ance to this treatment but was improved by IVIG. There was a temporal relationship between neurological and renal improvement and IVIG, and this could reflect a common pathogenesis. Recognition of such coexisting disorders provides further evidence of an immune dysfunction in CIDP.

A P Fèvè, G Rancurel, and J-M Liègeois
Service de Neurologie, Hôpital Salpétrière, Paris
Correspondence to: A P Fèvè, Service de Neurologie, Hôpital Tenon, 4 rue de la Chine, 75020 Paris, France.


Acute stage Bell’s palsy and narrowing of the palpebral fissure caused by drooping of the eyebrow and the upper eyelid

Wideened palpebral fissure on the affected side has been regarded as one of the hallmark of peripheral facial palsy. The palpebral fissure of the affected eye may, however, be narrower than that of the healthy eye. Twenty five patients with Bell’s palsy in the acute stage were examined at our outpatient clinic from January 1990 to May 1991. No other neurological signs were observed in these patients. Ten patients had a narrowed palpebral fissure rather than widened palpebral fissure on the affected side looking straight ahead. Their ages ranged from 32–59 years with a mean of 46–8 years. Seven patents were men and three women, four being diabetic. All 10 patients complained of a drooping of the upper eyelid on the affected side when opening their eyes. All had a furrowless forehead and the corner of the mouth drooped on the affected side, both characteristics of peripheral facial palsy. They were unable to close the affected eye tightly, and five patients had lagophthalmos (incomplete closure). The position of the eyebrow and the upper eyelid on the affected side was lower and the width between their lower margins was shorter than on the non-affected side (fig). The patients experienced difficulty in elevating the eyebrow and the upper eyelid when gazing upwards. The position of the eyes was intact, however, and the extraocular movements were full. The pupils were round, equal in size and reactive to light.

In Bell’s palsy, the principal muscle involved in opening the upper eyelid and maintaining normal eyelid posture is the superior palpebral levator, which is innervated by the oculomotor nerve. Two accessory muscles are Müller’s muscle via the oculospastisic pathway and the frontal muscle by the facial nerve. The upper eyelid is indirectly elevated by the attachments of the frontal muscle into the eyebrow with the superior orbital portion of the orbicularis oculi muscle. Thus it is possible that the weakness of the frontal muscle causes drooping of the eyebrow and the upper eyelid, resulting in narrowing of the palpebral fissure. This phenomenon might be more noticeable in Japanese than in white patients, because Japanese patients are more “heavy lidded”.

SHINO OHIKAWA
TAKASHI YOSHIDA
ATSUSHI YAMADORI
Neurology Service, Hyogo Brain and Heart Center at Himeji, 520 Satcho-Ko, Himeji 670, Japan

Correspondence to: Dr Ohikawa.


Familial recurrent cranial nerve palsy

Familial recurrent cranial nerve palsy is a syndrome of unknown etiology in which affected family members experience recurrent paresis of the oculomotor, abducens, and facial nerves in an apparent autosomal dominant inheritance pattern. Typically, these individuals experience acute onset of a cranial nerve palsy followed by gradual resolu- tion of the deficit over several months. This presentation and natural history is similar to that of patients with diabetic vasculopathic cranial nerve palsies and prompted us to search for a vasculopathy in the patient and family members we describe.

A 36 year old man developed binocular, horizontal diplopia in August 1990. He had experienced an idiopathic, peripheral, right facial nerve palsy (Bell’s palsy) in October, 1985 and a left Bell’s palsy in May 1986. He was an otherwise healthy nurse anesthetist taking no medications. On examination, best corrected visual acuity was 6/6 in each eye and colour vision was normal. Pupils were briskly reactive without anisocoria or a relative afferent papillary defect. Visual fields were full. Facial sensation was intact, and there was no nystagmus or proptosis. There was mild orbicularis weakness bilaterally and evidence of aberrant regeneration of both facial nerves. The patient had an ocular motility disturbance consistent with a left abducens nerve palsy: abduction of the left eye was decreased and there was a 25 prism diopeter (PD) esotropia in primary gaze that increased to 45 PD in attempted left gaze. The remainder of the examination was unremarkable.

