potential importance we have attempted to repeat this study using the traditional indirect immunofluorescence on cryostat sections of fresh human pituitary obtained from surgical operations. Undiluted sera were applied and sheep anti-human total Ig and specific IgA (Wellcome) were used as conjugates at the proper dilution. Normal human pituitary obtained from a patient with breast cancer was used throughout our study. The presence of prolactin, growth hormone, LH, FSH, TSH cells as well as the specific pituitary microsomal antigen was confirmed on every 30 sections with specific monoclonal antibodies and with prolactin cell positive serum as a positive control. Sera were collected from 20 patients with clinically diagnosed Alzheimer's type senile dementia and three patients with Down's syndrome who had had intellectual deterioration. Diagnosis was made on historical evidence of at least six months duration of symptoms and all patients had a cranial CT scan, were examined neurologically and had psychometry. Patients with a history of stroke, head injury, alcoholism, epilepsy, major psychiatric illness were excluded, as were those with focal neurological signs, hypertension, metabolic or endocrine illness. The Alzheimer group were aged between 63 and 95 (mean age 80 ± 9 yr), with a duration of illness of between 6 and 72 months (mean duration 33 ± 18 m). The Down's group were in their early sixties (mean age 61 ± 1 yr) with a duration of dementia of between 12 and 18 months.

None of the sera from either group contained autoantibodies to prolactin cells, although six had gastric parietal cell, five had thyroglobulin autoantibodies (titres 1:80–1:320) and five were positive for thyroid microsomal autoantibody (titres 1:400–1:1,600). The reproducibility of our results in relation to the use of different substrates was checked by testing serum randomly selected from patients affected by several endocrinopathies and normal controls on human, baboon and monkey pituitary sections. As shown in the table, sera tested on baboon and human pituitary gave virtually identical results for pituitary endocrine cell autoantibody screening, whereas the same sera tested on monkey

pituitary showed a much higher number of positive reactions. We postulate that although rhesus monkey is close to the human, some human gamma-globulins could show a heterophile specificity with monkey pituitary. When in fact the 20 Alzheimer's and the three Down's syndrome sera were re-tested on monkey pituitary, 4/23 sera gave positive reactions on endocrine cells, two of which reacted with prolactin cells. Our failure to replicate Pouplard's results could be due to substrate differences since the French group used post-mortem human glands. Further studies are warranted in this area before firm conclusions can be reached regarding the role of prolactin cell antibodies as markers for unexplained cerebral atrophies.

M PHILPOT

J COLGAN

R LEVY

A HOLLAND

Institute of Psychiatry,

R MIRAKIAN

CA RICHARDSON

GF BOTTAZZO

Department of Immunology,

Middlesex Hospital,

London, UK

References

- Rossor MN. Dementia. *Lancet*, 1982; ii:1200–4.
- Marsden CD. The diagnosis of dementia. In: Isaacs AD, Post F, eds. *Studies in Geriatric Psychiatry*, 1978. New York: John Wiley.
- Goudsmit J. The Dementia syndrome, *Lancet* 1983;i:187.
- Cohen D, Eisdorfer C. Serum Immunoglobulins and cognitive status in the elderly. *Br J Psychiatry* 1980;136:33–39.
- Pentland B, Christie JE, Watson KC, Yap PL. Immunological parameters in Alzheimer's pre-senile dementia. *Acta Psychiat Scand* 1982;65:375–9.
- Pouplard A, Emile J, Vincent-Pineau F. Circulating human pituitary prolactin cell autoantibodies and Alzheimer's disease. *Rev Neurol (Paris)* 1983;139:178–91.
- Bottazzo GF, Pouplard A, Florin-Christensen A, Doniach D. Autoantibodies to Prolactin secreting cells of the pituitary. *Lancet* 1975;i:97–101.
- Bottazzo GF, Vendelli C, Mirakian R. The detection of autoantibodies to discrete

endocrine cells in complex endocrine organs. In: Pinchera A, et al, eds. *Autoimmune Aspects of Endocrine Disorders*. London: Academic Press, 1980:367–77.

- * Mirakian R, Cudworth AG, Bottazzo GF, Richardson CA, Doniach D. Autoimmunity to anterior pituitary cells and the pathogenesis of insulin-dependent diabetes mellitus. *Lancet* 1982;i:755–9.

