Stroke due to atrial myxoma in a young woman with co-existing acoustic neuroma and pituitary adenoma

Sir: Cardiac myxomas are uncommon benign tumours with a gelatinous friable consistency. They may present in a wide variety of ways and are difficult to diagnose clinically owing to the lack of specific signs.1 The majority present with features of atrioventricular valve obstruction, systemic embolism, or constitutional disorder such as fever, malaise, leucocytosis, and an elevated erythrocyte sedimentation rate. Seventy-five per cent occur within the cavity of the left atrium, when systemic embolism is a major risk.2 Emboli to a wide variety of sites have been reported, some being massive, and complications include infarction and mycotic aneurysm formation.3 Cerebrovascular embolism is a relatively common mode of presentation.4

A 28-year-old woman was referred three weeks after suddenly developing slurred speech, diplopia, paraesthesiae on the left side of the face, and weakness of the left arm. She had vomited initially and developed a left sided headache later. Poor hearing in the left ear was noticed following a head injury three years before this presentation. Otherwise she had recently been well, was on no medication and did not smoke. Examination revealed acromegalic facies and hands. There were splinter haemorrhages in the fingers. She was normotensive and in sinus rhythm. Neurological examination revealed nerve deafness in the left ear, and mild left finger-nose ataxia. Power was reduced in the left leg, and the left calf was wasted following childhood poliomyelitis. Clinically a brainstem stroke was the initial diagnosis, but plain and computed tomography showed a large left acoustic neuroma complicated by hydrocephalus, and enlargement of the pituitary fossa. Following a decompressive shunt the acoustic neuroma was removed, and transtheoidal hypophysectomy was performed for a chromophobe adenoma during a later admission. Her subsequent progress was good and she returned to her job in Greece.

Two years later she was again referred, after suffering a major dominant hemisphere stroke, with dysphasia, facial weakness and a dense right hemiplegia. The heart was clinically normal, and the electrocardiograph within normal limits. Computed tomography showed a left hemispheric infarct, and angiography showed complete occlusion of the left internal carotid artery. In view of the probable embolic nature of the stroke and the history of splinter haemorrhages, echocardiography was performed and demonstrated a large left atrial myxoma. This was removed under cardiopulmonary bypass, and since then she has been making a slow but encouraging recovery from her stroke.

The initial clinical diagnosis in this case was brainstem cerebrovascular accident, and retrospectively this is still possible although the acoustic neuroma and hydrocephalus were the likely cause. Because of the rarity of cardiac myxoma little is known about associations with other conditions. In addition it does not qualify for registration in cancer registries which are gathering valuable epidemiological information on malignant tumours, including the coincidence of multiple primaries.4 Pituitary adenomas and acoustic neuromas occur as elements of multiple tumour complexes, for example, in the multiple endocrine adenopathy syndrome and neurofibromatosis respectively. However, although there is an association between cardiac myxoma, pigmented skin lesions and myxoid neurofibromata,5 there is no recognised association of these myxomas with other primary tumours.

When a cardiac cause for systemic or cerebral emboli is suspected, two dimensional echocardiography provides a reliable and noninvasive investigation which may demonstrate treatable causes, including endocarditis, mural thrombus, mitral stenosis and cardiac myxoma. However the yield is small unless there is already clinical evidence of heart disease.6 Even in this selected group, few patients have a myxoma; one case was found in 138 patients with suspected focal cerebral ischaemia,7 and one in 110 patients referred with suspected systemic embolism.8 Maroon and Campbell discussed features which should alert the physician to the presence of a cardiac tumour in patients with cerebral embolism: (1) a patient aged under 30 years in whom endocarditis has been excluded; (2) normal cardiac rhythm and no history of cardiac disease; (3) a cardiac dysrhythmia or murmur which varies with time and posture; (4) hyperglobulinaemia, elevated sedimentation rate, anaemia and leucocytosis with constitutional symptoms; (5) myxomatous tissue in an excised embolus; and (6) the finding of a filling defect or pseudoaneurysm on cerebral angiography. Now that surgery provides a potential cure for cardiac myxoma,9 although follow up is advisable, it is vital that the diagnosis is considered and confirmed as early as possible.

The neurosurgical operations in this case were performed by Mr MJ Torrens, and the cardiac surgery by Mr G Keen.

P GORMAN
R LANGTON HEWER
Department of Neurology,
Frenchay Hospital,
Bristol BS16 1LE. UK

References
Letters


Accepted 23 November 1984

Myasthenia gravis associated with a hormone producing malignant carcinoid tumour

Sir: Myasthenia gravis is often associated with thymic hyperplasia or thymoma,1 and an increased incidence of non-thymic malignancy has been reported.2 We describe a patient who developed myasthenia gravis 1 year after the diagnosis of a malignant carcinoid tumour had been made and 4 years after the onset of symptoms.

A 77-year-old retired insurance salesman was admitted to another hospital in March 1983 with acute urinary retention due to an enlarged prostate. Over the preceding 3 years the patient had complained of episodic diarrhoea, without the passage of blood or mucus, and accompanied by abdominal colic. He gave no history of flushing attacks or wheezing. Investigations over this period including sigmoidoscopy and barium enema examination were normal. A gastroscopy had demonstrated a small polyp which was removed and found to consist of hypertrophic gastric mucosa. The patient had undergone bilateral hernia repairs 15 years previously and a mastoid operation 10 years before that. Otherwise he had been well and took no medication. Transurethral resection of the prostate was performed, and histology showed benign prostatic hypertrophy. After operation he developed small bowel obstruction and a laparotomy was carried out. This revealed small bowel obstruction due to a tumour in the proximal ileum, with peritoneal seedings and enlarged mesenteric lymph nodes, but no obvious liver involvement. Resection included 1 m of ischaemic small bowel with end to side jejunoo-ileal anastomosis. Histology of the resected tumour was characteristic of a mid-gut carcinoid tumour resulting in ischaemic mucosal necrosis of the small bowel. In both the bowel mucosa and mesentery clumps of uniform tumour cells containing red granules staining positively with the diazonium technique were present.

