LETTERS TO THE EDITOR

Occipital neuralgia: another benign cause of "thunderclap headache"

Headaches characterised by paroxysmal onset of severe, generalised pain ("thunderclap headache") are regarded as early signs of subarachnoid haemorrhage. It has even been suggested, that despite normal cranial CT and CSF studies, thunderclap headaches may be a sign of an unruptured intracranial aneurysm therefore heralding an increased risk for subarachnoid haemorrhage.¹ However, recent reports on the long term follow up of patients with thunderclap headaches show that with normal CT and CSF studies, thunderclap headaches are usually benign,² often vascular in origin.³

Over the past four years we have seen 12 patients (ages 27-54 years; 7 women, 5 men) with occipital neuralgia presenting with excruciating, generalised headaches of explosive onset. None had rheumatoid arthritis, recent trauma, or cervical spine abnormalities. The pain was shooting in 8 and throbbing in 4. All patients described nausea, vomiting and photophobia. Eleven experienced blurry vision, 7 vertigo or dizziness, 5 neck stiffness, and 5 stuffiness of the nose. Pressure on the greater occipital nerve produced a positive Tinel's sign in all cases, and 9 had hypoaesthesia in the sensory distribution of C2. Within 30 minutes of an occipital nerve block with 1-2 cc of 1% lidocaine, complete pain relief was achieved in 7 patients and significant improvement (residual pain less than 10% severity) in all the others. CT scans in 12/12 and CSF analysis in 5/5 were normal. Cervical spine x rays were normal in all patients.

Occipital neuralgia is caused by irritation or injury to the greater occipital nerve, and is generally characterised by uni- or bilateral throbbing pain that frequently radiates to the forehead and around the eye.⁴ However, our patients illustrate a much more dramatic form of presentation mimicking subarachnoid haemorrhage. The greater occipital nerve is the continuation of the second cervical nerve root and receives branches from the superior cervical sympathetic ganglion, the trigeminal ganglion, the acoustic, and the vestibular nerves.⁵ Occipital neuralgia is therefore frequently associated, as in the case of our patients, with autonomic dysfunction in the neck and face, vertigo, nose stuffiness, and visual disturbances.⁴ The diagnostic clinical features for occipital neuralgia are the presence of a sharply circumscribed area of tenderness over the greater occipital nerve trunk as it crosses the superior nuchal line, sensory changes in the C2 distribution, and the response of the pain to infiltration of local anaesthetic near the tender area of the nerve trunk.⁶ If the diagnosis of occipital neuralgia is established, the only potentially serious underlying conditions are craniovertebral anomalies of the cervical spine and cervical arthritis, which can be detected with cervical spine x rays.^{6,7} Cervi-

cal spondylosis can present with thunderclap headaches and lead to chronic headaches associated with neck movements.²

Timely clinical recognition of the acute form of occipital neuralgia can help avoid unnecessary testing in patients with thunderclap headache and lead to the appropriate treatment with almost immediate pain relief.

A PASCUAL-LEONE
National Institute for Neurological
Disorders and Stroke, National
Institutes of Health, Bethesda,
MD, USA

A PASCUAL-LEONE PASCUAL
Departamento de Medicina Interna,
Sección de Neurología, Hospital
Clínico y Universitario de
Valencia, Valencia, Spain

- Day W, Raskin NH. Thunderclap headache: symptom of unruptured cerebral aneurysm. *Lancet* 1986;ii:1247-48.
- Harling DW, Peatfield RC, Van Hille PT, Abbott RJ. Thunderclap headache: is it migraine? *Cephalalgia* 1989;9:87-90.
- Widjicks EFM, Kerkhoff H, Van Gijn J. Long term follow up of 71 patients with thunderclap headache mimicking subarachnoid haemorrhage. *Lancet* 1988;i:68-70.
- Chouret EE. The greater occipital neuralgia headache. *Headache* 1967;7:33-4.
- Bogduk N. The anatomy of occipital neuralgia. *Clin Exp Neurol* 1980;17:167-84.
- Hammond SR, Danta G. Occipital Neuralgia. *Proc Austr Assoc Neurol* 1978;15:258-70.
- Ehni G, Benner B. Occipital neuralgia and the C1-2 arthrosis syndrome. *J Neurosurg* 1984;61:961-5.

