sclerosis, however, familial association is not an epidemiological feature of AIDP. In fact, familial cases have rarely been reported.²⁻⁴ We recently encountered a father and his daughter who developed an unusual but clinically similar course of AIDP after triggering factors associated with occurrence of AIDP.

The father, a 50 year old, previously healthy Kurdic Jew, complained of right hand weakness a few days after haemorrhoidectomy under general anaesthesia. Within a few more days, lower limb weakness and paresthesias ensued and slowly progressed. He did not consult a physician at that time as his condition had stabilised. A year later, he was first referred for consultation. On examination there was global areflexia, minimal distal weakness, atrophy of the right hand, and pronounced distal weakness in both legs, greatest on the left. Vibration and superficial sensation were impaired in a glove and stocking distribution in both legs and in the right hand. Cerebrospinal fluid (CSF) was acellular, with a protein concentration of 0.94 g/l and a normal glucose concentration. Nerve conduction velocities were 38, 37.5, and 39 m/s in the median, ulnar, and lateral popliteal nerves respectively; F wave latencies were prolonged in all extremities. Sensory potentials were reduced or could not be elicited and sensory conduction velocities were slowed (left sural nerve 22 m/s). Sural nerve biopsy showed massive loss of myelinated fibres with remyelination and some onion bulb formation, without an inflammatory reaction. During the next few years, his clinical condition improved slightly. Motor conduction velocities four years later were 53 and 46 m/s in the median and ulnar nerves respectively.

Four years later, at the age of 24, the first patient's daughter had serologically established Epstein-Barr virus infection. Two weeks later, she progressively developed lower limb weakness. On examination she had bilateral foot dorsiflexion weakness, more pronounced on the right with diminished deep tendon reflexes in the legs. Pinprick sensation was decreased in the right leg and the left hand in a glove and stocking distribution. During the next three days, her weakness progressed to involve also the plantar flexion of the feet and the iliopsoas on the right. Global areflexia appeared but sphincter functions were preserved. The CSF was acellular with normal protein (0.3 g/l) and glucose concentrations and no oligoclonal bands. Motor nerve conduction velocities were reduced (median nerve 45 m/s; tibial nerve 32 m/s; common peroneal nerve 28 m/s) and distal latencies were prolonged (right tibial nerve 17.5 ms; left tibial nerve 18.4 ms; right common peroneal nerve 20.8 ms; left common peroneal nerve 17.1 ms), as were F wave latencies in the legs (>60 ms). Right sural nerve conduction was 34 m/s. Electromyography was normal. Treatment with prednisone was initiated. Her condition gradually improved, but residual left dorsiflexion weakness was still evident after a year. Motor conduction velocities six months later were improved (38 and 37 m/s in the common peroneal and tibial nerve respectively).

Erythrocyte sedimentation rate, haemogram, serum electrolytes, liver and kidney function tests, latex, Rose Waler, antinuclear factor, VDRL, AST, creatine phosphate protein electrophoresis, HIV and hepatitis B surface antigen serology, and

chest radiographs were within normal limits or negative in both patients.

GM1 gangliosides, galactocerebroside (GalC), and myelin basic protein (MBP) autoantibodies were determined as previously described.⁵ The daughter had increased GM1 and MBP antibodies whereas levels in the father were normal. GalC antibodies were raised in both.

The disorder in these two patients fulfils the criteria required for the diagnosis of AIDP.¹ In both it occurred after an antecedent event associated with occurrence of AIDP, was diffuse, affecting all four limbs with widespread slowing of nerve conduction, and was followed by gradual improvement on follow up studies. Their disease course differed, however, from typical AIDP by several features: the disorder was clearly asymmetrical, did not involve respiratory and cranial nerves, and was associated with residual deficit for several years of follow up.