A complete blood count, thyroid function tests, rheumatoid factor screen, serum protein electrophoresis, assay for acetylcholine receptor antibodies, assay for angiotensin converting enzyme, Lyme titres, edrophonium test, chest radiograph, and magnetic resonance imaging were all normal with the exception of a mildly elevated cholesterol level in the serum (252 mg/dl) and a mildly elevated CSF protein (55 mg/dl). The left }

Figure Patient looking straight ahead. Narrowing of the palpebral fissure is seen on the left side.
abducens nerve palsy resolved gradually over 4 months.

The patient has one healthy son (age 8) and an unaffected mother, but his father and brother (the patient's only sibling) had experienced cranial nerve palsies in the past. At the age of 39 (5/89), the brother developed a right abducens nerve palsy that resolved over 6 weeks. Three years previously he had had a myocardial infarction and subsequent early coronary bypass surgery that required a 5-veesel coronary artery bypass. The patient's father, an only child, experienced cranial nerve palsies in the early 50's. His father had normal neuroophthalmological examinations in January 1991.

A large number of family members had a thorough haematological evaluation for disorders predisposing to thrombosis including assays for cardiopulmonary antibody, the Lupus anticoagulant (Russel viper venom time), prothrombin, partial thromboplastin, fibrinogen, fibrin split products, plasminogen, antithrombin III, protein C, and protein S. These tests were all normal or negative.

The familial occurrence of recurrent cranial nerve palsies has been described by Yawn and associates in 1950.1 Subsequently, several other families have been reported.2,3 The aetiology of the cranial nerve palsies remains unclear despite extensive, including glucose tolerance tests, serologic testing, erythropoiesis (Tension) tests, cerebrospinal fluid analysis, single fibre electromyography, CT scan, and cerebral angiography. Recently, haematological disorders predisposing to thrombosis have been associated with numerous neurological and systemic manifestations in young adults, including cerebral and myocardial infarction, deep venous thrombosis and migraine.4 Ophthalmic consequences (retinal arterial and venous occlusion, vitreous hemorrhage and ischaemic optic neuropathy) have also been reported in these patients.5 Thrombosis in these disorders may be caused by a deficiency of one or more proteins integral to the regulation of clot formation (antithrombin III, protein C, protein S, fibrinogen) or by the presence of antiphospholipid antibodies (cardiolipin antibody, Lupus anticoagulant) that interfere with phospholipid dependent activation complexes (prothrombinase) essential to clot regulation. These tests were not available for the evaluation of previously reported patients with the syndrome of familial recurrent cranial nerve palsy. We believe that the pathophysiology of neurovascular disease in this syndrome of vasculopathic disease because several of our patients had other evidence of vascular disease and vasculopathic disorders (diabetes, hypertension, coronary artery disease). These vasculopathic disorders are associated with cranial nerve palsies that frequently and improve in a similar manner. Our patients, however, had no laboratory evidence of antiphospholipid antibodies or deficiencies of coagulation cascade proteins and thus may constitute a new type of vasculopathy. Familial recurrent cranial nerve palsy may be caused by an as yet unidentified vasculopathic process.

KARL C GOLNIK
The Storm Eye Institute,
Medical University of South Carolina,
Charleston, South Carolina, 29125, USA
NEIL R MILLER
The Wilmer Institute
The Johns Hopkins Hospital,
Baltimore, Maryland, 21205 USA
Correspondence to: Dr Miller.

1 Stone TT. Peripheral facial palsy. Multiple attacks in three brothers. JAMA 1950; 143:1154-5.

Transient amnesia heralding brain stem infarction

Transient global amnesia (TGA) is a syndrome of acute, transient memory disturbance with severe anterograde and retrograde amnesia but no neurological signs and preservation of personal identity. Vascular, epileptic, migraine and hypoglycaemic attacks have been implicated in its aetiology.1,2 A recent case control study has shown that most episodes of TGA are not associated with risk factors for ischaemic cerebral disease.3

Whilst ischaemic and hypoglycaemic syndromes, previous reports of apparent TGA as a manifestation of transient ischaemic attacks, have rarely included witnessed accounts of the ischaemic events.4

A 46 year old man presented having had five episodes of vertigo, each lasting for several hours, over the previous six months. Examination between the attacks was normal apart from mild hypertension and marked symptoms of anxiety. When seen as an outpatient routine investigations were unremarkable. There was no evidence of a peripheral lathamitary disturbance and the brainstem evoked potentials, CT and MRI scans were normal. He was referred for a psychiatric opinion. There was no previous psychiatric history, although over the previous months, following his divorce and threats to his job, he had become anxious about his future. He was known to have drunk heavily in the past but had not done so in the previous year.

