Letters

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.1 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 paracellular2-5 or orbital tumours.1 Even when systemic 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 1½ 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 V cranial nerve was found. A plain radiograph of the skull showed destruction of the dorsum sellae and floor of the sella turcica (fig 1a). 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, 1½ 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.6 7 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 characteris-

Fig (a) Conventional lateral skull radiogram show enlarged pituitary fossa (arrows) with erosion of the dorsum sellae. (b) Coronal scan with enhanced intrasellar tumour (arrows).
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 attribution 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 choquinol sales were stopped in 1970. The reason for this disparity remains uncertain, but the greater consumption of choquinol in Japan is likely to have been the most important factor.

In our assessment of the cases reported since 1977, the semiological 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.

G BAUMGARTNER* O GILLAND H KAESER CA PALLIS F CLIFFORD ROSE HH SCHAUMBURG PK THOMAS NH WADIA
* Neurologische Universitätsklinik, Kantonsstital, Rämistrasse 100, Zürich, Switzerland.