Letters

Chronic focal polymyositis

Sir: The weakness in polymyositis is usually described as symmetrical and mainly proximal. Bharucha and Morgan-Hughes1 mentioned four patients with chronic focal polymyositis, in whom highly selective muscle wasting and weakness remained confined to the forearm flexors, brachio radialis and quadriceps muscles. We would like to report a patient presenting with chronic bulbar palsy.

In April, 1983, a 57 year old man presented with a 4 month history of slowly progressive dysphagia. The difficulty was associated with drinking liquids. Some of the liquid escaped through the nose and coughing or choking was frequent while drinking. On examination, his voice was soft and hypernasalised. Voluntary elevation of the palate was incomplete and the gag reflex was weak. The tongue was atrophic without fasciculation. Neck weakness was prominent, with atrophy of the sternocleidomastoid muscles. There was a positive snout reflex but absent jaw jerk. Ocular movements were full and no weakness was observed in the facial muscles and the four extremities. The deep tendon reflexes were hyporeactive in the upper limbs and normal in the lower limbs. The serum creatine kinase (CK) was 56 IU/l (normal, 47 to 210). Electromyography showed fibrillation potentials in the sternocleidomastoid muscles but little abnormality in the individual motor units. Muscle biopsy was not performed. Wasting and weakness persisted, confined to the bulbar and neck muscles. The patient was tube-fed but suffered from recurrent aspiration pneumonia between September, 1983 and April, 1986. In 1987, minimal weakness in the deltoid muscles was noted. A biopsy of the left deltoid muscle showed marked variation in muscle fibre size with large collections of inflammatory cells which were mainly interstitial. A small number of regeneration fibres were present. The CK was 57 IU/l. The patient was treated with prednisone 60 mg daily and during the next 3 months there was improvement in swallowing. He became able to drink liquids without nasal regurgitation and aspiration.

In polymyositis dysphagia is unusual as the first symptom and prolonged bulbar palsy without limb girdle weakness has rarely been reported.2,3 Our patient with morphologically proven polymyositis had selective bulbar palsy of 4 years' duration. The symptoms improved following steroid therapy. Ueno et al4 described a remarkably similar patient who had dysphagia of 2 years' duration and showed a good response to steroids. Although CK is useful in diagnosis and the assessment of activity of polymyositis, the enzyme was normal both in our case and the patient described by Ueno et al.

Polymyositis is one of the few neuromuscular diseases in which drug therapy is of value. One should therefore be alert to the possibility and be aware of unusual manifestations of the disease. Chronic focal polymyositis should be considered in the differential diagnosis of bulbar palsy.

SHOUSAUKI NODA
HIROTOSHI UMEZAKI
HIROAKI ITOH
KENJI HIROMATSU
Department of Neurology,
Kyushu-Kosei-Nenkin Hospital,
Kishinoura, Yahata-Nishiku,
Kitakyushu, 806, Japan

TATSUNORI YAMAMOTO
Department of Neurology,
University of Occupational and
Environmental Health,
School of Medicine,
Kitakyushu, 806, Japan

References


False positive diagnosis of phaeochromocytoma in a patient with Parkinson's disease receiving levodopa

Sir: Phaeochromocytoma may present in patients aged over 70 years, but is uncommon at this age.1 We here report the case of a 71 year old lady who narrowly escaped an unnecessary laparotomy because of a false-positive diagnosis of phaeochromocytoma.

At age 66 this lady developed dragging of her left leg and a mild postural tremor of the left hand. Parkinson's disease was diagnosed, and for 18 months a low dose of Sinemet (levodopa with carbidopa) virtually abolished her symptoms. At this time, she began to develop throbbing headaches which would begin 45 minutes after each dose, and caused her to limit her levodopa intake, with resultant worsening of mobility. At age 68 she was referred to our department. On examination, intellectual function was normal, and there were no symptoms of autonomic impairment. Eye movements were normal. In the OFF condition there was marked bilateral akinesia, moderate rigidity, and a fine postural tremor. Total KCH disability score was 40, which fell to 22 on higher doses of Sinemet. Autonomic function tests off treatment showed evidence of a partial vagal neuropathy, but with a normal Valsalva ratio and an absence of postural hypotension. A clinical diagnosis of idiopathic Parkinson's disease with a mild autonomic neuropathy and dose-related wearing-off phenomenon was made, and anti-parkinsonian treatment needed repeated titration over the next 2 years.

By mid-1985, now aged 70, she had developed new symptoms. In addition to her headaches, she also complained of facial flushing, and severe burning and prickling dysesthesiae affecting the entire body. These were present during OFF periods, exacerbated at the beginning and end of action of individual doses of levodopa (coinciding with an increase in dystonic dyskinesias), but considerably relieved at the peak of action of her dosage. There was no evidence of depression, and the pain was resistant to numerous treatments including amitriptyline and lithium, and persisted throughout a 5-day levodopa "holiday". She was referred to a pain clinic, but then lost to follow-up when, in January 1986, she was admitted to another hospital with a fractured left femur and given an Austin-Moore arthroplasty. Four months later, she was admitted to the same hospital, and her treatment was changed to Sinemet 110 tds, with the addition of selegiline 10 mg mane. On account of her complaints of facial flushing and somatic pain, two 24 h urinary vanillylmandelic acid (VMA) estimations were made, using the standard Pisano method:2 both were raised (to 119 and 125 μmol/24 h; normal range for the laboratory less than 35). Renal ultrasound was normal. CT body scan was reported as showing possible enlargement of the right adrenal gland (later reviewed and considered to show a partial volume effect of the right kidney). The patient was therefore re-admitted for removal of a right adrenal phaeochromocytoma, and started pre-operatively on phenoxybenzamine. The evidence for a phaeochromocytoma was reviewed, and further investigation was felt to be indicated. Twenty four hour urinary 5-hydroxyindole acetic acid levels and iodine-131-metiodobenzylguanidine scan were both normal. Whilst still taking Sinemet 110 tds, a pentoctinolinium suppression test was performed.

728
Chronic focal polymyositis.

S Noda, H Umezaki, H Itoh, K Hiromatsu and T Yamamoto

J Neurol Neurosurg Psychiatry 1988 51: 728
doi: 10.1136/jnnp.51.5.728

Updated information and services can be found at:
http://jnnp.bmj.com/content/51/5/728.1.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/