Improvement in the POEMS syndrome after administration of tamoxifen

The POEMS syndrome is an acronym for a multisystem autoimmune disorder comprising polyneuropathy, organomegaly, endocrinopathy, serum M-band, and skin changes. About 40% of patients may respond transiently to prednisolone and cyclophosphamide, but others are refractory to all treatment and die after a mean of 33 months. In 1989, Barrier et al reported a patient with the POEMS syndrome who had been progressive and refractory to all treatment but improved after tamoxifen was given. We report a second patient in whom resolution of most of the features of the syndrome occurred after tamoxifen.

In 1985, a 32 year old Indian man presented with a one year history of weakness, numbness, and pain in his lower legs associated with sweats, skin hyperpigmentation, and weight loss of 20 kg over two years. He had generalised lymphadenopathy and signs of a sensorimotor peripheral neuropathy which was demyelinating in type on electrophysiological assessment (table). Sural nerve fascicular biopsy specimens showed demyelinated expanded nerve fibres with no inflammatory infiltrates, amyloid deposits, or evidence of vasculitis. CSF protein was 0.84 g/l. By 1987 the neuropathy involved the upper limbs. He developed impotence, increased lymphadenopathy, fever, orthopnoea, ascites, pleural effusions, peripheral oedema, diffuse skin hyperpigmentation, hirsutism, leucocytosis, and finger and toe clubbing. A polyclonal increase in serum IgG was found, but no paraprotein was detectable on immunoelectrophoresis of serum or urine. Primary hypothyroidism and primary hypogonadism were discovered. Skeletal survey showed considerable periosteal bone formation around the femoral shafts and osteosclerosis of the spine. He was treated with prednisolone 40 mg/day and then also azathioprine 150 mg/day. His fever and lymphadenopathy resolved but there was no neurological improvement.

By 1988 he had also mild chronic bilateral papilloedema, distinct distal limb wasting, and severe limb weakness: MRC grade 4 proximally, grade 3 in his hands, and grade 1 around his ankles and feet. The reflexes were absent. Proprioreception was lost to wrist and knee, as were vibration sensation to shoulders and costal margins, and pinprick, temperature and, light touch sensations to midforearm and knee. A lymph node biopsy specimen showed features of the multicentric type of Castleman's disease. Cyclophosphamide 120 mg/day was added, but the deterioration continued (figure). Vital capacity fell to 1.31 sitting and 0.91 lying. Two 81 courses of plasma exchange in August and September 1988 produced marginal improvement. The azathioprine was stopped. Tamoxifen 10 mg twice daily was commenced in late September 1988. Cyclophosphamide was discontinued in October 1988 because of a chest infection. The prednisolone was reduced such that he remains on 4 mg/day 20 months later.

Drastic improvements have occurred since autumn 1988. Lymphadenopathy, the skin and nail changes, the ascites and pleural effusions, and most of the ankle oedema have resolved, and limb pain has lessened. Strength in all muscles, except those that were grossly wasted, and vital capacity have increased strikingly (figure). Knee and biceps jerks have returned. Light touch and pinprick sensations are now lost only over the feet and ankles. Proprioreception is mildly impaired in the toes. He is able to wash, feed, and dress himself, and walk with a rollator frame. Motor nerve conduction velocities (MNCV) and compound muscle action potential (MAP) amplitudes have also increased (table). Haemoglobin and platelets have risen to 173 g/l and 526 x 10^9/l respectively. Bone marrow examination was normal. Most recently, the patient complained of painful spasms of his shoulder girdle and proximal upper limb muscles. They arose after he had been using his arms in an elevated position—for example, combing his hair or washing his face. They lasted about five minutes during which the arm became cyanosed with venous engorgement. Some myokymia was observed as these spasms receded, though not at other times. EMG revealed sustained motor unit activity not under voluntary control in these muscles. Carbamazepine was started, and the spasms ceased.

This man undoubtedly has the POEMS syndrome, with clinical features of all seven categories outlined by Nakanishi et al. The electrophysiological, histopathological, CSF, and blood abnormalities are all typical, as are the stripling radiological changes. A serum M-protein is absent in about a quarter of cases, but these have a polyclonal increase of immunoglobulins, usually of IgG, as was found in our patient. We suggest that tamoxifen was responsible for his improvement. There was no response to prednisolone, azathioprine, and cyclophosphamide in adequate doses and combinations over 1988–9. Marginal change may have followed the two courses of plasma exchange, one month apart, but it is difficult to attribute the continuing and much greater improvement over the following 22 months to this, which has had little success in previous reports.

Neither spontaneous remission nor stabilisation have been reported in this progressive syndrome. Tamoxifen is used in patients with breast cancer for its anti-oestrogenic properties and has been useful in treating refractory lymphoma. Barrier et al reported a patient with the POEMS syndrome who was successfully treated with tamoxifen. He had a plasmacytoma which had proved refractory to radiotherapy and chemotherapy, and an anti-tumour effect of tamoxifen was proposed. Both in the POEMS syndrome and the related condition of neuropathy associated with osteosclerotic myeloma or plasma- cytoma, successful treatment of the tumour by surgery or radiotherapy may effect a cure or prolonged remission. There is no evidence that our patient has a plasma cell tumour or even monoclonal proliferation, but it seems reasonable that some plasma cell dyscrasia is present and that tamoxifen somehow acted on this. Continuous motor unit activity has not been reported before in the POEMS syndrome. We did not determine the origin of the abnormal activity precisely, but in the clinical context we strongly suspect that it was of peripheral origin. Carbamazepine abolished it.