Accepted 25 June 1984

Spinal angiomatosis presenting as a sensory neuropathy

Sir: Spinal angiomatosis¹ is a rare condition which usually presents with progressive symptoms suggesting a radiculomyelopathy. It may also cause subarachnoid haemorrhage.^{2,3} The unusual presentation of our patient with symptoms suggestive of a sensory neuropathy appears unique.

An Irish building labourer, aged 61 years, was admitted to hospital because of progressive tingling of his legs and unsteadiness in walking for ten days. He had not previously had any significant illnesses and there was no family history of neurological disease. There was no suggestion of any exposure to known neurotoxins. His alcohol consumption was considered moderate, but he smoked 80 cigarettes daily. He appeared unkempt, with markedly carious teeth. In other respects general medical examination was normal. His intellectual function had always been below average and he was vague about the details of his medical history. The cranial nerves and tone and power of his limb muscles were normal. All the tendon reflexes were reduced, while the right biceps, left knee and ankle reflexes were absent, but plantar responses were flexor. Modalities of sensation were impaired in the fingers as far as the metacarpophalangeal joints, and in the feet, as far as the ankles. Position sensation was impaired at the toes and ankles. He was unable to stand still with his eyes closed, and his gait was wide based and unsteady. Extensive haematological, biochemical and radiological investigation for a possible carcinoma gave normal findings. A CT scan of the brain was normal. Nerve conduction studies showed normal motor conduction velocities but reduced sensory action potentials (right radial 1 μ V; right sural 2 μ V; left sural absent). The F latency to the right tibial muscles was 34 ms and to the left abductor hallucis was 55 ms. Lumbar puncture revealed a clear fluid containing no cells

Table Unselected cases tested by IFL on three different pituitary substrates

Pituitary substrates	Total cases tested	Positive reactions
Normal human	100	17
Baboon	100	18
Rhesus monkey	51	28

and protein 1.6 g/l. Sural nerve biopsy showed gross loss of myelinated fibres and an excess of collagen. The appearance of axonal degeneration was confirmed by electron microscopy, which also showed occasional demyelinated axons. Sensory loss gradually became more severe and extended proximally. After one month the reflexes all disappeared, including the plantar responses which had remained flexor until that time. In addition, weakness of the intrinsic hand muscles and hip flexion developed. The neurological deficit continued to increase and, after three months, wasting of the intrinsic hand muscles, tibialis anterior and peroneal muscles had become evident. Finger clubbing had also developed. By this stage the right median motor conduction velocity slowed to 40 m/s, the median nerve action potential—which had been 11 μ V disappeared completely and the F wave latency remained 36 ms. A course of prednisolone and azathioprine failed to influence the progress of the condition. After six months, ataxia of the limbs became evident with pseudo-athetosis of the outstretched fingers. After eleven months he became incontinent of urine and after a year he died from a chest infection.

At necropsy severe bronchitis and bronchopneumonic consolidation were found in both lungs. In the ascending aorta there was a 1 cm horizontal tear, as is associated with dissecting aneurysm, but dissection had not occurred. Other systems were normal, apart from the nervous system to be described. The cerebrum, brain stem, cerebellum, pons, medulla and cranial meninges were normal on both gross and histological examination. The spinal theca was somewhat thickened, while a number of prominent tortuous vessels were noted under the leptomeninges covering the spinal cord from its mid cervical to lower thoracic regions. The spinal cord itself was distorted and softened in its mid thoracic part. The roots in the cauda equina were noted to be either of normal pale-cream colour or pinkish-grey in approximately equal proportion. The sciatic nerve was remarkably fibrous in texture and grossly resembled a frayed tendon.

Histology showed that both the spinal theca and leptomeninges were thickened: iron deposits (Perl's ferrocyanide test) in these membranes suggested that the thickening had resulted from previous haemorrhage. There was an excess of leptomeningeal vessels with arterial structure, while the veins were dilated, tortuous and markedly more collagenous (fig a) than

Fig. (a) Dilated leptomeningeal vessels over posterior surface of the cord. Note collagenous thickening of prominent veins and reduplication of the regional spinal artery. *Buk's modified Verhoeff elastic-Lendrum trichrome*, $\times 15$. (b) Thick-walled collagenised vessels of uncertain type in posterior column of cord. Note surrounding myelomalacia. Same stain as (a) $\times 60$.