The fact that familial cases have not been described more often in AIDP, a relatively common condition of putative immune mediated pathogenesis, is intriguing. The clinical history and the laboratory work-up in these patients did not support a possible common basic autoimmune predisposition. There was no evidence for an immunological abnormality on auxiliary and serological studies or for an associated autoimmune condition. GM1 antibodies, present in about a quarter of patients with AIDP were significantly raised only in the affected daughter's serum; her serum also contained detectable MBP antibodies. Human leucocyte antigen (HLA) analysis of the affected father and daughter did not show any of the previously reported HLA alleles associated with systemic or neurological autoimmune conditions. The presence of serum GalC antibodies in patients with AIDP might be an indicator of peripheral nerve injury, but whereas they may induce experimental peripheral nerve demyelination, there is no evidence to link them with AIDP pathogenesis.

Thus the unusual course, the lack of serological abnormalities, and the absence of immunological features associated with AIDP suggest that the familial predisposition in these two patients may be coincidental or belong to a different pathogenetic mechanism. The paucity of reported familial cases and a lack of any immunological disturbances in our patients seems to indicate that AIDP is different from other general and neurological autoimmune disorders.

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- 1 Arnason BGW. Acute inflammatory demyelinating polyradiculoneuropathy. In: Dyck PJ, Thomas PK, Lambert EH, Bunge R, eds. *Peripheral neuropathy*. 2nd ed. Philadelphia: WB Saunders, 1984:2050-100.
- 2 Margairaz A, Poyart C. Forme familiale de polyradiculonevrite aigue spontanement resolutive. *Rev Neurol* 1965;113:475-9.

- 3 Saunders M, Rake M. Familial Guillain-Barré syndrome. *Lancet* 1965;ii:1106-7.
- 4 McGregor GA. Familial Guillain-Barré syndrome. *Lancet* 1965;ii:1296.
- 5 Wirgin I, Brenner T, Argov Z, Steiner I. Multifocal motor nerve conduction abnormalities in amyotrophic lateral sclerosis. *J Neurol Sci* 1992;112:199-203.

Peripheral neuropathy in association with insulinoma: clinical features and neuropathology of a new case

Peripheral neuropathy associated with hypoglycaemia secondary to insulinoma is unusual and just 30 patients have been described.¹

Three years before admission this 60 year old woman started episodes of confusion, self abuse, and irrational behaviour. Six months before entry, she noted progressive loss of sensation and weakness below the knees and hands. Neurological examination disclosed a mild paresis with mild impairment of all sensations in a glove and stocking distribution. The deep tendon reflexes were decreased. Laboratory investigations showed a fasting plasma glucose concentration below 50 mg/dl and a serum insulin concentration of 18.1 µU/ml. A fasting test showed hypoglycaemia associated with inappropriately raised insulin levels. Coeliac angiography, abdominal echography, and abdominal CT showed a vascular tumour in the head of the pancreas. The motor nerve velocities were slightly decreased and there was an important reduction in muscle action potential amplitude. Sensory nerve action potentials were absent with the exception of the right median nerve; F wave velocities were decreased except for the left tibial nerve. Electromyography showed evidence of denervation in the right first dorsal interosseous, tibialis anterior, and both extensor digitorum brevis muscles. No denervation was found in the left vastus medialis. These abnormalities were compatible with an axonal sensorimotor neuropathy. Sural nerve biopsy showed axonal degeneration. Other causes of acquired polyneuropathy such as carcinomatous polyneuropathy, paraproteinaemias, uraemia, alcohol, diabetes mellitus, connective tissue diseases, amyloidosis, and hypothyroidism were excluded but a 4 cm insulinoma was identified and surgically removed. Hypoglycaemic attacks have not recurred and there has been motor improvement but paresthesiae have persisted during the next three years. Electrophysiological studies demonstrated no significant change.

