal area, with frequent epileptiform discharges arising from the F3 and T4 electrodes. Computed tomography showed postoperative changes in the right frontal region and right frontotemporal low density, with enlargement of the right lateral ventricle. Cerebrospinal fluid was normal. The patient’s family had observed no previous seizures.

Letters

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