Persistent segmental cerebral artery constrictions in coital cephalgia

Onset of severe headache during sexual intercourse can result from subarachnoid haemorrhage, but more commonly it has a benign aetiology.1-7 In the majority of cases, such coital cephalgia has a vascular character, although it may also relate to muscular contraction or, rarely, to low pressure in the cerebrospinal fluid.1 There are similarities with the headache of pheochromocytoma,6 and a hyperdynamic circulation has been implicated in some instances.7 In addition, coital cephalgia has been regarded as a migraine variant because of its character, its response to beta blockade, the frequent past or family history of migraine in affected individuals, and the rare development of residual neurological deficits.7,8 There have recently been several reports of angiographic abnormalities in migraine consisting of multiple segmental narrowings of cerebral arteries.5-6 However, cerebral angiography in coital cephalgia has previously been reported to be normal,4,10 to the extent that its use has been reserved for those patients in whom a more sinister pathology is suspected. However, we now report a case of coital cephalgia with radiological findings which resembled those in migraine, but which were found to persist when angiography was repeated after several months.

A 35-year-old right-handed white woman was admitted to hospital with a severe bifrontal headache which began 10 days previously. Initially there was a throbbing pain that later became a dull ache which fluctuated in intensity. It was accompanied by vomiting but there was no photophobia or neck stiffness. She later described its onset as associated with orgasm, and admitted that she had experienced similar headaches for 18 months, always precipitated in the same way. Previously they had resolved after a few hours. There was no family history of migraine, but the patient described violent, unilateral, throbbing headaches during her twenties which had not been associated with sexual intercourse. There was no history of head injury and she was not taking oral contraceptives or any other regular medications.

Initial examination showed no neck stiffness or Kernig's sign, normal fundi and no focal neurological deficits. The blood pressure was 150/90 and subsequently remained stable. Because of the duration of symptoms, it was thought necessary to exclude a subarachnoid haemorrhage. A cranial CT scan showed no evidence of intracranial bleeding. At lumbar puncture the pressure was 175 mm CSF. The fluid was not xanthochromic and analysis was otherwise normal. Four vessel angiography was performed under general anaesthetic but this did not reveal angioma or aneurysm. However, there were segmental narrowings involving cortical branches of many of the vessels in the supratemporal and infratemporal compartments bilaterally. Subsequent blood tests were normal, including the erythrocyte sedimentation rate (4 mm/hour), C-reactive protein (less than 6 mg/L), antinuclear and rheumatoid factors. Twenty four hour urinary excretion of vanillylmandelic acid (VMA) was also normal (31, 29 and 28 µmol/24 hr).

The patient was treated with clonidine (50 µg bid) with subsequent resolution of her headaches despite resumption of sexual activity. After three months, however, her clonidine compliance became erratic and the headaches recurred in an attenuated form without orgasm. However, they again subsided when treatment was resumed.

Intravenous digital subtraction angiography was performed, eight months after presentation. Despite the poorer resolution obtained by this method compared with the arterial study, some of the middle cerebral cortical branches were observed without significant overlap, and they still had regions of narrowing similar to those seen previously.

Despite the duration of her presenting headache, its onset and character were typical of this patient's coital cephalgia. This diagnosis was further supported by the response to migraine prophylaxis and the relation of symptoms to compliance with treatment. Lundberg and Osterman11 regarded persistence of pain beyond 24 hours as favouring subarachnoid haemorrhage rather than benign orgasmic cephalgia. However, subsequent authors have reported cases of the latter experiencing pain for several days.12 In our case there was no evidence of intracranial bleeding and the clinical picture, negative autoantibody tests, and normal erythrocyte sedimentation rate did not support the presence of a vasculitic illness to explain the segmental cerebral arterial constrictions. These changes have also been reported in pheochromocytoma,13 but again the patient's blood pressure and normal urinary VMA excretion were not in favour of this diagnosis. Finally, her benign clinical course was unlikely to be due to an isolated cerebral angiitis.14

Patients with coital cephalgia usually present after many attacks of pain, and angiography is not generally performed since the diagnosis is then apparent. Nevertheless, in at least 15 previous cases having cerebral angiography the appearances were normal.14,15 Segmental arterial narrowing has been reported in three patients with
migraine and one with atypical cluster headache. These changes are compatible with arterial spasm of vessel wall oedema and were found to be transient whenever angina was not repeated. In our case, however, at least some of the arterial constrictions persisted. It is possible that they were not related to the patient’s symptoms. Nevertheless, it is also known that the spasm following subarachnoid haemorrhage can leave the vascular lumen permanently narrowed because of ensuing mural fibrosis. The possibility arises that similar structural changes occurred in our patient, and the spasm described in his associated head ache could occasionally lead to permanent narrowing of the affected vessels.

R KAPOOR
BE KENDALL
MG HARRISON
Departments of Neurology and Radiology,* The Middlesex Hospital, Mortimer Street, London, United Kingdom.