normal veins around the cord. One of these leptomeningeal veins contained a small organising thrombus filling half its lumen. Numerous dilated tortuous thick walled small collagenous vessels were seen within the cord (fig b) and were particularly associated with the necrosis (myelomalacia) of the posterior columns. The upper thoracic cord was cavitated at its centre and resembled a syrinx but the cavity was not lined with epithelium and was clearly necrotic in origin. The region of the right anterior spinocerebellar tract showed a modest degree of pallor. Perl's ferrocyanide test was negative in the cord, indicating the absence of previous haematomyelia. The dorsal spinal roots contained frequent dilated sinuses lined by endothelium only; a considerable number (up to 90%) of nerve fibres were lost in many dorsal roots. Ventral spinal roots showed less loss of nerve fibres. The sciatic nerve examined was partly replaced by collagen and showed substantial fibre loss.

Spinal angiomas—originally described by Foix and Alajouanine¹ has been characterised in depth by others.²⁻⁶ The present case conforms pathologically to

these previous descriptions, but is presented here because of the unusual and unsuspected clinical presentation.

The disease usually presents as a progressive radiculomyelopathy at an average age of 40 years (range 21–75). It is two and a half times more common in men than in women. It is complicated by spinal subarachnoid haemorrhage in about a quarter of cases and sometimes by development of haematomyelia. It may be associated with capillary telangiectasia in the affected dermatomes.

Our patient's symptoms and signs were much more suggestive of a sensory neuropathy than spinal cord disease. In a heavy smoker the appearance of finger clubbing appeared to support a diagnosis of non-metastatic sensory neuropathy, but it is possible that the finger clubbing was related to the arteriovenous shunting in the spinal cord.

The pathological findings were typical of spinal angiomas. Most vein walls were markedly tortuous and collagenised or hyalinised. This, together with the apparent arterialisations of some veins, indicates that blood enters such angiomas at high pressure. However the "river-delta" haemodynamic effect of angiomas on the vessels might have reduced perfusion pressure in some parts of the tumour. Aminoff² suggested that the ischaemic necrosis (myelomalacia) of the affected areas of cord are caused by reduction in the arteriovenous gradient across the angioma. Thrombosis is a further factor, and one thrombosed vein was noted in the present case. Aminoff² further suggested that the shunt through the angioma may in effect "steal" from the cord. The relative sparing of the lateral column explains the lack of corticospinal tract signs in the developing stages of our patient's illness. In the later stages the associated degeneration of the dorsal, and to a lesser degree, ventral roots might have masked signs of a specific corticospinal tract deficit.

We thank Drs JC Houston and RK Knight for allowing us to report this case.

RAC HUGHES*

CWM ADAMS

Departments of Medicine* and Pathology,
Guy's Hospital Medical School,
London. SE1 9RT, UK

References

- 1 Foix C, Alajouanine T. La myélite nécrotique subaiguë. Myélite centrale angiohypertrophique à évolution progressive.

Paraplegie amyotrophique lentement ascendante, d'abord spasmodique, puis flasque, s'accompagnant de dissociation albumino-cytologique. *Revue Neurol (Paris)* 1926;33 (2):1-42.

² Aminoff MJ. *Spinal Angiomas*, Oxford: Blackwell, 1976:82-85.

³ Antoni N. Spinal vascular malformations (angiomas) and myelomalacia. *Neurology (Minneapolis)* 1962;12:895-904.

⁴ Greenfield JG, Turner JWA. Acute and subacute necrotic myelitis. *Brain* 1939; 62:227-52.

⁵ Hughes JT. *Pathology of the Spinal Cord*, 2nd Edition, London: Lloyd-Luke, 1978:81-83.

⁶ Jellinger K. Pathology of spinal vascular malformations and vascular tumours. In: Pia HW, Djindjian R, eds. *Spinal Angiomas. Advances in Diagnosis & Therapy*. Berlin: Springer-Verlag (1978):18-44.

three days after admission, being asymptomatic three months later. A further EMG study after 33 days evolution showed a clear reduction in spontaneous activity and a normal blink reflex.