Fourteen months later he was referred to this hospital because of progressive weakness in the trunk, neck, and limbs developing over several months. For 4 weeks prior to admission he had noticed ptosis, double vision, difficulty in chewing and swallowing. Examination showed bilateral ptosis, worse on the right, limitation of eye movements and diplopia in all directions, an inability to close his jaw fully and bilateral facial weakness. Weakness of his neck, trunk, and limb muscles was such that he was unable to stand or maintain his head in an upright position. These symptoms and signs were improved dramatically by an intravenous injection of edrophonium and treatment with pyridostigmine was started.

Investigations were as follows: haemoglobin 9-5 g/dl, a dimorphic blood film picture, serum iron 2-5 μmol/l, Total Iron Binding Capacity 72·6 μmol/l, % saturation 3·4%, positive faecal occult blood, serum B12 25 ng/l. A Schilling test showed reduced absorption of B12 both with and without intrinsic factor. Urinary 5-hydroxyindoleacetic acid (5 HIAA) excretion 205 μmol/24 h (normal less than 75 μmol/24 h), gamma GT 76 u/l (normal less than 50 u/l). Auto antibodies to striated muscle negative, to acetylcholine receptor 4·9 nmol/l (normal controls less than 0·2 nmol/l), and to gastric parietal cell weakly positive. Rapid Plasma Reagin Test and Total Iron Binding Capacity negative, with a chest radiograph, isotope liver scan, urea and electrolytes, thyroid function tests, serum folate, serum aldolase and creatine kinase all normal. Electrophysiological studies showed normal motor and sensory conduction in the right median nerve. With repetitive stimulation of the right ulnar nerve at the wrist recording from right adductor digiti minimi showed initial response with amplitude 11·2 mV and classical myasthenic decrement at stimulation rates from 1–10/Hz. The decrement of the 4th response was as follows; 1/Hz 10·5%, 2/Hz 39·3%, 3/Hz 39·3%, 5/Hz 28·3%, 10/Hz 24·7%. Stimulating at 50Hz for 128 stimuli produced a small increment of the second response and then a progressive decrement to under 50% of the initial amplitude. Microscopic examination of muscle obtained at biopsy of palmarus longus showed variation in size of muscle fibres which were generally slightly smaller than normal. There was no significant small fibre grouping, no lymphorrhages and no inflammation. Histiochemistry showed no fibre type preponderance, degeneration, nor fibre type grouping. Methylene blue staining of motor end plates showed elongation and branching of terminal expansions compatible with myasthenia gravis.

The patients anaemia was treated initially with oral iron and parenteral hydroxyecobalamin. Prednisolone and azathioprine were started; his condition improved gradually over the next 2 months and he was able to leave hospital 11 weeks after admission.

Myasthenia gravis is associated with a higher than expected incidence of non-thymic malignancy. A retrospective study of 1243 patients with myasthenia gravis revealed 94 non-thymic neoplasms, most commonly affecting breast, lung, colon and rectum.2 In particular the incidence of malignancy within 2 years of the diagnosis of myasthenia gravis was much greater than expected. Thymic disorders remain the most consistently recognised abnormalities associated with myasthenia, thymic germinal centres or hyperplasia being present in 65% of cases and a further 10% having a thymoma.1 Thymic carcinoid should be differentiated from thymoma, having a significant malignant potential, being associated with Cushings syndrome, and occurring with multiple endocrine adenomatosis.3

Features of the carcinoid syndrome occur in about 10% of carcinoid tumours, usually those in the jejunum and ileum.4 Systemic symptoms, usually attacks of diarrhoea, abdominal colic, flushing, or wheezing are due to the production of a variety of secretory products, most often 5-hydroxytryptamine (5-HT) and kinins. A proximal myopathy in some patients with the carcinoid syndrome has been attributed to excess 5-HT,5 and thus treated with cyproheptadine.

The patient we describe complained of attacks of diarrhoea and abdominal colic for 3 years before the diagnosis of a carcinoid tumour became evident after laparotomy, when an excess urinary excretion of 5-HIAA was demonstrated. Just over 1 year later he developed myasthenia gravis. He did not have a carcinoid myopathy and muscle biopsy changes were compatible with myasthenia gravis. When myasthenia gravis develops in association with a malignant tumour weakness may be wrongly attributed to the malignancy or general debility, the diagnosis missed, and the patient denied effective treatment for his condition.
Stroke due to atrial myxoma in a young woman with co-existing acoustic neuroma and pituitary adenoma.

P Gorman and R L Hewer

*J Neurol Neurosurg Psychiatry* 1985 48: 718-719
doi: 10.1136/jnnp.48.7.718

Updated information and services can be found at:
[http://jnnp.bmj.com/content/48/7/718.citation](http://jnnp.bmj.com/content/48/7/718.citation)

**Email alerting service**

These include:

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](http://group.bmj.com/group/rights-licensing/permissions)

To order reprints go to:
[http://journals.bmj.com/cgi/reprintform](http://journals.bmj.com/cgi/reprintform)

To subscribe to BMJ go to:
[http://group.bmj.com/subscribe/](http://group.bmj.com/subscribe/)