Downloaded from http://jnnp.bmj.com/ on December 9, 2016 - Published by group.bmj.com

also improved. Two weeks after operation her serum thyroxine rose to 72 nmol/l; basal TSH was minimally elevated at 6·5 mu/l and after 200 μg TRH, rose to 9·8 mu/l at 20 minutes. Basal prolactin was elevated at 38 μg/l. Her basal LH (4 u/l) and FSH (2 u/l) showed no response to 100 μg LHRH, and her plasma oestradiol was undetectable. During an insulin tolerance test her plasma glucose fell to 1·2 mmol/l and her basal morning plasma cortisol of 175 nmol/l rose to 420 nmol/l, a substantial response. During a water deprivation test her plasma osmolality rose to 303 mosm/kg, her urine rising to a maximum of 404 mosm/kg indicating diabetes insipidus. Two months after operation her serum thyroxine had risen to 85 nmol/l and TSH fallen to 1·6 mu/l. She had mild symptoms of diabetes insipidus and still had amenorrhoea with galactorrhoea. Her basal prolactin was elevated at 28 μg/l. Two months later her thirst and polyuria had improved, though she still had no periods and her prolactin remained elevated at 51 μg/l, whilst at 8 months she had no further symptoms attributable to diabetes insipidus although her galactorrhoea and amenorrhoea remained.

Isolated granulomatous lesions of the pituitary are rare and the differential diagnosis includes sarcoidosis, tuberculosis, syphilis and giant cell granuloma.[2] Although acid fast bacilli were not seen in this case, the patient's past history, results of investigation and improvement with anti-tuberculous treatment made a pituitary tuberculoma the likely diagnosis.

There have been few reports of pituitary tuberculoma in the past[3-5] although there have also been reports of giant cell granuloma of unknown origin[6-8] which might represent a solitary tuberculoma. Hassoun *et al*[8] claimed that the diagnosis of a pituitary granuloma is suggested by marked hypopituitarism, out of proportion to the size of the tumour. Clearly in our case, there was significant suprasellar enlargement, and the degree of hypopituitarism was not inconsistent with this.

Our patient presented as a neurosurgical emergency and although the diagnosis of tuberculoma was not considered preoperatively, the appropriate treatment was operative decompression of the optic chiasm. Post-operatively she still has some endocrine dysfunction as shown by the mild hyperprolactinaemia, gonadotrophin deficiency and mild diabetes insipidus. However, her hypothyroidism has improved, and although we do not have a pre-operative insulin stress test for comparison of dynamic pituitary adrenal function, morning cortisols have returned towards normal.

The potential importance of recognising a tuberculoma as a possible cause of a space occupying lesion within the pituitary fossa is that it may be treated conservatively with less hazard to the patient than operative intervention. Clearly, however, if the patient presents as a neurosurgical emergency, there may be no alternative to surgical decompression.

We thank Mr A Crockard who performed the trans-sphenoidal surgery on this patient and Dr RD Barnard for the pathological interpretation, and Professor WI McDonald for allowing us to report his patient.

DJA ECKLAND
JH O'NEILL*
SL LIGHTMAN
Medical Unit,
Charing Cross and Westminster
Medical School,
Westminster Hospital,
Page Street,
London SW1P 2AP
and
The National Hospital for Nervous
*Diseases**
Maida Vale,
London W1, UK

References

1 Maurice-Williams RS. Tuberculomas of the brain in Britain. *Postgrad Med J* 1972;**48**: 678–81.
2 Richards AG, Harvey PW. "Giant Cell Granuloma" and other pituitary granulomata. *Q J Med* 1954;**23**:425–39.
3 Coleman CC, Meredith JM. Diffuse tuberculosis of the pituitary gland simulating tumor, with post-operative recovery. *Arch Neurol* 1940;**44**:1076–85.
4 Brooks MH, Dunlao JS, Bronsky D, Waldstein SS. Hypophysial tuberculoma with hypopituitarism. *Am J Med* 1973;**54**:777–81.
5 Sheehan HL, Summers VK. The syndrome of hypopituitarism. *Q J Med* 1949;**18**:319–78.
6 Taylor C, Duff TA. Giant cell granuloma involving the pituitary gland. Case Report. *J Neurosurg* 1980;**52**:584–7.
7 Delpozo JM, Roda JE, Montaya JG, Inglesias JR, Hurtado A. Intrasellar granuloma. *J Neurosurg* 1980;**53**:717–9.
8 Hassoun P, Anayssi E, Salti I. A case of granulomatous hypophysitis with hypopituitarism and minimal pituitary enlargement. *J Neurol Neurosurg Psychiatry* 1985;**48**:949–51.