T P ENEVOLDSON
A E HARDING
National Hospital for Neurology and Neurosurgery
Queen Square
London WC1N 3BG

Correspondence to: Dr Enevoldsen.
Traumatic basal ganglia haemorrhage with slight clinical signs and complete recovery

A traumatic basal ganglia haemorrhage is a rare but serious complication of head injury. Recognition of its prevalence and clinical features has been possible only with the advent of CT. We describe a patient with a large traumatic basal ganglia haemorrhage with slight neurological signs and complete recovery.

A 15-year-old right-handed young woman sustained a left frontotemporal injury in a motorcycle accident. Witnesses reported a short loss of consciousness (lasting a few seconds) accompanied by a sudden and brief extensor "stiffening" of all limbs and followed by a confusionable state (few minutes). On admission to the emergency department an hour later she was awake and fully oriented and reported retrograde amnesia of a few minutes duration. General physical examination and neurological examinations were normal, as were X-ray pictures of skull, chest, and cervical spine, routine laboratory investigations and EKG. The next day she was still alert and cooperative, but complained of diffuse, moderate to severe, band-like headache. She had a very slight weakness of her left lower facial muscles. Her EEG showed a drowsy pattern (flattening with bilaterally random 4-7 Hz low voltage waves, with inverted arousal reaction) without clear cut abnormalities. Two days later a repeat EEG showed right temporo-frontal 1-3 Hz high voltage waves, spreading mainly to the ipsilateral hemisphere. A brain CT scan showed a medium sized haemorrhage surrounded by a slight oedema in the anterior half of the right lentiform nucleus, with a slight bilateral random 4-7 Hz low voltage waves, with inverted arousal reaction) without clear cut abnormalities. Two days later a repeat CT scan showed a reabsorption of the haemorrhage. The EEG had reverted to normal. A right carotid angiogram did not show a vascular lesion.

Traumatic basal ganglia haematomata is a rare (3%) complication of severe closed head injury, occurring mainly in the young,1 the proposed underlying mechanism is shearing of an anterior choroidal or lenticulostriate artery due to violent deceleration, deceleration brought about by a high velocity injury.2 3 In almost every case the haemorrhage is accompanied by the usual pathological features of severe head injury—for example, diffuse axonal injury, multiple contusions, and epidural or subdural haematomata.4 In one large series patients with a traumatic basal ganglia haematomata had a poor prognosis5 but cases with a favourable outcome have been reported.6 Basal ganglia vascular lesions that do not involve the internal capsule may be asymptomatic,7 and subcortical vascular lesions of the dominant hemisphere may bring about only aphasic disturbances8 or even be clinically silent.9 Small basal ganglia haemorrhages in the non-dominant hemisphere may not be associated with the typical cognitive and behavioural syndromes (left neglect, visuospatial impairment etc).10 The interest of the present case lies in its favourable outcome. Although an early CT examination was not performed, we suggest that the early absence of neurologic and EEG abnormalities reflected a slow development of the haematoma.

CARLO-ITALIO PARODI, SERGIO CAMMARATA, NICOLA PIZIO, GIANDOMENICO SACCO
Divisione di Neurologia, Ospedale Galliera, Mura delle Cappuccine 14, 16128 Genova, ITALIA

Correspondence to: Dr Parodi, Via Assarotti 38/21 16122 Genova, ITALIA


Transient pure sensory strokes in patient with aneurysm of rostral basilar artery

Pure sensory stroke (PSS) usually results from a lacunar infarct in the sensory nuclei of the thalamus; however, ischaemic and haemorrhagic lesions with various locations have also been reported.1 We studied a patient with PSS in whom an aneurysm of the rostral basilar artery was disclosed by CT scan and MRI.

On the day of admission a 78-year-old right-handed man developed three brief episodes of numbness and unpleasant dysesthesia on the right side of the body. He had no headache, stiff neck, dizziness, or visual symptoms. He was in good general health, without hypertension, which was well controlled with medication. Neurological examination performed during one of the episodes showed that he was conscious, well oriented, and aware of his disorder. There was loss of temperature and pain sensation affecting the left side of the body including the face. Touch, vibration, position sensation, graph-aesthesia, and stereognosis were normal, and no other neurological deficit was found. A few minutes later the symptoms resolved spontaneously, and the neurological examination showed no objective sensory disturbances. Speech and language and other cranial nerve functions were normal. General physical examination was unremarkable and laboratory studies showed normal results. Electroencephalogram, somato-sensory, brainstorm auditory, and visual evoked potentials were also normal. The CT scan showed a round area of contrast enhanced density in the region of the interpeduncular fossa, with the CT features of a rostral basilar artery aneurysm (figure, top). MRI confirmed the presence of an aneurysm extending from the upper pons to the inferior aspect of the third ventricle without affecting the thalamus and compressing the left cerebral peduncle slightly strophic (figure, middle). MRI disclosed hypointense images within the aneurysm, suggesting a clot inside its lumen (figure, bottom). Both CT scan and MRI did not show any abnormality of a focal nature in the brainstem, internal capsule, basal ganglia, or cerebral hemispheres. A digital venous angiogram showed no stenosis or ulceration in the carotid or basilar arteries. Reassessment 4 months later showed a normal neurological examination, and the patient reported that no other similar disturbances had occurred.

The neurologic disorder was in this patient the meet the established criteria for transient ischaemic neurological deficit (TIA) as they resolved within a few minutes after onset. Both CT scan and MRI showed a small intracerebral haemorrhage of the rostral basilar artery six months after the onset of the neurological disorder, without any other pathological change elsewhere in the brain. Therefore the precise vascular territory affected cannot be identified, but on the basis of the anatomic location either the vascular supply of the thalamus or of the upper midbrain explains the symptoms and signs presented.2 Asymptomatic aneurysms
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T P Enevoldson and A E Harding

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