Pure sensory Guillain-Barré syndrome

The existence of a purely sensory form of Guillain-Barré syndrome is still subject to controversy, although the criteria for its

diagnosis have been established. We report the case of a patient who had acute sensory neuropathy which, due to its clinical, cerebrospinal fluid and electrophysiological characteristics, may be considered a sensory form of Guillain-Barré syndrome.

Three days before admission, a 69 year old woman developed a sensory deficit, with a sensation of tightness and dysesthesias in her feet and hands, which increased in intensity and extension during the following days. Two days later she showed a markedly unsteady gait and clumsiness in handling her upper extremities. She was admitted to our centre the next day. She had been vaccinated against influenza a month before and had suffered from intense sore throat, specially on swallowing, without fever during the two weeks before admission.

On neurological examination, her mental status, cranial nerves, strength, light touch and pinprick sensation were normal, while there was a complete loss of vibration and arthrokinetic sense. There was dysmetria of the four extremities, the intensity of which rose considerably when she closed her eyes, hypotonia of the upper extremities, pseudoathetoid movements and total areflexia with cutaneous plantar reflexes in flexion. She showed marked truncal ataxia, a broad-based gait, and was unable to stand or walk without support. Romberg's sign was positive.

Routine laboratory tests (except for an ESR of 37 mm/h), levels of folic acid, vitamin B12, tumour markers (alfa-fetoprotein, CA-125, CEA), HIV and syphilis antibody tests, ANA, antiDNA, anti-Ro, anti-LA and anti-mitochondria antibodies, were normal. Serum IgG anti-Hu were negative. ECG, chest x ray, abdominal ecography and a thoracoabdominal CT scan were also normal. Two CSF examinations were carried out, on the

Table Nerve conduction values

	Day 6 ¹	Day 18	Day 110	Lower limit of normal range
Motor				
Right median				
Amplitude (mV)	3.08	1.6	1.25	5.8
Distal latency (ms)	4.8	4.4	5.2	3.2
Conduction velocity (ms ⁻¹)	45.6	48	51.0	33.8
F wave latency (ms)	nd	nd	26.8	28.7
Left median				
Amplitude (mV)	2.96	nd	nd	5.8
Distal latency (ms)	3.8	nd	nd	3.2
F wave latency (ms)	23.6	nd	nd	28.7
Right ulnar				
Amplitude (mV)	6.44	nd	nd	5.3
Distal latency (ms)	2.8	nd	nd	2.8
Conduction velocity (ms ⁻¹)	50	nd	nd	47.4
Right peroneal				
Amplitude (mV)	2.28	nd	3.08	1.2
Distal latency (ms)	5.6	nd	5.6	4.2
Conduction velocity (ms ⁻¹)	42.3	nd	43	42.2
Sensory				
Right median (III-Wrist)				
Amplitude (μV)	ns	3.3	14.6	12.3
Distal latency (ms)	ns	3.7	3.1	2.2
Right ulnar (V-Wrist)				
Amplitude (μV)	ns	1.1	4.4	4.2
Distal latency (ms)	ns	3.1	1.9	1.8
Right sural				
Amplitude (μV)	ns	nd	2.6	6.4
Distal latency (ms)	ns	nd	3.7	3.3
Conduction velocity (ms ⁻¹)	ns	nd	37.8	41
Left sural				
Amplitude (μV)	nd	nd	2.9	6.4
Distal latency (ms)	nd	nd	3.9	3.3
Conduction velocity (ms ⁻¹)	nd	nd	35.8	41
Mixed				
Right median				
Amplitude (μV)	ns	1.2	1.8	5.5
Conduction velocity (ms ⁻¹)	ns	48.9	55.6	51.7
Left median				
Amplitude (μV)	ns	nd	nd	5.5
Conduction velocity (ms ⁻¹)	ns	nd	nd	51.7

LL = lower limit of normal range; ns = not seen; nd = not done.

¹ Days from the onset.

third and nineteenth day from the beginning of the symptoms, the first one being normal and the second one showing an increase in protein (0.95 g/l) with 11 cells and negative titres to brucella, syphilis, toxoplasma, herpes simplex, varicella-zoster, and a negative anticryptococcal antigen determination.

The patient's clinical pattern began to improve two weeks after admission; she was discharged a week later. Follow up a month later revealed only a generalised areflexia and a slight dysmetria on the heel-knee test.

