

Figure CT scan showing right temporal lobe and left cerebellar hemisphere haemorrhages.

### Meningitis and spinal subdural empyema as a complication of sinusitis

Spinal subdural empyema is a rare event. We describe the case of a young man who developed sinusitis while recovering from a routine operation followed by a dramatic meningitic illness with formation of a spinal subdural empyema.

A previously fit 23 year old man had an elective left temporal mandibular joint meningo-plasty for Costen's syndrome. Two days later he developed a mild headache with a slightly purulent nasal discharge. Within the following 24 hours his headache became bifrontal and more severe. He developed visual hallucinations though he remained orientated and there was marked neck stiffness, severe low back pain with very limited straight leg raising and a high fever. There was no previous history of sinus disease or immunosuppression. His lumbar puncture showed turbid green cerebrospinal fluid (CSF) with 284 polymorphs and 134 lymphocytes with a CSF protein of 1.5 grams per litre and a low CSF glucose.

Plain skull radiographs showed thickening of the right maxillary sinus with a small gas bubble in the supra-sella cistern. A CT scan confirmed these findings and showed a completely opaque sphenoid sinus. Cultures from his CSF grew *streptococcus milleri* (a micro-aerophilic haemolytic streptococcus) and a bacteroides of the corrodens type. Both these organisms are compatible with an origin in the upper respiratory tract. Ampicillin, metronidazole and gentamicin were given intravenously. His sinuses were drained and a considerable amount of green pus was obtained. His temperature gradually settled though his severe back pain persisted and his tendon jerks in both lower limbs became depressed. A cervical myelogram was performed and this showed somewhat irregular subarachnoid space in the lower cervical region and a complete block at the level of D7. A subsequent CT scan of the same region confirmed a posteriorly situated mass causing forward displacement of the cord but no bony erosion. He had a laminectomy from D7 to D10. A subdural mass with much granulation tissue was identified and suitable decompressive measures were undertaken. Following this he made a slow but progressive recovery although he was troubled with low back pain and was discharged home after completion of a month of antibiotic therapy.

The association between sinusitis and intracranial empyema is well established. Spinal subdural empyema is a much rarer occurrence and has not been previously described as a complication of sinusitis.

In a 1973 review of the literature only ten cases were described.<sup>1</sup> Five of these patients died, three were left with significant neurological deficits and only two fully recovered. Prompt surgical drainage and antibiotics have obviously improved prognosis, and more recently eight additional cases have been reported.<sup>2</sup> Seven of these patients made full or fair recoveries.

Typically, spinal subdural empyema presents with a fever, backache, and radicular symptoms and signs. There have been claims that the absence of percussion tenderness helps to distinguish it from an epidural collection of pus<sup>1,3,4</sup> which is usually associated with vertebral osteomyelitis or bacteremia. The diagnosis has recently been facilitated with the introduction of spinal CT.<sup>5,6</sup>

In our patient no clear collection of intracranial subdural pus was identified with a CT brain scan with contrast though this may appear normal in the early stages.<sup>7</sup>

The striking feature in this case is the finding of an extensive spinal subdural empyema. It remains unclear whether it was formed by haematogenous spread from an intracranial or paranasal source despite the relative avascularity of the subdural space. The empyema may have derived from extension through the arachnoid in the presence of meningitis. Seeding of the subdural space by lumbar puncture has been described as a possible cause.<sup>8</sup>

Paranasal sinus pathology should be excluded as a cause of pyogenic meningitis even in the absence of a clear history of sinusitis, and should now be considered as a possible cause of spinal subdural empyema.

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### Familial cerebellar ataxia and possible cosegregation with an inversion in chromosome 4

We have recently encountered a family which may shed light on the gene locus of an inherited form of late onset cerebellar ataxia.

The family pedigree (fig) reveals that three of seven siblings have or have had late onset cerebellar ataxia. Case II2 died aged 65 years. From the age of 55 years relatives noticed that she had a progressively unsteady gait, and slurred speech. None of her medical records can be traced. Case II4 died aged 74 years. She was first referred to a neurologist at the age of 60 years with a one year history of unsteady gait and dysarthria, which showed slow but steady deterioration culminating in marked disability. Relevant findings on examination at referral included nystagmus on lateral gaze, normal optic discs, and normal tone, power, tendon reflexes and sensation in all limbs. Air encephalography did not show any obvious cerebellar abnormality nor was any noted at necropsy examination.

