

electroretinogram was normal. Brain stem auditory evoked potentials were poorly formed but of normal latency. Audiometry confirmed severe sensory neural deafness and a CT scan was normal.

The association of hereditary motor and sensory neuropathy with optic atrophy was first reported in 1889,³ and a number of other families in which both disorders occur have since been described.⁴⁻⁶ Likewise there is an association between hereditary motor and sensory neuropathy and deafness. Of a series of 225 patients with hereditary motor and sensory neuropathy, four were found with sensori-neural deafness.⁷ Rosenburg and Chutorian reported a family in which three members had optico-acoustic atrophy and hereditary motor and sensory neuropathy.¹ These patients had moderate slowing of the motor conduction velocities without thickened peripheral nerves. They discussed the relationship between the hereditary spino-cerebellar degenerations, optic atrophy, nerve deafness, hereditary motor and sensory neuropathy and hereditary sensory neuropathy and concluded that a considerable overlap between these conditions exists. Iwashita *et al*, described a pair of siblings with the same triad of conditions.² One of these also had evidence of sensory involvement. These cases had normal motor conduction velocities and evidence of widespread denervation. The amplitude of the sensory action potentials was not reported in either of these patients. Both groups note the superficial similarity of these conditions to Refsum's disease but consider that the differences are more fundamental, with no increase in serum phytanic acid or CSF protein, no retinitis pigmentosa, anosmia, ichthyosis or ECG abnormalities.

Our case is similar in many respects to those mentioned above. The onset of symptoms in the present case is, however, slightly later than in those previously reported. The progression of the disease in our case appears to have arrested and to have been stable for the past 30 years or so. No long term follow-up data is available on the other cases for comparison.

Although the electrophysiological data are not described in detail, there is clinical evidence of sensory involvement in the previously reported families. Our patient had no clinical or electrophysiological evidence of sensory neuropathy thus favouring a diagnosis of distal spinal muscular atrophy.

This case suggests that optic atrophy and deafness are not only an occasional feature in hereditary motor and sensory neuropathy, but also a rare finding in the distal form of spinal muscular atrophy.

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A case of tabes dorsalis with tonic pupils and lightning pains relieved by sodium valproate

Sir: We report the case of a woman who had had tabes dorsalis accompanied by lightning pains for 30 years. The pains were relieved within a few weeks by sodium valproate, but an attempt to replace this drug by carbamazepine was unsuccessful. The patient also has tonic (Holmes-Adie) pupils. The patient is a married woman born in 1914 who was first seen in August 1982, when aged 68 years, at the Centre for Pain Relief, Mersey Department of Medical & Surgical Neurology, Walton Hospital, Liverpool. She complained of "aching legs" and lightning girdle pains in the chest which she had been having for 30 years. The pains came in bouts lasting two or three weeks; she experienced four or five bad bouts and two or three less bad attacks per annum. The pains were exacerbated by cold, "tension", and

anger; and ameliorated by warmth (hot water bottle) and rest. She experienced "severe tingling" on sponging herself.

She had acquired a treponemal infection from her husband at the end of the war, during which he had served in the Merchant Navy. He was reported in 1983 as having serology consistent with having had a treponemal infection in the past; he died in October 1985 of renal and cardiac failure complicating carcinoma of the bladder. The patient herself was first seen at the Special Clinic in the Liverpool Royal Infirmary in 1958, when tabes was diagnosed. Irregular pupils, described as Argyll-Robertson, were reported at that time. She was treated with penicillin, and found to be Wassermann-negative in 1960 (blood and CSF). Further serological examinations in 1980 and 1984 confirmed only a past infection. The patient has a family history of hypertension, and is herself hypertensive, her blood pressure having been as high as 200/120 mm Hg; this is being treated with propranolol 40 mg/day. She has also been investigated for peptic ulcer, as she has a long-standing (and familial) history of heartburn. She has bilateral cataracts of long standing. Only one of her five siblings is still alive. The patient has a son and daughter, both born before the Second World War. Both are healthy and married, and each has two healthy children.

At presentation in 1982, the patient underlined 15 words on the McGill Pain Questionnaire. Conventional analysis showed that 90% of word groups in the somatic category were chosen, 100% in the evaluative category, 100% in the miscellaneous category, but only 25% in the affective category. The (rank) intensity score was 45, or 75%. Overall, the picture was one of severe organic pain with remarkably little functional overlay, in sharp contrast to the score patterns obtained in cancer² or backache.³ When asked which pain descriptors she would most like to be rid of, she unhesitatingly replied "stabbing and scalding".