Electrophysiological studies (table) were carried out on the sixth and eighteenth day, which revealed absent sensory potentials. Motor NCS showed less abnormality than that of sensory nerves. There was delay in the distal motor latency in all the nerves tested and there was also reduction in motor amplitude in both median nerves, with a 25.3% drop in peak-to-peak amplitude between wrist and elbow, and a 39.2% drop in amplitude between wrist and axilla in the first right median nerve conduction study. Neither temporal dispersion nor motor conduction block in the remaining nerves was noted. EMG studies were normal. A third electrophysiological study performed a month after her discharge showed the recovery of the sensory nerve action potentials in the lower and upper extremities, although the amplitude and sensory conduction velocity of the nerves studied were lower than the normal values. The motor NCS, however, were similar to previous values. The EMG study of the right anterior tibialis and right first dorsal interosseous muscles showed compatible changes with minimum denervation.

Until recently the Guillain-Barré syndrome was an entity whose nosological characterisation relied upon a purely descriptive base, with relatively widely accepted diagnostic criteria.¹ Among these, the presence of muscular weakness is the most noticeable, indicating the main affection of the motor roots. However, a purely sensory clinical variant of this illness is also believed to be possible and the following features are necessary for it to be accepted:² rapid onset, distribution widespread and symmetrical, complete or near recovery, high CSF protein content, with few or no cells, and an electrophysiological study compatible with a demyelinating process in the peripheral nervous system.

There is still a certain controversy as to whether this clinical variant occurs. In a recent review of 42 patients with acute sensory neuropathy, the authors concluded that this condition is not part of the spectrum of inflammatory demyelinating neuropathies;³ nevertheless, it is noticeable that only 2 of the 42 patients had complete remission of symptoms and that the course of their disease was very protracted—from six to nine months.

A case of acute polyneuropathy has been published in which the sensory disorders were also prominent and very similar to those of our patient.⁴ The necropsy carried out after the patient's death from pulmonary embolism revealed inflammatory infiltration with segmental demyelination in the peripheral nerves, posterior roots and spinal ganglia, with a marked preservation of the anterior roots. These findings, except for the preservation of the motor roots, are identical to those which are described in the Guillain-Barré syndrome.⁵

Our patient had an acute sensory neuropathy after a presumed viral illness and vaccination with a monophasic course and nearly

complete recovery—both clinical and electrophysiological—in a short time; CSF protein content was high with a mild pleocytosis. The evidence suggests that this was probably an acute sensory inflammatory demyelinating polyneuropathy.

FRANCESC MIRALLES
JORDI MONTERO
RAMON RENÉ
JUAN A MARTINEZ MATOS
Department of Neurology,
Hospital Princes d'Espanya,
Ciutat Sanitària de Bellvitge,
Barcelona, Spain

Correspondence to: Dr Miralles, Department of Neurology, Hospital Princes d'Espanya, Ciutat Sanitària de Bellvitge, Feixa Llarga s/n, 08907 L'Hospitalet de Llobregat, Barcelona, Spain

- 1 Asbury AK, Bolis L, Gibbs CJ. Workshop on autoimmune neuropathies: Guillain-Barré syndrome. *Neurology* 1990;40:381.
- 2 Asbury AK. Diagnostic considerations in Guillain-Barré syndrome. *Ann Neurol* 1981;9(suppl):1-5.
- 3 Windebank AJ, Blehrud MD, Dyck PJ, Daube JR, Karnes JL. The syndrome of acute sensory neuropathy: clinical features and electrophysiologic and pathologic changes. *Neurology* 1990;40:584-91.
- 4 Dawson DM, Samuels MA, Morris J. Sensory form of acute polyneuritis. *Neurology* 1988;38:1728-31.
- 5 Prineas JW. Pathology of the Guillain-Barré syndrome. *Ann Neurol* 1981;9(suppl):6-19.

Cerebral venous thrombosis in paroxysmal nocturnal haemoglobinuria

Paroxysmal nocturnal haemoglobinuria (PNH) is a rare acquired disease of the haematopoietic system manifested by chronic haemolytic anaemia, leukopenia, and thrombocytopenia. Patients with PNH have an increased risk of developing systemic venous thrombosis, and obstruction of the hepatic veins, or Budd-Chiari syndrome (BCS) which is a highly fatal complication in about 30% of cases. PNH is also a well-established though extremely uncommon cause of cerebral venous thrombosis (CVT).¹⁻³ We describe a patient with PNH and BCS in whom CVT was angiographically documented. We comment upon the risks of anticoagulants in this unusual situation and the need to remember PNH among the possible causes of CVT.

