Letters

Stabilisation of a severe case of P.O.E.M.S. syndrome after tamoxifen administration.

Sir: In the absence of consistently effective treatment for plasma cell dyscrasia with polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes (P.O.E.M.S. syndrome), it seemed of interest to report a possible response to tamoxifen therapy.

A 56 year old man had been treated since 1982 for a sensorimotor neuropathy with oedema of the lower limbs. Since 1980 he had suffered from impotence, testicular atrophy, hypertrichosis and gynaecomastia. Nerve biopsy showed moderate decrease of myelinated fibres, without specific uptake in immunofluorescence and with no amyloid deposits. A monoclonal lambda IgG dysglobulinaemia (17 g/l, IgA 2.5 g/l, IgM 0.15 g/l) appeared as a complication after 6 months. Vertebra D12 was radio-opaque, and the diagnosis of plasmacytoma was confirmed by guided biopsy (the specimen showed preferential, especially lambda, IgG uptake in plasma cells). Iliac bone biopsy showed moderate plasmacytic infiltration. The C.S.F. protein level was 3 g/l, with an immunoelectrophoretic profile comparable to that of serum (IgG = 0.40 g/l).

Different treatments were undertaken: radiotherapy of vertebra D12 (3,300 rads) in 1982; sequential chemotherapy associating melphalan and prednisone (3 treatments) or melphalan, lomustine, cyclophosphamide and prednisolone (6 treatments); and combinations of vincristine, doxorubicin and dexamethasone according to the protocol of advanced multiple myeloma refractory to alkylating agents.¹ This last treatment resulted in medullary aplasia with septicæmia. All these treatments were ineffective. Weakness worsened in the lower limbs and spread to the upper limbs, so that in 2 years' time the patient was bedridden with intense pain refractory to symptomatic therapy. Generalised melanoderma developed, followed by pleural and peritoneal effusions and pericarditis with tamponade. Throughout this period the electrophoretic peak remained stable.

Out of desperation, after having read an article by Narasimhan² on the use of tamoxifen in the treatment of refractory lymphoma, we undertook this therapy (10 mg twice a day) in February 1985. Within 3 months, clinical improvement was spectacular, with disappearance of the oedema

and serous effusions and with partial and gradual regression of neurological signs (with resumption of physical activities, particularly walking, and disappearance of pain). There was moderate improvement in nerve conduction velocities. Two and a half years later, the general state of the patient is good, and his biochemical values remain stable.

In a study of 102 Japanese cases of P.O.E.M.S. syndrome,³ T. Nakanishi noted the possibility of improvement of neurological and other symptoms in a minority of patients by administration of prednisone and cyclophosphamide alone or in association. However, the majority of patients died (often of heart failure) after a mean survival period of 33 months. Several cases of improvement after irradiation of a solitary plasmacytoma have also been reported.⁴

The improvement in our patient after tamoxifen administration, when all other treatments had failed, suggests that this agent may be another therapeutic arm to combat the rapidly developing P.O.E.M.S. syndrome. Tamoxifen is ordinarily used as an antioestrogen in certain breast cancers and has also demonstrated its efficacy in a few cases of malignant lymphopathies.² Its action seems to stimulate the activity of natural-killer lymphocytes. Might there be a relationship with the gonadotrophic endocrine component of the P.O.E.M.S. syndrome, which has been particularly studied by Bardwick *et al*?⁵

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Accepted 12 September 1988.

Unilateral essential tremor after wrist immobilisation: a case report

A 62 year old right handed decorator fell down stairs and fractured his wrist. This was treated by immobilisation in a scaphoid plaster from thumb to elbow for 6 weeks. When the plaster was removed the subject immediately noticed a tremor in his right hand which was intermittent and posturally dependant. With the wrist in a neutral or flexed position no tremor was present, but when the wrist was extended a marked tremor arose.

Prior to the accident he had been aware of some tremor in the outstretched fingers but it was so small that it did not interfere with living and was too slight for him to seek advice. There was no family history of tremor, and no problem with the left hand.

On examination the only abnormality was in the right arm. The right hand was normal in flexion and the neutral position. As soon as a posture involving extension was assumed a severe flexion-extension tremor appeared at the wrist. Writing could also provoke it. The frequency of the tremor was 7Hz, and it was absent during other postures of the wrist. The left hand was tremor free. There was no dystonia, no Parkinsonian features and no loss in dexterity in the fingers.

