nerosyphilis, the patient was treated with procaine penicillin. Four months later there had been no significant change in either symptoms or signs.

Ossification of the posterior longitudinal ligament is of unknown aetiology and may have an incidence as high as 1–3% in the Japanese who have cervical radiology. It may occur at any spinal level but is usually maximal in the cervical region. Although there is frequently no neurological involvement, the condition may cause a progressive myelopathy requiring decompressive laminectomy. As would be expected, myelopathy tends to occur with greater degrees of encroachment of the cervical canal. Severe myelopathy was noted where ossification occupied more than 30% of the cross-sectional area of the spinal canal but there are reported several cases of gross radiological change without symptoms. In this patient neurological disturbance was mild and not obviously progressive.

Occasionally ossification of the posterior longitudinal ligament is seen as a feature of diffuse idiopathic skeletal hyperostosis (Forestier’s Disease) which is characterised by bone proliferation in both axial and extra-axial sites. Ossification of the posterior longitudinal ligament is distinct from cervical spondylitis although the two conditions may co-exist. In ossification of the posterior longitudinal ligament there is ectopic bone formation rather than calcification of the posterior longitudinal ligament. The ossification exceeds the anatomical limits of the posterior longitudinal ligament and is continuous with the vertebral bodies whereas the posterior longitudinal ligament itself attaches only to the annulus. Lastly, ossification of the posterior longitudinal ligament tends to be most marked at C3 to C5, a higher level than cervical spondylitis. Reports of pathological examination of the cord are limited but the findings appear to be a non-specific combination of demyelination, axonal loss and vascular changes, all of which are maximal in the dorsal and lateral region of the cord.

Early changes are inconspicuous on lateral films of the cervical spine and are unlikely to be recognised unless the possibility of ossification of the posterior longitudinal ligament is considered. The extent of ossification is best shown by CT. In this case CT was combined with metrizamide myelography which demonstrated clearly the level and extent of cord compression.

BRF LECKY
JA BRITTON

Departments of Neurology and *Radiology,
Guy’s Hospital,
London SE1 9RT, UK

Address for reprint requests: Dr BRF Lecky, Maida Vale Hospital, London W9 1TL.

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Wernicke-Korsakoff syndrome with bilateral facial nerve palsies.

Siri: In 1881 Carl Wernicke described the first three cases of the syndrome which has subsequently borne his name. The studies of Harper indicate that the necropsy incidence of this condition is 2–8%, with only 20% of such cases being correctly diagnosed on clinical grounds. Nevertheless in the series of Vctor et al, the incidence of ocular disorders and ataxia was high (96% and 87% respectively), and even no sign of other cranial nerve palsies was present. A classical case of the Wernicke-Korsakoff syndrome was recently encountered, accompanied by severe bilateral facial weakness of lower motor nerve type. A housewife aged 29 years had had considerable alcohol intake for at least 20 years, consuming about 6 bottles of beer (approximately 200 grams of alcohol) each day. She presented to her general practitioner some ten weeks before admission with right facial hyperaesthesia, and bilateral parotid gland enlargement was noted. Two weeks prior to admission she was noted to be emotionally unstable and withdrawn, and became progressively confused and immobile. A right facial weakness was noted one week before admission. Examination revealed the patient to be obese and afebrile. She was dehydrated with bilateral parotid gland swelling and tender hepatomegaly. Neurological examination showed that she was drowsy and aphatic, and she obeyed only simple commands and mumbled occasional words. The fundo showed oedema of the nerve fibre layer with some linear haemorrhages, the discs appearing normal. Prominent horizontal and vertical nystagmus was present, with normal oculo-ocular movements. Severe bilateral facial weakness was evident involving all muscle groups; subsequent examination revealed no impairment of taste. Minimal lid movement was observed with corneal reflex testing, but there was no blink to menace. No apparent limb weakness could be detected, the reflexes in the arms being reduced and the leg reflexes were absent. The plantar responses were flexor. Sensory examination at a later date showed only a minimal distal impairment of pain in the toes.

Investigations showed a haemoglobin of 16.3 g/100 ml, a packed cell volume of 49%, and a mean corpuscular volume of 88 (range 80–96). The erythrocyte sedimentation rate was 52 mm/h. The white cell count was 20.4 10⁹ mm⁻¹ with a neutrophilia of 72%. Serum biochemistry was consistent with pre-renal failure with hypernatraemia of 154 mmol/l (range 137–145), a high urea of 58.5 mmol/l (range 3–8), and a high creatinine of 0.55 mmol/l (range 0.05–0.12). Bilirubin was normal, gamma glutamyl transpeptidase