A 34 year old woman was admitted to hospital because of malaise, low-grade fever, pleural effusion, hepatomegaly and ascites. The diagnosis of PNH was established by the presence of haemosiderinuria, positive Ham acid haemolysis and sucrose lysis tests. Hepatic venogram confirmed the existence of BCS. She received repeated blood transfusions and a year later a portocava shunt was performed. Three years later she complained of intensive fronto-occipital pain and clumsiness of the left limbs. Examination showed a mild distal paresis of the left leg with only minimal slowness of alternating movements in the left arm. There was impairment of position and pain perception with sensory extinction on the left side. The optic discs were slightly blurred. The visual fields were full with a moderately enlarged blind spot on both sides. CT scanning showed a right parietal infarction. Cerebral angiography revealed thrombosis of superior sagittal sinus, straight sinus, lateral sinuses, and internal cerebral veins. The patient made a complete recovery with dexamethasone and coumarin therapy.

In a comprehensive review of 38 cases, Bousser *et al*⁴ failed to list PNH among the

possible causes of CVT. PNH is commonly undiagnosed for a period of months to years, and its true incidence among patients with CVT is probably underestimated because detailed coagulation studies are not performed in most cases. This may be of the utmost importance when treatment with heparin is under consideration. The recent German randomised trial demonstrated the benefit of high-dose heparin in patients with CVT.⁵ Conversely, therapy with heparin has caused exacerbation of the hemolytic process in some patients with PNH,^{1,2} and thus might be deleterious in this particularly difficult situation.

PNH should be considered among other possible causes of CVT and ruled out by the appropriate laboratory investigations, especially when treatment with heparin is contemplated.

A ALFARO
Service of Neurology,
University Hospital La Fe,
Valencia, Spain

- 1 Johnson RV, Kaplan SR, Blailock ZR. Cerebral venous thrombosis in paroxysmal nocturnal hemoglobinuria. *Neurology* 1970;20:681-6.
- 2 Spencer JAD. Paroxysmal nocturnal haemoglobinuria in pregnancy: case report. *Br J Obstet Gynaecol* 1980;87:246-8.
- 3 Benoit P, Lozes G, Destée A, *et al*. Hypertension intracranienne bénigne et maladie de Marchiafava-Micheli. *Rev Neurol (Paris)* 1986;142:782-5.
- 4 Bousser MG, Chiras J, Bories J, Castaigne P. Cerebral venous thrombosis. A review of 38 cases. *Stroke* 1985;16:199-213.
- 5 Villringer A, Garner C, Meister W, *et al*. High-dose heparin treatment in cerebral sinus-venous thrombosis (SVT). *Stroke* 1988;19:135.

Toxic reaction following the combined administration of fluoxetine and phenytoin: two case reports

Fluoxetine is a new antidepressant agent unrelated to the tricyclic antidepressants, whose structure corresponds to a straight chain phenylpropylamide. The drug selectively inhibits reuptake of serotonin but not noradrenaline and has a minimal muscarinic, dopaminergic, histaminergic or serotonergic effect.¹ The only described interaction^{1,3} is with L-tryptophan which enhances its therapeutic effects, but produces symptoms and signs of intoxication.² Presently there is no published work on the possible interaction between fluoxetine and anticonvulsant drugs.

We describe two patients, who developed symptoms and signs of intoxication with phenytoin a few days after initiating the use of fluoxetine.

An 84 year old woman was treated with phenytoin 300 mg daily, after removal of a chronic subdural haematoma. Two months later she developed a depressive syndrome. CT showed no alteration, and the plasma level of phenytoin was 15 µg/ml. Treatment with a dose of 20 mg/day of fluoxetine was given, increasing the dose to 40 mg/day after 10 days. Five days after the beginning of treatment she developed gait ataxia, vertigo, diplopia and alteration of consciousness. Examination also showed dysmetria of the limbs, multidirectional nystagmus, and alteration of judgement with visual hallucinations.

The plasma level of phenytoin was 35 µg/ml. The dose of phenytoin and fluoxetine was reduced gradually and there was progressive recovery from the signs and