Hepatitis B tests, six serum and tibial tiated. Her nerve still normal. Treatment protein oneal veloices. The appeared aminated and lateral velocities 18-4 m/s; tibial a wave progressed. After and slowly did his condition affer, his condition did not improve on follow up studies. Their disease course differed, however, from typi- cal 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 etiologic deficit. There was no evidence for an immunologi- cal abnormality on auxiliary and serological studies or for an associated autoimmune condition.

The presence of serum GaC 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 pathogen- esis.

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 families may be coinciden- tal or belong to a different pathogenic mechanism. The paucity of reported familial cases and a lack of any immunological disturbances in our patients seems to indi- cate that AIDP is different from other general and neurological autoimmune dis- orders.

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-ahne, bilateral facial, upper and lower limbs weakness for a few months before entry, she noted progressive loss of sensation and weakness below the knees and hands. Neurological examination disclosed a mild paresis with mild impair- ment of all sensations in a glove and stock- ing distribution. The deep tendon reflexes were decreased. Laboratory investigations showed a fasting plasma glucose concentra- tion below 50 mg/dl and a fasting insulin concentration of 18.1 μU/ml. A fasting test showed hypoglycaemia associated with inappropriately raised insulin levels. Coelic angiography, abdominal echography, and abdominal CT showed a normal 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 lateral nerve. Electromyography showed ev- idence of denervation in the right first dorsal interosseous, tibialis anterior, and both extensor digitorum brevis muscles. No der- envation was found in the left vastus medi- alis. These abnormalities were compatible with an axonal sensorimotor neuropathy. Sural nerve biopsy showed axonal degenera- tion. Other causes of acquired polyneuropa- thy such as carcinomatous polyneuropathy, paraproteinemia, uraemia, alcohol, dia- betes mellitus, connective tissue disease, amyloidosis, and hypothyroidism were excluded but a 4 cm insulinoma was identi- fied and surgically removed. Hypoglycaemic attacks have not recurred and there has been motor improvement but pareses 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 hypo- glycaemia rather than secondary to hyper- insulinemia. After excision of a pancreatic tumour, sensory symptoms disappeared 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. In one case, there was an analysis of nerve histology and there were 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 size and fibre type distri- bution.1,3 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. A sural nerve biopsy study indicated axonal neuropathy. A necropsy study showed destruction of anterior horn cells but no abnormalities of dorsal root ganglia and peripheral nerve. 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 were characteristic of a pure axonal degeneration, but a coexisting lesion in the anterior horn cell cannot be ruled out.

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Trigeminal neuroma due to contralateral meningioma of the posterior cranial fossa

Posterior fossa meningiomas are uncommon intracranial tumours, accounting for about 10% of all intracranial meningiomas. They are now easily diagnosed by CT, but sometimes they grow very large and become symptomatic. Impression 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. Infrequently the tumour may cause contralateral cranial nerve involvement, which confuses the diagnosis.

We report a case of meningioma of the posterior clinoid with contralateral typical trigeminal neuroma as the main symptom. A 58 year old married woman complained of a right typical trigeminal neuroma 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 hypalgesia or hypesthesia of the face and forehead. EEG and otopathological findings such as audiometry, caloric responses, electroneystagmography, and brainstem auditory evoked response were normal.

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

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The head of the showed a contralateral (left sided) high density lesion of the posterior fossa enhancing uniformly with intravenous contrast medium. The tumour, measuring 4 x 4 x 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. 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 neuroma is the only symptom for a long time. Haddad and Tahb summarised 21 such cases from the medical literature. Six of these patients had a typical trigeminal 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.

In conclusion, an isolated trigeminal neuroma, 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.

Panhypopituitarism after cavernous sinus thrombosis

Anterior hypopituitarism is a rare late complication of cavernous sinus thrombosis.1-3 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 Addisonsian 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/mm³. 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 no oedema, 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 dilated disc and a white spot. 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, without obstruction 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 sphenoidal sinus yielded pus and a pneumococcus was grown from blood cultures. She improved...