The necropsy report recorded that this woman had a chromosome abnormality although the precise nature of this was not specified. Review of her medical records and personal contact with all known locally sited

Her limbs were flaccid with symmetrically diminished reflexes and equivocal plantar responses.

She made no response to deep painful stimuli. Initial blood results revealed a leucocytosis of  $48.9 \times 10^9/l$  with a differential of eosinophils 32.81 and neutrophils 12.18  $\times 10^9/l$  respectively. Her haemoglobin, biochemical screen and liver function tests were all normal. A full coagulation screen including platelets, KPPT, fibrinogen and Protein C were also normal.

A chest radiograph showed cardiomegaly and old calcified apical foci. Her CT scan (figure) showed a right temporo-parietal haematoma with spread into the right lateral ventricle and associated mass effect, and a left cerebellar haematoma with compression of the fourth ventricle but without resultant hydrocephalus. She failed to regain consciousness and died 10 weeks later. Permission for a necropsy examination was refused.

HES is a term which was introduced by Hardy and Anderson in 1968.<sup>1</sup> It is defined as persistent eosinophilia ( $> 1500 \text{ mm}^3$ ) in a patient in whom no known underlying cause can be found and who develops end organ injury. Central nervous system disturbance is the second most important clinical manifestation.

In a recent study<sup>2</sup> 65% of patients were affected. Central nervous system dysfunction was seen in seven patients (15%), four of whom had a distinctive encephalopathy. Peripheral neuropathy was found in 27 patients (52%). Embolic phenomena including anterior and middle cerebral artery occlusions were seen in six (12%).

This is the first reported case of HES presenting with intracranial haemorrhages.

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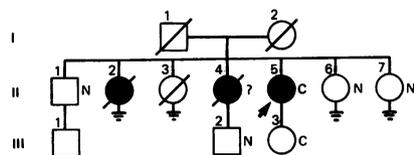


Figure The family pedigree. N = pericentric inversion of chromosome 4 not present. C = carrier of pericentric inversion. ? = ? inversion 4 carrier.

cytogenetic laboratories has not as yet resulted in any further information. Detailed neuropathological examination was not performed.

Case II5 had bilateral cataracts removed aged 55 years. Shortly after she was first observed to have an unsteady gait and slurred speech. On examination aged 66 years optic atrophy and horizontal nystagmus were documented and CT scan was reported as showing marked cerebellar atrophy. Now aged 74 years she cannot walk without support but otherwise manages well with the routine tasks of daily life. Chromosome studies undertaken on cultured lymphocytes showed a pericentric inversion of one number 4 chromosome with break points at p14 and q21 that is, 46,XX,inv(4)(p14q21).

The father (I1) and mother (I2) of these individuals were unrelated and allegedly without signs of cerebellar dysfunction until their deaths aged 63 and 87 years respectively. No signs of cerebellar ataxia have been noted in the surviving sibs who are aged 83 years (II1), 67 years (II6) and 64 years (II7), or in individuals III2 and III3 who are both aged 51 years; only one (III3) of these individuals carries the inversion 4 chromosome.

Precise classification of hereditary ataxia is notoriously difficult and this family is no exception in that the disease in Case II4 might reasonably be labelled as pure cerebellar ataxia whereas the description of optic atrophy in Case II5 suggests a more widespread disease process. Variation of this nature within families is well documented.<sup>1</sup>

Whatever its precise classification, familial late onset cerebellar ataxia usually, if not invariably, shows autosomal dominant inheritance,<sup>2</sup> whereas the apparent involvement in only one generation of this family is more consistent with autosomal recessive inheritance. However, the father of these affected women died at the relatively young age of 63 years and might therefore have developed the disorder had he survived longer so that autosomal dominant inheritance cannot be excluded.

