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. 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, often vascular in origin.

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 blurring of vision, 7 vertigo or dizziness, 5 neck stiffness, and 5 stiffness 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. 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. 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.

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. 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.

1. Cervical 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.

LETTERS TO THE EDITOR

Headaches and lead up to subarachnoid haemorrhage.


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 dysesthesia in her feet and hands, which increased in intensity and extension during the following days. Two days after she showed a marked 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, specifically 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 arthrogenic sense. There was asymmetry of the four extremities, the intensity of which rose considerably when she closed her eyes, hypotonia of the upper extremities, pseudoneurotoid movements and total areflexia with cutaneous plantar reflexes in flexion. She showed marked truncal atasia, 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 folie 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
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 simples, varicella-zoster, and a negative antinuclear 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 a generalised areflexia and a slight dysmetria of the heel-keen 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 velocities of the nerves were abnormal. 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 Guillain-Barré syndrome was an entity whose nosological characterisation relied upon a purely descriptive base, with relatively widely accepted diagnostic criteria. 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: 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 current 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. 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.

The onset of acute polyneuropathy has been published in which the sensory disorders were also prominent and very similar to those of our patient. 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 axonal component. However, the preservation of the motor roots, are identical to those which are described in the Guillain-Barré syndrome.

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 changes. The 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, JURES MONN RENE
JUAN A MARTINEZ MATOS
Department of Neurology, Hospital Principal de l’Esparro
Ciutat Sanitari de Bellvitge, Barcelona, Spain

Correspondence to: Dr Miralles, Department of Neurology, Hospital Principal de l’Esparro, Ciutat Sanitari de Bellvitge, Feixa Llarga 88070 L’Hospitat de Llobregat, Barcelona, Spain


Cerebral venous thrombosis in paroxysmal nocturnal haemoglobinuria

Paroxysmal nocturnal haemoglobinuria (PNH) is a rare acquired disease of the haemopoietic system manifested by chronic haemytic anaemia, leukaemona, 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).1,2 We describe a case with PNH and BCS in whom CVT was angiographically documented. We comment upon the risks of anti-coagulants in this unusual situation and the need to recognise 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 portacava shunt was performed. Three years later she complained of intense fronto-occipital pain and clumsiness of the left limbs. Examination showed a mild distal paraesthesia of the leg only with only minimal slowing 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 al4 failed to list PNH among the possible causes of CVT. PNH is commonly undiagnosed for a period of time and death from hepatic failure is under consideration. The recent German randomised trial demonstrated the benefit of high-dose heparin in patients with CVT.2 Conversely, therapy with heparin has caused exacerbation of PNH in some patients with PNH,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 de Neurologia, Hospital Universitari de la Santa Creu i de la Santa Pau, Barcelona, Spain


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 seratoninergic effect. The only described interaction is with L-tryptophan which enhances its therapeutic effects, but produces symptoms and signs of intoxication.5 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 in 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 fluoxetine was reduced gradually and there was progressive recovery from the signs and symptoms of intoxication.