Stuttering pituitary apoplexy resembling meningitis

In the first clear description of pituitary apoplexy Brougham *et al*[1] suggested that the syndrome should be considered as the abrupt development of headache, amblyopia, diplopia, drowsiness, confusion or coma. These clinical features have now become well established.[2,3] However, Brougham *et al* mention one case of haemorrhage into a pituitary tumour simulating rupture of an intracranial aneurysm where ophthalmoplegia was delayed several days.[4] We have seen two cases where delay in diagnosis occurred as a result of the subacute onset of symptoms. The clinical picture was further confused by the finding of a highly cellular cerebrospinal fluid (CSF).

The first case was a 50 year old warehouse attendant who had a five day history of generalised headache and vomiting with two days of double vision. His relatives had noticed an intermittent droop of the right eyelid. He had previously been in good health apart from the drainage of a "cold abscess" in early childhood. He drank about 20 pints of beer each week.

Examination revealed a slight pyrexia (37·5°) with limitation of neck flexion but no neck stiffness. He had a partial left third nerve palsy with a dilated but reactive left pupil and a partial left ptosis. There was no field defect and extra-ocular movements appeared full. Power and tone was normal in the limbs. The biceps reflexes could be elicited on reinforcement but otherwise tendon reflexes were all absent. Plantar responses were flexor and sensory testing was normal. An unenhanced CT scan was performed to exclude an abscess. Lumbar puncture revealed opalescent CSF under 19 cms pressure which contained a concentration of 77 mg% of protein with 52 white cells (5% polymorphs, 91% lymphocytes, 4% macrophages). No organisms were seen and the concentration of sugar was normal. An EEG showed an excess of mainly posteriorly distribute slow waves.

A provisional clinical diagnosis of a subacute meningoencephalitis was made and treatment was started with intravenous Acyclovir and thiamine. Over the next three days there was a deterioration in his clinical state with a rising pyrexia to 39°C. He developed bilateral ptosis with impaired adduction and elevation of both eyes. Visual acuity in the right eye was reduced to counting fingers at one metre and a right altitudinal field defect developed which extended below the horizontal meridian.

A high resolution enhanced CT scan of the orbits revealed a pituitary adenoma with some suprasellar extension. Treatment with Prednisolone was started immediately with rapid improvement of visual acuity to 6/7·5 in the effected eye. Endocrine studies revealed a normal prolactin (< 65 units) but low TSH (0·2 uU/L) and free T4 (5·5 pm/L). He subsequently had an uneventful transphenoidal hypophysectomy and pathological examination of the specimen was consistent with the eventual diagnosis of infarction in a pituitary adenoma.

The second case was a 45 year old printer's assistant who presented with symptoms of the carpal tunnel syndrome bilaterally. It was noted, a year later, that he had large hands and other features of acromegaly. A pituitary adenoma was confirmed on CT scan. Whilst awaiting a transphenoidal hypophysectomy he awoke with sudden onset frontal headache and vomited. The following day he was a little better but three days later he became confused and complained of visual impairment. He was admitted to a local hospital where he was found to have neck stiffness and a temperature of 40°C. A lumbar puncture revealed CSF which was xanthochromic and under increased pressure with raised protein concentration and a mixed pleocytosis. The following day he developed peri-orbital and subconjunctival oedema and bilateral sixth nerve palsies. His deep tenden reflexes were absent and his conscious level began to deteriorate. He was transferred to a specialist unit where he was found to be disoriented in time and place and pyrexial. Periorbital oedema, bruising around the left eye and marked neck stiffness were noted. Visual acuity was 3/60 on the right and 6/18 on the left and he was unable to identify any of the Ishihara test plates. There was a temporal hemianopia to colour in the left eye and a dense central scotoma on the right on confrontation. There was mild disc swelling with absent venous pulsation. Lateral rectus weakness was complete and bilateral and upward movement of the right eye was limited. The only other abnormal finding was total areflexia.

The full blood count was normal as was the urea although the serum sodium was low at 126 mm/L. The CSF contained 1·2 G/L of protein with a pressure of 30 cms of CSF and a white cell count of 900 (10^6/L) of which 79% were polymorphs and the rest lymphocytes. There were 20 (10^6/L) red cells and a normal CSF glucose. Urine and serum osmolarities suggested inappropriate secretion of ADH. A plain skull radiograph showed an enlarged pituitary fossa and opacification of the sphenoid sinus. CT scan revealed a suprasellar mass with lateral extension. Serum cortisol was low (60 mu/L) as were levels of total thyroxine, TSH, prolactin, FSH and LH confirming panhypopituitarism. A random estimation of growth hormone was slightly elevated (16·6 mu/L). In view of the gradual evolution of symptoms and the cellular CSF a provisional diagnosis of meningitis was made by the referring hospital. The CSF proved to be sterile and field defects and cranial nerve signs resolved when antibiotics were withdrawn and he was treated with intravenous steroids and thyroxine.