Resolution of a severe sensorimotor neuropathy following resection of an extranodal malignant lymphoma

Lymphoma may cause a peripheral neuropathy by nerve infiltration or by non-metastatic effects and occasionally this may be the presenting feature. We describe a patient presenting with a severe sensorimotor axonal neuropathy which resolved following the resection of an asymptomatic localised gastric lymphoma. Over a period of two months a 75 year old woman developed difficulty in walking followed by distal paresthesiae and increasing weakness in all four limbs. She had lost 6 kg (13.2 lb) of body weight and had no other symptoms. General and cranial nerve examinations were normal. There was generalised muscle wasting in the limbs, particularly the lower limbs, with atrophy of the hand muscles and quadriceps. There was hypotonia and symmetrical proximal and distal weaknesses of all limbs (MRC grade 3-4). All tendon reflexes were absent with plantar responses flexor. Sensation was intact except for minor distal impairment of light touch and pinprick. There was no ataxia. During the next two months weakness and ataxia developed in the upper limbs but similar proximal and distal sural and biceps, and lower limb, muscles were affected. Power became grade 2 or less in all limb muscles and light touch and pin prick loss ascending to mid-limb level with complete loss of proprioception to wrists and ankles. Cranial nerves were normal. The patient was unable to walk and the patient was unable to walk. Electromyography on admission showed fibrillation potentials, positive sharp waves and a reduced interference pattern in upper and lower limb muscles. Sensory action potentials (SAP) were normal in the upper limb muscles but sural SAPs were not obtainable. Motor conduction velocities were slightly reduced: right median 47 ms, ulnar 43 ms and left lateral popliteal 41 ms. Routine blood investigations were normal including serum electrophoresis, heavy metals and Borrelia serology. Urinary porphyrins were negative. Cerebrospinal fluid (CSF) cell count, protein and cytology, chest radiographs and pelvic ultrasound, sigmoidoscopy and rectal biopsy, bone marrow aspirate and trephine were all normal.

A partial thickness sural nerve biopsy was performed. Paraffin sections revealed severe axon loss and demyelination with no inflammatory cells or amyloid. Plastic sections confirmed extensive loss of myelinated fibres and some remaining were thinly myelinated. There were numerous Schwann cell processes but very few axon sprouts. Teased fibre preparations showed most fibres to be columns of Schwann cells with no debris. The small number of myelinated fibres were all regenerative fibres. On immunohistochemistry, there were no deposits of IgG, IgM, C1q or C3d or cells showing the leucocyte common antigen. At gastroscopy the suspicious area in the gastric fundus was biopsied. This revealed extensive infiltration by B-cell monoclonal lymphocytes (markers M1, negative, UCHL1 negative, MOPC positive, MOPC positive, Kappa negative, lambda positive). A proximal gastrectomy was performed, resecting all the macroscopically abnormal stomach. The left gastric artery was divided but all other nerves, the liver and spleen and the rest of the bowel were macroscopically normal. Histology confirmed a well differentiated low grade B-cell lymphocytic lymphoma, extranodal, involving at least four lymph nodes. It stained with monoclonal antibodies to lambda light chains and IgM heavy chains. Her neuropathy improved dramatically over the next three months. She could stand unaided after four weeks, walk with a Rottor six weeks later and manage stairs and live independently by three months. Improvement was confirmed myelometrically. Sensation became normal except for minimal proprioceptive impairment in the fingers and toes.

This elderly woman presented with a sensorimotor neuropathy which was of subacute onset, progressive and eventually very severe. The clinical suspicion of lymphoma malignancy was eventually confirmed by the finding of a B-cell gastric lymphoma. The lack of demonstrable spread despite extensive investigation, the normal CSF findings and the slow clinical course. This latter group was characterised by frequent spontaneous stabilisation and minor sensory involvement both clinically and at necropsy. Although the early electrophysiological studies demonstrated a predominantly motor neuropathy, our case is distinguished from those of Schol and others by the severity of the eventual clinical sensory loss and by the very severe axonal loss in the biopsy of the sural nerves. Furthermore, the neuropathy did not merely stabilise but resolved dramatically. This occurred after the removal of the lymphoma and although proof of a causal relationship is lacking, the timing between improvement of the neuropathy and the resection of the lymphoma is compelling. Furthermore, the patient has not relapsed in over eleven months of observation. There have been no previously reported cases of a paraneoplastic lymphomatous neuropathy resolving with treatment of the tumour, although this has been noted without such precise timing in neuropathies associated with benign extranodal carcinomas. This improvement in our case was all the more surprising in view of the severe axonal loss on sural nerve biopsy. Presumably, in other cases axonal loss could not have been so severe or alternatively their function may have been impaired by an element of demyelination, though conduction velocities were only slightly reduced.

The stomach is the commonest extranodal site for non-Hodgkin’s lymphoma and resection with or without radiotherapy may be curative. Given the gratifying response of this patient’s neuropathy to treatment of her extranodal lymphoma, we advocate that when investigation in such cases is extended to the