Thus it may be that the pericentric inversion detected in one and possibly two affected individuals is a marker for the disorder with one of the breakpoints indicative of its genetic locus. Inevitably we can only speculate at present since the information as it stands is far from conclusive. Thus we are simply presenting our findings as an observation which may be of interest at a time when attention has focused on molecular biology as a means of mapping the disease loci of inherited neurological disorders.<sup>3</sup>

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## MATTERS ARISING

### Magnetic resonance imaging in Behçet's disease

We have read with interest the recent study by Besana *et al*<sup>1</sup> analysing the neurological involvement of eight patients with Behçet's disease (BD) by means of electrophysiological and MRI evaluation. They found two patients with MRI abnormalities. In one case there was a small lesion in the periventricular white matter and in the second there were two small lesions in the left periventricular white matter. They pointed out that characteristics of the observed lesions on MRI could be typical of ischaemic pattern, possibly due to vasculitis. Nevertheless, the authors did not exclude a demyelinating injury. We think that in some cases this last mechanism is the most likely one.

Recently, we have seen a 28 year old male with a history of recurrent oral and genital ulcers, arthritis and uveitis who was admitted because of occipital cephalgia, mild lethargy, oral aphthous stomatitis and gait disturbance. Physical examination showed a slight right hemiparesis with right patella and ankle clonus. The right plantar response was

extensor. There was evidence of dysmetria and ataxia. His temperature was 37.6°C. There was no clinical evidence of meningeal irritation. The optic fundi appeared normal. The cerebrospinal fluid (CSF) was clear and contained 70 cells/mm<sup>3</sup> of which 75% were lymphocytes; the glucose was 3.7 mmol/L (normal = 2.2-3.9 mmol/L) and the protein 60 mg per 100 ml. The basic myelin protein was normal. There was no evidence of infection caused by bacteria, fungi or neurotropic viruses in the CSF. There was no increase in the serum antibody titres against respiratory, cytomegalovirus, measles, herpes simplex, or varicella-zoster viruses. CT brain scan was normal. MRI showed a pathological area in the pons extending upwards to the cerebral peduncles and downwards to the medulla oblongata, which was clearly showed by the higher T2 intensity (fig a). Clinical symptoms promptly disappeared after treatment with steroids, and MRI two months later showed no abnormalities (fig b).

The full recovery in some patients with neuro-Behçet's disease after steroid therapy suggests a lesional mechanism different from ischaemic necrosis due to vasculitis. An immunologically mediated demyelinating process seems to be probable. In our case, the wide extension of pathological T2-weighted images and their resolution after treatment support this mechanism. Taking into account the higher value of MRI over radiograph CT examination in the diagnosis of demyelinating lesions, MRI could be the method of choice in the diagnosis of neurological involvement in BD.

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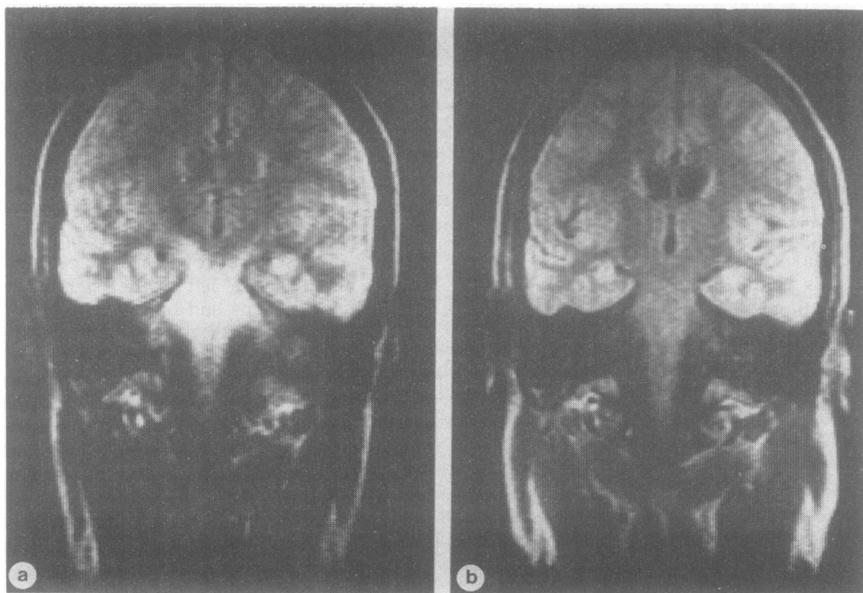


Figure (a) MRI of T2-weighted images showing a pathological area in the pons extending upwards to the cerebral peduncles and downwards to the medulla oblongata. (b) MRI performed two months later showing no abnormalities.