Accepted 26 July 1986

Protein C deficiency: a cause of amaurosis fugax?

Sir: We evaluated a 45 year old man who was admitted to hospital with a 2 day history of recurring left eye symptoms. He had experienced more than a dozen episodes of transient visual loss. He described "a shade coming down", sometimes over the entire left eye and sometimes over only the upper or lower half-field. His symptom lasted for 5 to 10 minutes. He had no history of migraine and there were no associated symptoms suggestive of migraine. A left Marcus-Gunn pupil was noted in the emergency room during an episode of amaurosis, but the neurological examination after admission was normal. Cardiac and vascular exams were normal except for chronic venous insufficiency in the legs. There was no evidence of acute deep venous thrombosis. Examination by a neuro-ophthalmologist revealed no abnormalities. The patient's medical history was unremarkable except for recurring thrombophlebitis in the legs. He had no history of hypertension, cardiac disease or diabetes mellitus. He smoked one-half pack of tobacco daily. He took no medication. His family history was noteworthy: both father and sister had a history of recurring thrombophlebitis.

Normal studies included: complete blood count, platelet count, biochemical survey, prothrombin time, partial thromboplastin time, platelet aggregation studies, serum antithrombin, erythrocyte sedimentation rate, sonoclot, fibrin split products and fibrin monomer. Cranial CT scan, electrocardiogram, echocardiogram and left carotid angiogram (including views of the ophthalmic artery) were normal. Protein C determination by immunological testing was abnormal at 46% (normal 70–180%). Testing of the patient's family disclosed similarly low values of protein C for five of 10 blood relatives. He was treated with coumadin and has been symptom-free in one year of follow-up.

Amaurosis fugax has been reported for more than 100 years. In 1952, Fisher [1] drew attention to the aetiological role of retinal emboli from the ipsilateral carotid. Others[2] have emphasised the heart as a source of emboli. Yet, in some series, 25–50% of the patients have neither carotid nor cardiac disease.[3,4] Reported additional mechanisms for amaurosis fugax include ocular disorders, vasculitis, platelet hyperaggregability, hypercalcaemia, myeloproliferative disorders, sickle cell disease, multiple myeloma, carotid artery trauma and dissection,

Downloaded from http://jnnp.bmj.com/ on December 9, 2016 - Published by group.bmj.com

migraine, Raynaud's disease, papilloedema, ophthalmic artery atheromata and osteoma. To our knowledge, no case has been ascribed to protein C deficiency.

Protein C is a vitamin K dependent plasma glycoprotein. When activated, it inhibits coagulation by inactivating factors V and VIII. It facilitates fibrinolysis by elevating plasminogen activator levels. Deficiency is inherited as an autosomal dominant trait. Since 1981, a number of families have been reported to have protein C deficiency and recurring thrombo-embolic events. Like antithrombin deficiency, protein C deficiency is typically associated with venous thrombosis and pulmonary emboli. The problem has generally not been associated with arterial disease or neurological symptoms. Recent reports indicate, however, that deficiency of either protein C or serum antithrombin can cause stroke by venous thrombosis.[5 6] Paradoxical embolism from a venous source might also produce neurological symptoms in this setting. A causal relationship between protein C deficiency and amaurosis fugax cannot be proved in our patient, but we submit that the connection is plausible.

Protein C deficiency may go unrecognised for many years and does not inevitably present thrombotic complications. Some patients with amaurosis fugax have no clear explanation for their symptoms. Some of them may have hypercoagulable states characterised by deficiencies of clotting inhibitors. If so, appropriate laboratory screening may lead to a more definitive diagnosis and therapy for these individuals.

DON B SMITH, MD
601 E Hampden Ave, Englewood, Colorado 80110
GORDON E ENS, MT(ASCP)
Colorado Coagulation Consultants Inc, 800 Clermont St, Denver, Colorado 80220, USA

References

1 Fisher M. Transient monocular blindness associated with hemiplegia. *Arch Ophthalmol* 1952;**47**:167–203.
2 Zimmerman LE. Embolism of central retinal artery secondary to myocardial infarction with mural thrombosis. *Arch Ophthalmol* 1965;**73**:822–6.
3 DeBono DP, Warlow CP. Potential sources of emboli in patients with presumed transient cerebral or retinal ischaemia. *Lancet* 1981;**1**:343–5.
4 Sorenson PN. Amaurosis fugax: unselected material. *Acta Ophthalmol* 1983;**61**:583–8.
5 Wintzen AR, Broekmans AW, Bertina RM, *et al*. Cerebral haemorrhagic infarction in young patients with hereditary protein C deficiency: evidence for "Spontaneous" cerebral venous thrombosis. *Br Med J* 1985;**290**:350–2.
6 Ambruso BR, Jacobsen J, Hathaway WE. Inherited antithrombin III deficiency and cerebral thrombosis in a child. *Pediatrics* 1980;**65**:125–31.