Peripheral neurological symptoms in the course of hypoglycaemic attacks are unusual and are probably due to maintained hypoglycaemia rather than secondary to hyperinsulinaemia. After excision of a pancreatic tumour, sensory symptoms tend to regress but definite improvement in muscle power is uncommon. After three years of follow up, no objective sensory impairment was evident in our patient. Motor improvement was also found. Electrophysiological studies were carried out in only 10 of the 30 cases previously reported.¹ In one case, there was an analysis of nerve histology and there are two necropsy studies.¹⁻³ The neuropathology of this syndrome, however, remains controversial. Muscle histology has been described in three cases, showing different degrees of fibre atrophy indicative of denervation.^{1,4,5} Pathological studies disclosed degeneration of pyramidal tracts, myelin

sheaths, and anterior horn cells but no abnormalities in dorsal and ventral roots or in dorsal root ganglia.³ A sural nerve biopsy study indicated axonal neuropathy.¹ A necropsy study showed destruction of anterior horn cells but no abnormalities of dorsal root ganglia and peripheral nerve.⁴ Therefore the damage in this neuropathy could be located in the axon or in the nerve cell body. The studies carried out on our patient are characteristic of a pure axonal degeneration, but a coexisting lesion in the anterior horn cell cannot be ruled out.

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- 1 Jaspan JB, Wollman RL, Bernstein L, Rubenstein AH. Hypoglycemic peripheral neuropathy in association with insulinoma: implication of glucopenia rather than hyperinsulinism. Case report and literature review. *Medicine (Baltimore)* 1982;61:33-44.
- 2 Moersch FP, Kernohan JW. Hypoglycemia: neurologic and neuropathologic studies. *Arch Neurol Psychiatry* 1938;29:242-57.
- 3 Tom MI, Richardson JC. Hypoglycemia from islet cell tumor of pancreas with amyotrophy and cerebrospinal nerve cell changes: a case report. *J Neuropathol Exp Neurol* 1951;10:57-66.
- 4 Bruyn GW, Garland H. Neuropathies of endocrine origin. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*. Vol VIII. Amsterdam: North Holland Publishing Co, 1970:52-6.
- 5 Mulder DW, Bastron JA, Lambert EH. Hyperinsulin neuropathy. *Neurology* 1956;6:627-35.

Trigeminal neuralgia due to contralateral meningioma of the posterior cranial fossa

Posterior fossa meningiomas are uncommon intracranial tumours, accounting for about 10% of all intracranial meningiomas.¹ They are now easily diagnosed by CT, but sometimes they grow very large before becoming clinically evident.² Impairment of ipsilateral cranial nerves (most often the 5th, 7th, 8th, and 9th nerves), cerebellar ataxia, and signs of increased intracranial pressure are the symptoms presenting most often.³ Infrequently the tumour may cause contralateral cranial nerve involvement, which confuses the diagnosis.^{4,5}

We report a case of meningioma of the posterior fossa, presenting with contralateral typical trigeminal neuralgia as the main symptom. A 58 year old married woman complained of a right typical trigeminal neuralgia of about one year's duration, involving all three divisions of the nerve and described as "electric-like bursts", triggered by eating. She had received 600 mg carbamazepine daily without effect.

Neurological examination showed only a minimal unsteadiness of gait and a fine horizontal nystagmus. The rest of the neurological examination was normal; in particular the corneal reflexes were present and there was no hypesthesia or hypalgesia over

CT scan of the head showing a contralateral (left sided) high density lesion of the posterior fossa.

the face and forehead. EEG and otological findings such as audiometry, caloric responses, electronystagmography, and brainstem auditory evoked response were normal.

CT of the head showed a contralateral (left sided) high density lesion of the posterior fossa enhancing uniformly with intravenous contrast medium. The tumour, measuring 4 × 4 × 3 cm, distorted the brainstem and caused an initial triventricular hydrocephalus (figure). At surgery, the mass, removed in its entirety, was located in the posterior portion of the tentorium, close to the left transverse sinus and the confluence of the sinuses. Histological examination showed a meningioma of transitional type. The patient's early postoperative course was favourable. The facial pain disappeared completely and a neurological examination after four months only showed a slight unsteadiness of gait. She did not complain of facial pain but six years later she was operated on for a recurrence of the tumour in the same region. The patient is still free of symptoms.