ase was elevated at 219 units/l (range 0–30), and there was a raised alkaline phosphatase of 135 units/l (range 30–110) and an elevated lactate dehydrogenase of 332 units/l (range 110–230). A red cell transketolase estimation was not performed. A chest radiograph was normal. A computed tomographic head scan with magnified views of the brain stem was normal. Lumbar puncture revealed cerebrospinal fluid with a normal pressure, cell count, and glucose content, and an elevated protein of 1-1 G/l (range 0-1-0-65). Electromyography and nerve conduction studies showed no evidence of denervation in the small muscles of the right hand or right foot. Fibrillation were present in the right frontalis and right orbicularis oris muscles, with no motor units present under voluntary control in either muscle. The right facial nerve was inactive, motor conduction velocities and distal latencies being normal in the right median and right lateral popliteal nerves, and normal sensory conduction studies were obtained from the right median and right proximal sural nerves. The motor and sensory conduction studies were repeated in the limbs two weeks later and were unchanged.

Initial management consisted of rehydration and intravenous thiamine and a progressive improvement in the patient’s mental state occurred. Two weeks later she could converse readily, but confabulated, remained disoriented in place and time, and had a marked short term memory defect. At this stage the nystagmus had disappeared, and the retinal changes had reverted to normal. She still exhibited severe bilateral facial weakness, and a gross ataxia of stance and gait could now be demonstrated. The leg reflexes remained absent. Repeat electromyography and nerve conduction studies in relation to the face ten weeks after presentation showed motor units under voluntary control on both sides, of greater amplitude and in higher numbers on the left side. The left facial nerve was now excitable, the latencies to frontalis and orbicularis oris being 8 and 7 ms respectively (normal less than 4 ms). The right facial nerve remained excitable. Review four months after presentation demonstrated correct orientation and there was no confabulation. A short term memory defect of mild degree was still present. The left facial muscles now appeared normal, but there was a residual moderate right facial weakness involving the lower more than the upper facial muscles. She could walk unsupported, but the heel-toe gait was ataxic. The leg reflexes remained absent.

In the present case there appears little doubt of the diagnosis of Wernicke-Korsakoff syndrome in view of the long history of excessive alcohol consumption, the prominent short term memory defect with confabulation, nystagmus, ataxia of stance and gait, and associated mild peripheral neuropathy. The partial improvement with thiamine further supports the diagnosis. The cerebrospinal fluid protein may be raised. The striking feature of this case was the presence of severe bilateral facial palsies of lower motor neuron type. Such palsies have not previously been described in this syndrome to our knowledge. In the monograph of Victor, Adams and Collins' encompassing 245 patients, no case of facial palsy was encountered, nor has this ever been observed subsequently (Victor, personal communication). The development of the facial weakness in conjunction with the other classical features of the Wernicke-Korsakoff syndrome, and the gradual improvement following thiamine suggest a similar pathological process. Periventricular haemorrhages in acute cases are a typical pathological finding, and it is well known that the fibres of the facial nerve course around the sixth nerve nucleus, forming the facial colliculus in the floor of the fourth ventricle. In view of the fact that sixth nerve palsy and horizontal conjugate gaze palsy are common ocular manifestations of the syndrome, due to lesions in the floor of the fourth ventricle, speculation may occur as to why facial palsy should in fact be so uncommon.

Other causes of bilateral lower motor neuron facial palsy were considered, but felt to be unlikely. No evidence of sarcoidosis was present, and the Guillain-Barré Syndrome was felt to be excluded by the presence of nystagmus and memory disturbance, and the preservation of the arm reflexes, together with the normal nerve conduction studies in the limbs. A coincidental bilateral Bell’s palsy appeared improbable, and in this context the preservation of taste was felt to represent a central lesion, taking into account the other findings such as nystagmus.

Central pontine myelinolysis is a rare disease that typically occurs in malnourished alcoholics. The lesion usually lies centrally in the basis pontis, clinical manifestations (if present) being spastic bulbar paralysis and quadriplegia. Whilst it has occurred in association with the Wernicke-Korsakoff syndrome in necropsy cases, lesions of the tegmentum of the pons usually occur only in the presence of severe involvement of the basis pontis (Victor, personal communication), which appears unlikely in the present case. Retinal oedema and haemorrhages were the other unusual features of this case. One of Wernicke’s original cases exhibited massive optic nerve swelling with associated streaky haemorrhages, and another had redness of the discs with an isolated retinal haemorrhage. In the series of Victor et al no retinal abnormalities were seen other than small haemorrhages, this being an uncommon finding and occurring in only 3% of all cases. The retinal changes in the present case are presumably causally related to the Wernicke-Korsakoff syndrome, as they improved with thiamine, and no other cause for them was determined. They may represent similar changes in the retinal capillaries as occur in the cerebral blood vessels, with endothelial hypertrophy, microhaemorrhages and oedema.2

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JP RICE
M HOROWITZ
D CHIN

Departments of Neurology and Medicine, Royal Adelaide Hospital, North Terrace, Adelaide, South Australia 5000

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