Accepted 20 July 1980.

Exercise provoked faecal incontinence in spinal stenosis

Sir: Lumbar canal stenosis, first delineated by Verbiest in the 1950s,[1] has been linked for many years with intermittent neurological symptoms in the lower limb. Blau and Logue, in 1961[2] introduced the term "claudication of the cauda equina", because of similarities with symptoms caused by peripheral vascular disease in the legs.

Symptoms classically associated with lumbar canal stenosis include pain, paraesthesiae and/or weakness in the legs, occurring on walking or sometimes with a change in posture, which are relieved by rest, and are associated with minor sensorimotor asymmetry but normal pedal pulses. We report a case of faecal incontinence provoked by exercise in a patient with lumbar canal stenosis, whose symptoms remitted following lumbar decompression. Such an association has not previously been described.

Mr H.H. first presented in April 1979 aged 62 years, with a three year history of dragging of the right leg occurring after he had walked several miles. In March 1978 he became suddenly impotent, and had noticed increased leg weakness since that time. He had had low back pain for 10 years but was otherwise asymptomatic, and had suffered no bladder or bowel disturbance.

Examination revealed reduced muscle bulk in the right thigh, and increased right knee and ankle reflexes, with normal sensation and intact cremasteric and anal reflexes. CSF studies and visual evoked responses were unhelpful, and metrizamide myelography showed a small arachnoid cyst posteriorly in the mid-dorsal region, with minor disc prolapse at the L 4/5 level. He was followed up in outpatients.

He re-presented in early 1985 with a two year history of faecal incontinence provoked by walking. Initially this only occurred if he walked several miles, but the distance had gradually decreased to 400 yards and he was forced to wear incontinence pads throughout the day. He was fully continent at rest. His right leg felt stiffer than before, but he had no urinary or other symptoms.

Examination revealed decreased muscle bulk in the right thigh as before, but there were no new sensory or motor signs, and anal sphincter tone was normal at rest. The distressing nature of his symptoms precluded provocation testing.

A cerebral CT scan was unremarkable. Iohexol myelography showed a greater than 50% reduction in the sagittal diameter of the subarachnoid space at L 4/5, associated with disc degeneration (see fig). Lesser narrowing of the spinal canal due to disc disease was also present at L 3/4. There was no significant abnormality in the dorsal and cervical regions, and the dorsal arachnoid cyst was unchanged. A lumbar decompression was performed, with removal of the spinous processes and laminae of L 4/5 and the upper part of S 1. At operation gross osteoarthropathy and hypertrophy of the ligamentum flavum was noted. The postoperative period was uneventful. One month after decompression the patient was able to walk more quickly than he had for three years. He had not been faecally incontinent for several weeks, and was confident enough to have discarded his incontinence pads. Three months later he had had no further episodes of incontinence, and his only residual symptom was slight stiffness of the right leg.

While exercise provoked symptoms in the legs related to stenosis of the lumbar canal are well described, the association of incontinence with the syndrome is less certain. Sharr, Garfield and Jenkins[3] found radiological evidence of lumbar stenosis and symptoms of "neurogenic claudication" in some women with urinary incontinence aggravated by exercise and posture. However, Hawkes and Roberts[4] found a 50% incidence of urological symptoms both in men and women with "neurogenic claudication" and equally in a control group with peripheral vascular disease.

Faecal incontinence has not previously been reported in this context, and its occurrence in our patient requires explanation. Maintenance of faecal continence is now thought to depend chiefly on the puborectalis muscle, which acts as a sling round the distal rectum. Although the internal smooth muscle sphincter makes the largest contribution to intraluminal rectal pressure, its division causes little functional disability.[5] Both the puborectalis and the external sphincter muscles are supplied by S 2–4 nerve roots and show tonic activity which ceases during defaecation.[6 7] La Fuente, Andrew and Joy[8] have shown in a cadaver

Downloaded from http://jnnp.bmj.com/ on December 9, 2016 - Published by group.bmj.com

Protein C deficiency: a cause of amaurosis fugax?

D B Smith and G E Ens

J Neurol Neurosurg Psychiatry 1987 50: 361-362
doi: 10.1136/jnnp.50.3.361

Updated information and services can be found at:
http://jnnp.bmj.com/content/50/3/361.citation

These include:

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/