On examination (which was during a bout, but not during an attack, of pain) she was 150 cm tall and weighed 55.2 kg. The painful area was symmetrically distributed in the T5 segments; the right upper thorax and both legs exhibited allodynia. Tone and power were normal for her age, but tendon reflexes were universally absent, and no superficial reflexes could be elicited. She did not exhibit the slowed withdrawal times from noxious stimulation which are usually associated with tabes dorsalis. Sensation of passive movement was normal and symmetrical at all joints tested. Pinprick sensa-

tion was diminished over both breasts, both hands, and both feet; application of a pin to the feet (particularly the left) provoked dysaesthesia. Quantitative somatosensory threshold testing revealed normal thresholds for touch (von Frey filaments) and for two-point discrimination. The vibrometer⁴ yielded normal threshold values for both hands; on the right leg it was at the upper limit of normal, and significantly above normal on the left leg. Somewhat surprisingly, normal values were found for skinfold pinch (= tissue-damage pain). Thresholds for hot pain, cold pain, and cold were normal, as tested with the Marstock⁵ apparatus, but thresholds for warmth were greatly raised in both feet (43.7°C on the right and 42.4°C on the left), so as to approach the hot pain thresholds (45°C and 45.7°C on the right and left respectively); the latter are in fact at the upper limits of normality.

Motor and sensory conduction velocities in the left median, ulnar, lateral and medial popliteal nerves were within normal limits. F wave latencies were normal for all four muscles tested; no consistent H response could be obtained. Somatosensory evoked potentials were elicited on the right side from the median and lateral popliteal nerves; they were recorded over SI cortex, the C5 spinous process, and Erb's point (median nerve only). Latencies were within normal limits. Since only large myelinated fibres are involved in all these responses, the small axons subserving the abnormal warmth sensation were not tested electrically.

With best spectacle correction, visual acuity was 6/60 on the right and 6/36+1 on the left. Lens opacities and senile macular degeneration were present in both eyes. The pupils were large (right 5.7 mm, left 5.5 mm) with a mild degree of anisocoria in normal room lighting. They did not constrict directly or consensually when a bright light was shone in one eye; no response to accommodation could be elicited. Instillation of two drops of 2.5% methacholine chloride into the conjunctival fornix of each eye reduced the pupillary diameter to 4.3 and 5.0 mm on the right and left respectively, using flash pupillometry under standard lighting conditions after 30 minutes. Pupilloconstriction was also obtained by instillation of 1/8% pilocarpine. Biomicroscopy with a slit lamp revealed vermiform movements in the sphincter muscles of both eyes; these are said to be pathognomonic of tonic pupils. These pupil abnormalities, coupled with the loss of deep tendon reflexes, would suggest the diagnosis of tonic pupils (Adie's syndrome). However, complete absence of all tendon reflexes is

reportedly rare, though it occurred in one eighth (3/24) of bilateral cases, which themselves only accounted for 24 out of 122 cases (of whom 22 were female).⁶ Argyll-Robertson pupils are small (less than 3 mm) and react to accommodation but not to light; while tonic pupils are large, do not react to light, relax slowly after accommodation, and are constricted by methacholine and physostigmine.⁷ One sixth of the cases in Thompson's⁶ series of 150 cases of Holmes-Adie syndrome had positive treponemal serology, including seven out of 21 cases of "neuropathic tonic pupils" with bilateral eye signs.

While small myelinated (A delta) fibres are notoriously difficult to test by objective examination in man, we were struck by the patient's report of pain exacerbation by cold, and her obvious dislike of cold stimuli. In our experience (Bowsher and Lahuerta, unpublished observations), cold allodynia is characteristic of peripheral nerve and dorsal root entry lesions. The production of dysaesthesia by pinprick to the feet, as well as the diminished pinprick sensation in the painful segments, also suggests the possibility of an A delta abnormality. The patient could induce paraesthesiae by sponging herself, and felt pain when her right thorax or legs were rubbed. This allodynia, together with the fact that her pain was exacerbated by autonomic events (anger and "tension"), point to a central neurogenic lesion, though peripheral pathology cannot be ruled out (cf. causalgia).

For the 30 years prior to her first attendance at the centre for Pain Relief, during which this patient had suffered lightning girdle pains, many analgesics had been tried. They all failed to relieve her pain, and many "upset her stomach". She did, however, derive some comfort from a combination of dextropropoxyphene hydrochloride 32.5 mg and paracetamol 325 mg; she had been taking some six tablets a day to "take the edge off the pain". Following her first interview and examination in 1982, she was started on sodium valproate 200 mg and amitriptyline 25 mg at night, increasing to 600 mg and 75 mg respectively in divided doses over two weeks. When seen one month later (September 1985), she stated that her pain had disappeared in ten days, but agreed that it might have been the end of a bout. She complained of heartburn and weight gain. The drugs were reduced to 400 mg and 50 mg per day. Another month later she was still complaining of heartburn, and now had mouth ulcers; she had stopped taking valproate a few days before attending the clinic. She was advised to stop tricyclics

and persevere with the anticonvulsant. The heartburn and mouth ulcers gradually disappeared, and tricyclics were cautiously reintroduced. Nine months after her first visit she had experienced no further pain, but was greatly concerned about her weight, which had increased from 55.2 kg to 60.1 kg over the period. Weight gain during valproate treatment is well known, and has recently been reviewed by Dinesen *et al.*,⁸ who reported a gain of more than 4 kg in 57% of patients, irrespective of dose.