Three days later, when attending the outpatient department, he suddenly complained of memory loss. This episode lasted for 5 hours during which time psychometric assessment illustrated the fluctuating nature of the memory deficit. He initially appeared agitated and bewildered, repeatedly asking the date, where he was and why he was there. He was alert and could recall his name and date of birth. One hour later a neuropsychological assessment showed that he performed satisfactorily on a relatively easy, three choice recognition memory test comprising coloured photographs. His ability to recognise and name pictured objects was within the average range. Further testing was not possible as he began to complain of blurred vision and loss of memory. On questioning during the next half an hour he appeared to experience total lapses in cognition and memory with reasonable accounts of his present circumstances. Two hours later there was a profound anterograde amnesia and patchy retrograde amnesia for events that were no other abnormal neurological signs. An EEG performed at the height of his symptoms showed a few slow wave transients in the fronto-temporal region. On the following morning (that is, 14 hours later) his ability to recall new information had returned to normal.

There remained, however, an amnesic gap for a short period of his train journey to the hospital on the previous day.

Three days later, while still in hospital, he had a brief episode of dysphasia, difficulty in understanding the tongue, weakness and clumsiness of the left arm. There were no neurological abnormalities when he was examined shortly after this episode, which was diagnosed as a transient ischaemic attack. Towards the end of the week he had a further transient episode of dysarthria and right hemiparesis, again with no signs on examination immediately after the event.

Twenty four hours later he became drowsy and developed periods of sleep in which he had evoked nystagmus to the right with delayed adduction of the right eye on leftward gaze, preserved upgaze with upbeat nystagmus, contraband nystagmus, dissociated gaze palsy, and right facial weakness. He exhibited diminished pharyngeal reflex, slow tongue movements and a dense right hemiparesis. CT scan showed low attenuation in the right inferior cerebellar hemisphere and right occipital lobe consistent with infarction. MRI confirmed the diagnosis of brainstem stroke, with extensive high signal at the right cerebellar hemisphere, brain stem and medial part of right occipital lobe suggestive of occlusion of right posterior inferior cerebellar artery and right posterior cerebral artery.

There was gradual neurological recovery over two months. He often appeared disoriented when discussing events during this period but his mental state was otherwise considered to be normal and he performed satisfactorily on both verbal and visual recognition memory tests.

This patient presented with an episode of amnesia with features of TGA.1 There was a witnessed attack of definite amnesia, resolving within 24 hours, without disturbance of consciousness, focal neurological signs, epileptic features or a recent head injury. During the attack the patient developed a clear anterograde amnesia with disturbance of long term memory without loss of personal identity, complex cognition or language.5 He also exhibited the phenomenon of "meta-memory", an awareness that memories ought to be readily recalled. Following recovery a small gap of retrograde amnesia remained.

The newly neurologically impaired patient was transiently blured vision but examination was normal during the episode.

This case serves to emphasise that transient amnesia resembling TGA may occur as a manifestation of transient ischaemic attacks in the vertebrobasilar territory and that there is a risk of subsequent major ischaemic deficit.

R S HOWARD
R FESTENSTEIN
M RON
The National Hospital for Neurology and Neurosurgery, London, UK

Correspondence to: Dr Howard, The National Hospital for Neurology and Neurosurgery, Queen Square, London WC1N 3BG.

Familial recurrent cranial nerve palsy.

K C Golnik and N R Miller

*J Neurol Neurosurg Psychiatry* 1992 55: 976-977
doi: 10.1136/jnnp.55.10.976-a

Updated information and services can be found at:
http://jnnp.bmj.com/content/55/10/976.2.citation

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Notes**

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/