In the subsequent 4 years the tremor has not spread to the opposite arm nor worsened in the right arm. It has not been improved by mysoline.

The clinical diagnosis was of an essential tremor, revealed clinically by the injury or more likely by the immobilisation. The history of some finger unsteadiness previously might suggest that the effect of the injury and its treatment was to raise a subclinical tremor to the clinical level. Its exquisite dependence on wrist posture might suggest that a peripheral trigger, possibly from an unbalancing of muscular afferents was important, although a central origin related to motor command cannot be excluded.

Dystonia have been described following peripheral injury,¹ and there is a case of an intention tremor following trauma but this was in association with clinical evidence of a cerebellar lesion.² The present subject's clear relation between injury and tremor appears to be a novel observation and begs the

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question as to why the phenomenon is not seen more widely, since many patients with subclinical tremor must suffer wrist injuries of this type.

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Accepted 21 October 1988

Superior sagittal sinus thrombosis in Wegener's granulomatosis

Sir: We report the occurrence of sagittal sinus thrombosis in a patient with systemic micropolyarteritis, an association we believe not previously described.

A 57 year old man had developed insulin dependent diabetes mellitus at the age of 19. He first experienced headaches in 1977. He became systematically unwell in 1981 with a rising plasma creatinine and an ESR of 123 mm/h. A renal biopsy was performed, and an arteriovenous fistula was fashioned but not used because renal function reverted to normal spontaneously.

In 1983 he again became unwell with repeated epistaxes, facial numbness, headache and persistently high ESR. His symptoms responded to corticosteroids.

In 1984 he developed visual distortions and subsequently visual obscurations. CSF pressure was normal though protein was 0.66 g/l.

In 1987 he presented with papilloedema. CSF pressure was 30 cm H₂O. There were 7 white cells/mm³, protein 1.77 g/l. CSF electrophoresis showed an oligoclonal pattern indicative of local synthesis. Cranial CT (fig. a) revealed thickening of the meninges and low density in the sinus confluence after intravenous contrast, consistent with sagittal sinus thrombosis ("empty delta" sign). This was confirmed by angiography (fig. b).

A lumbo-peritoneal shunt produced an improvement in the visual symptoms and headaches, though recovery was complicated by two focal seizures.

Further renal biopsy and review of the 1981 histology showed a focal necrotising micropolyarteritis. Anti-neutrophil cytoplasmic antibodies were positive at 20% (normal <15%). Other auto-antibodies including anti-nuclear antibodies were negative. Coagulation was normal and the lupus anticoagulant was absent. Examination of the nasal septum revealed bilateral ulcerating lesions. Biopsy showed chronic inflammatory changes. Otological examination was normal.

These clinical features and investigations suggest a systemic micropolyarteritis of the Wegener's type. Wegener's granulomatosis involves the nervous system in up to 50% of cases,¹ typically causing cranial and peripheral neuropathies, aseptic meningitis, diabetes insipidus and focal lesions of brain and spinal cord.² The pathogenesis of these complications is by granuloma formation, or more commonly by focal arteritis. Small arteries are most commonly affected.

In our patient we postulate the sagittal sinus thrombosis to be a result of vasculitis, either by extension of thrombus from inflamed small vessels, or as a consequence of chronic aseptic meningitis. Lateral sinus thrombosis has been described in Wegener's granulomatosis but only in a septicæmic patient with otitis media and mastoiditis.³ We are not aware of any other previous report of involvement of the large intracranial veins and sinuses in Wegener's granulomatosis, though systemic lupus erythematosus may rarely produce this complication.^{4,5}

We thank Dr R Ross Russell and Dr D Taube for permission to publish details of a patient under their care.

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Accepted 6 September 1988

Fig 1a CT cranial scan after intravenous contrast enhancement (September 1987) showing thickening of the meninges and "empty delta" sign at sinus confluence.

Fig 1b Digital subtraction angiogram (March 1988). Iohexol administered via catheter in superior vena cava. Demonstrates underfilling of superior sagittal sinus including non-opacified segment in posterior frontal region, consistent with thrombotic occlusion.