The use of anticonvulsants, and of valproate in particular, for lancinating pains other than trigeminal neuralgia, and including tabes dorsalis, has subsequently been reviewed by Swerdlow.⁹ Rapid relief with carbamazepine had been reported in several cases,¹⁰ but an attempt to replace valproate with it in the present case was soon followed by a severe attack of pain, such as she had experienced before anticonvulsant therapy was instituted. The patient returned to valproate on her own initiative, and the pains soon subsided.

One year after her first visit, when taking valproate 400 mg and amitriptyline 50 mg per day, she reported a slight recurrence of pain in her right scapula, but volunteered that she had been worrying about her family holiday arrangements. Another exacerbation occurred three months later, including allodynia and hyperpathia in the right arm, when she was concerned about the health of the brother (who has since died) to whom she was closest. In the 18 months since that time, the patient has had no serious recurrence of pain, despite the death of her favourite brother in 1984, and of her husband and oldest sister in 1985, within a few days of each other. Amitriptyline was tailed off and stopped late in 1984. Liver function tests were at first performed three monthly, but are now carried out twice a year. The patient continues on a twice-daily dose of sodium valproate 200 mg; she no longer has any gastroenterological side effects; and is now a cheerful and virtually pain-free woman.

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Clostridium perfringens brain infection following a penetration wound of the orbit.

Sir: We present the first reported case whereby the response of clostridial cerebritis to antibiotics and hyperbaric oxygen was monitored using computed tomography. The mechanism of infection illustrates the danger of concealed penetrating injury and its possible consequences.

A 47 year old company director presented at his local hospital having tripped and fallen into a rose bush. A branch had pierced the inner aspect of his right eye. He had lacerations of the bulbar conjunctiva and the lower lid. The globe was intact. Tetanus toxoid was administered and he was admitted for repair of his injuries. An epistaxis that night and haematemesis the next day caused

postponement of surgery. On the third day following injury he developed marked lid oedema and chemosis, with a pus-like discharge. The wound was explored and multiple fragments of wood were found together with pus. After irrigation and repair of the lid laceration, treatment with metronidazole, erythromycin and chloramphenicol was commenced. After operation the eye was stable, but he developed pyrexia. On the sixth day following injury, he became aphasic with a right hemiparesis. Culture of pus from the wound showed a pure growth of *Clostridium perfringens*. He was then referred to Atkinson Morley's Hospital.

On admission, he was conscious, but aphasic and obeying only simple commands. His right eye had lid swelling and chemosis. There was a right hemiparesis: Grade IV of the arm, Grade 0 of the leg. Radiography revealed bony injury to the supero-medial aspect of the right orbit and the upper and lower walls of the right frontal sinus, which was opaque. There was a small gas loculus in the right frontal pole. Computed tomography showed oedematous changes in the medial aspect of both frontal lobes, more extensive on the right, where a gas bubble was present in the frontal pole (fig). Treatment was started with intravenous penicillin, metronidazole and chloramphenicol. Later that day hyperbaric oxygen therapy was commenced; three treatments over three days at 250 KPa (2½ atm.) pressure for three hours was given.

Thereafter steady, uneventful improvement took place. Following review by an ENT surgeon the right fronto-ethmoidal sinus was explored. A fracture of the orbital roof was found, the frontal sinus containing

pus and herniating cerebral tissue. A fracture of the superomedial aspect of the orbit was displaced into the ethmoid from which were removed two pieces of foreign material, one measuring 1.2 cm × 4 mm. The pus was sterile and biopsy of the brain hernia demonstrated only necrosis. Eighteen days following his injury the dysphasia and hemiparesis had resolved considerably. He was discharged for convalescence on antibiotics. On review a month later, speech was normal, although there was a flatness of affect. The right eye was normal and there was no limb weakness. A CT scan showed resolution of the intracranial gas, but with some residual frontal lobe oedema. Full recovery subsequently occurred.

Although potentially fatal, intracranial infection with *Clostridium perfringens* can be successfully treated following prompt aspiration, where indicated, and antibiotic therapy.¹ The mortality rate from cerebral abscess due to clostridial infection has been assessed at 24%.²

In this case, only computed tomography made possible the diagnosis of "clostridial cerebritis" (with potential for abscess formation). Intensive therapy was clearly indicated and the patient's condition improved markedly during the use of hyperbaric oxygen in conjunction with antibiotics, although Keogh³ has suggested that there is no place for the former. We would emphasise the importance of investigating the possibility of retained foreign material within penetrating wounds around the orbit, which may act as a source of infection.

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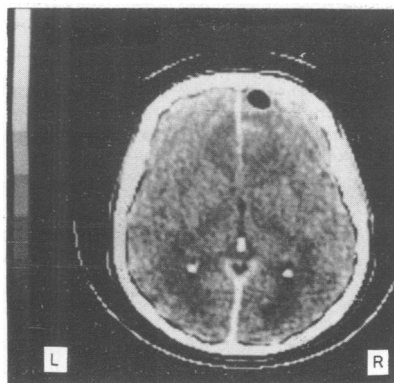


Fig Computed tomography of the brain showing a gas bubble in the right frontal pole and oedematous changes in the medial aspect of both frontal lobes.