Facial palsy in the course of cephalic tetanus usually appears when there is already evidence of trismus^{3,5,8} or simultaneously with it.^{4,6,9-11} Thus the interest in our case lies in the fact that facial paralysis was the only clinical sign for the first eight days of evolution, being diagnosed understandably as Bell's palsy. Although mistaken diagnoses of facial paralysis at the onset of cephalic tetanus have been mentioned occasionally,^{2,12} the pattern of evolution described here has only been reported in one case.¹⁰ This was a 14-year-old patient who developed left facial paralysis 3 days after suffering an eye injury on the same side; trismus and dysphagia were present the following day. Our EMG findings are consistent with those previously described.^{2,3,5,11} Consequently, when Bell's palsy and craniofacial injury are found together, the possibility of cephalic tetanus should be considered.

J MAYO

J BERCiano

Department of Medicine
(Section of Neurology),
NMC Marqués of Valdecilla,
Faculty of Medicine,
Santander, Spain

References

- ¹ Boudouresques J, Khalil R, Vigouroux RA, Daniel F, Goset A. Infectious diseases of nerves. In: Vinken PJ, Bruyn GW, eds. *Handbook of Clinical Neurology*. Vol 7. Amsterdam: North Holland Publishing Company, 1970:473-494.
- ² Vakil BJ, Singhal BS, Pandya SS, Irani PF. Cephalic tetanus. *Neurology (Minneapolis)* 1973;23:1091-1096.
- ³ Dastur FC, Shanani MT, Dastoor DH, et al. Cephalic tetanus: demonstration of a dual lesion. *J Neurol Neurosurg Psychiatry* 1977;40:782-6.
- ⁴ Farquharson RG. Cephalic tetanus. *Proc R Soc Med* 1972;65:875-6.
- ⁵ García Mullin R, Daroff RB. Electrophysiological investigations of cephalic tetanus. *J Neurol Neurosurg Psychiatry* 1973;36:296-301.
- ⁶ Abde VW, Dekate MP. Cephalic tetanus (clinical analyses of 9 cases). *J Indian Med Assoc* 1980;74:111-3.
- ⁷ Mishra M, Sinha BN. Facial palsy in tetanus. *Br Med J* 1972;3:475-6.
- ⁸ Ahuja GK, Kamala G. Cephalic tetanus. *Neurol India* 1978;26:10-13.
- ⁹ Bagratuni L. Cephalic tetanus with report of a case. *Br Med J* 1952;1:461-3.
- ¹⁰ Park DM. Cranial nerve palsies in tetanus and cephalic tetanus. *J Neurol Neurosurg Psychiatry* 1970;33:212-5.
- ¹¹ Mamoli B, Ludin HP. Electrophysiological investigations in a case of cephalic tetanus. *Neurol* 1977;214:251-5.
- ¹² Struppler A, Struppler E, Adams RD. Local tetanus in man. *Arch Neurol* 1963;8:102-78.

Accepted 6 August 1984

Accepted 6 August 1984

Cephalic tetanus presenting with Bell's palsy

Sir: Cephalic tetanus is a clinical variant characterised by paralysis of the cranial nerves.¹⁻⁵ Weakness of the muscles innervated by the seventh nerve is the most frequently reported symptom. However, cephalic tetanus commencing with facial paralysis, as in the case described here, is unusual.

A 53-year-old man suffered a wound on the right side of his chin, caused by a stick. Two days later, there was inflammation at the site of the injury and weakness of the right orbicularis oris muscle. Later the paralysis increased, with weakness of the ipsilateral orbicularis oculi, but without facial stiffness. On the fifth day, the patient was seen by his general practitioner who diagnosed right-sided Bell's palsy and prescribed prednisone 15 mg daily. Five days later, the patient had difficulty in swallowing and in opening his mouth, and was referred to our hospital. Examination revealed severe lockjaw and left facial hemispasm, combined with right facial paralysis, predominantly affecting the lower portion (fig). Taste was not affected. The wound was infected. Treatment included debridement of the wound, tracheotomy, penicillin, antitetanic gamma globulin, tetanus toxoid and diazepam. At rest, the electromyogram showed bilateral continuous abnormal activity in the masseters, orbicularis oris and orbicularis oculi muscles; electrical silence was not obtained. The patient began to improve

Fig Patient 20 days after onset of symptoms. Note the healed wound on the right side of the chin, right peripheral facial palsy and spasm of the left orbicularis oris and orbicularis oculi muscles.