Contralateral involvement of the trigeminal nerve due to a mass occupying the posterior fossa space (such as meningioma), has been reported.^{1,5} In these cases, however, the involvement of the 5th nerve was generally manifest by early impairment of facial sensation and decreased corneal reflex. Very rarely, a contralateral trigeminal neuralgia is the only symptom for a long time. Haddad and Taha⁴ summarised 21 such cases from the medical literature. Six of these patients had a typical neuralgia, 12 atypical, and in four cases the information available was not complete. There was a predominance of females and meningioma was the most frequent tumour.

In our case, the most plausible mechanism to explain the pathogenesis of the contralateral trigeminal involvement is that the tumour, situated in the back portion of the posterior fossa, pushed the brainstem and caused compression of the nerve root at its point of entry into the tentorial foramen.⁴

In conclusion, an isolated trigeminal neuralgia, especially if carbamazepine resistant, may be due to a contralateral tumour of the posterior fossa, such as a meningioma, and

requires CT of the head. Surgical treatment relieves pain in almost all cases.

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- 1 Cushing H, Eisenhardt L. *Meningiomas: their classification, regional behaviour, life history and surgical end results*. Springfield, IL: Charles C Thomas, 1938:169-223.
- 2 Hirsh LF, Mancall EL. Giant meningiomas of the posterior fossa. *JAMA* 1978;240:1626-7.
- 3 Martinez R, Vaquero J, Areitio E, Bravo G. Meningiomas of the posterior fossa. *Surg Neurol* 1983;19:237-243.
- 4 Haddad FS, Taha JM. An unusual cause for trigeminal neuralgia: contralateral meningioma of the posterior fossa. *Neurosurgery* 1990;26:1033-8.
- 5 O'Connell JEA. Trigeminal false localizing signs and their causation. *Brain* 1978;101:119-42.

Panhypopituitarism after cavernous sinus thrombosis

Anterior hypopituitarism is a rare late complication of cavernous sinus thrombosis.¹⁻⁵ Posterior hypopituitarism has not as yet been described. We report a 27 year old woman who presented eight months after septic cavernous sinus thrombosis with an Addisonian crisis and diabetes insipidus in association with a urinary tract infection.

The previously well woman gave a six month history of headache, increasing in the week before admission, and was found to have meningism. Her menses had been normal up to the time of admission. Measurements in CSF obtained by lumbar puncture gave a protein concentration of 43 mg/l, a normal glucose concentration (5.2 mM with a blood glucose of 7.5 mM), and a white cell count of less than 5 cells/mm.³ No organisms were found. The next day she became drowsy (but obeyed commands) and pyrexial (38°C). She had a pulse of 80 beats/min and a blood pressure of 130/80 mmHg and was transferred for further assessment. There was bilateral periorbital swelling with a right proptosis. She had no perception of light on the right but normal acuity (4/5) and pupillary responses on the left. The right fundus showed retinal venous congestion, with a fixed and dilated pupil. There was a complete right external ophthalmoplegia, with limitation on the left of elevation and depression with absent abduction, but normal adduction. The remainder of the examination was normal. A cranial CT showed opacification of the sphenoid sinus and part of the ethmoid sinuses, filling defects in the cavernous sinuses after intravenous contrast, with congestion of the orbital veins and proptosis. The pituitary appeared normal. There was a small low density area in the right frontal lobe consistent with ischaemia. A diagnosis of septic cavernous sinus thrombosis secondary to sphenoid sinusitis was made. Her electrolytes were normal. She was started on penicillin, chloramphenicol, and metronidazole. Exploration of the right sphenoid sinus yielded pus and a pneumococcus was grown from blood cultures. She improved