The presence of inflammatory changes in the CSF in some cases of pituitary apoplexy is well documented in the literature. Bjerre and Lindholm[5] have recently emphasised the occurrence of "sterile meningitis" as an important feature of pituitary apoplexy and report six cases where CSF examination revealed an elevated leucocyte and or polymorph count. Four further cases in the literature are cited. The presence of CSF pleocytosis usually presents little diagnostic concern in patients with a typical acute onset where the diagnosis is confirmed by urgent CT scanning. The subacute or stuttering onset in our cases made diagnosis difficult and prompted this report. In the first case the absence of a field defect on presentation compounded the difficulty in diagnosis. In the second case the lack of awareness of this mode of presentation of pituitary apoplexy delayed establishing an accurate diagnosis despite the previous diagnosis of a pituitary tumour.

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JB WINER, G PLANT
National Hospital for Nervous Diseases, Queen Square, London

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Intracranial haemorrhages occurring in the idiopathic hypereosinophilic syndrome

The hypereosinophilic syndrome (HES) can produce a range of neurological disorders. Three major patterns of neurological involvement have been described: encephalopathy, sensory polyneuropathy, and central nervous system thrombo emboli.

We describe an elderly woman with this syndrome who presented with right temporal lobe and left cerebellar hemisphere haemorrhages.

An Indian woman aged 72 years first presented to our hospital in 1980 with general malaise and a nocturnal cough. Examination at that time revealed a mild expiratory wheeze. Her haemoglobin was normal but the white cell count was elevated at 46 000 × 10^9/l (78% eosinophils and 15% neutrophils). Her platelets, coagulation screen and auto antibodies were normal. A chest radiograph showed old calcified apical foci with some ill defined shadowing in the left mid zone. A bone marrow examination revealed eosinophilia with both the red and white cell series severely depressed. There was no excess of blasts and no abnormal cells.

A diagnosis of HES was made and she was started on a reducing dose of prednisolone. On discharge her white cell count was 10·7 × 10^9/l with 1% eosinophils. Symptomatically she had improved and her nocturnal cough had disappeared.

She remained well on a small dose of prednisolone and continued to have a normal eosinophil count over the next five years. She was lost to follow up in 1985 but according to her general practitioner she continued to take prednisolone until 1987. She was admitted in February 1989 after collapsing at home. According to her son the event was sudden and without warning. There was no history of head trauma or of a seizure. She was a non smoker, non drinker and was not on any medication. Systems enquiry, by proxy, was normal.

On examination she was unconscious (Glasgow coma scale Grade IV), apyrexial but with no neck stiffness. General examination was normal, BP 170/80.

On neurological examination, her pupils were mid point and fixed. Fundoscopy was normal and she had no obvious facial asymmetry. Her eye movements were difficult to assess and she had a depressed gag reflex.

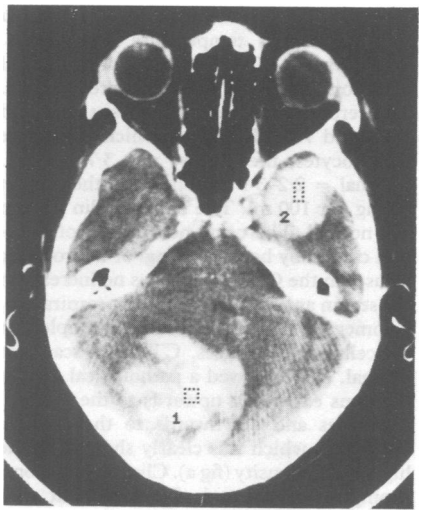

Figure CT scan showing right temporal lobe and left cerebellar hemisphere haemorrhages.

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 × 10^9/l with a differential of eosinophils 32·81 and neutrophils 12·18 × 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.[1] It is defined as persistent eosinophilia (> 1500 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[2] 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.

S ROCHE,
S CROSS,
B KAUFMAN,
Department of Medicine for the Elderly, Central Middlesex Hospital, Acton Lane, London

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Correspondence to: Dr Roche, Department of Medicine for the Elderly, St Charles Hospital, Exmoor Street, London W10, United Kingdom.

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 meningoplasty 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 cerebospinal 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 microaerophilic 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.[1] 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.[2] 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[1 3 4] which is usually associated with vertebral osteomyelitis or bacteremia. The diagnosis has recently been facilitated with the introduction of spinal CT.[5 6]

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.[7]

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.[8]

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.

R HARRIES-JONES
M HERNANDEZ-BRONCHUD
P ANSLOW
CJ DAVIES
Radcliffe Infirmary, Oxford

Correspondence to: Dr R Harries-Jones, Hereford General Hospital, Hereford HR1 2PA.

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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