Four days later CT scanning was repeated, without contrast, to assess the extent of infarction. An ill-defined hypodense region was visible in the basal part of the left temporal lobe. The adjacent slice showed a slightly haemorrhagic component, with some compression of the sulci and ambient cistern (fig 1a). This was even clearer on the next slice (fig 1b). Blood pressure was 150/90 mmHg and the patient's level of consciousness suddenly deteriorated (Glasgow Coma Scale Score E1 M3 V1, with small, unreactive pupils).

CT scan fifteen minutes later showed fresh haemorrhage, heterogeneously distributed over at least three foci, with a marked shift to the right (fig 1, C and D). At this time the level of anticoagulation remained appropriate (2.4 INR). The anticoagulation was reversed with vitamin K, the coagulation factors II, VII, IX, X, and fresh frozen plasma. Nevertheless, the patient died after several hours. Permission for necropsy was not obtained.

In this patient with bacterial endocarditis the brain infarct may have been caused by an infected embolus from the heart. It is uncertain whether arteritis was a factor in the chain of events. CT scanning provided some information on the origin of the haemorrhagic infarction. Necropsy demonstration of remnants of old haemorrhages at the periphery of the ICH, in the shape of fibrin globes and deposits of haemosiderin suggest^{2,6} that intracerebral bleeding can originate at more than one site. As soon as a small blood vessel has ruptured, the pressure around the centre of the bleeding would increase, which would cause other small arteries in the periphery of the haematoma to be compressed and damaged, with further haemorrhages as a result. In our case this process may have been completed within a few minutes, under the influence of anticoagulant treatment. An alternative explanation is that the haemorrhages were not consecutive but truly simultaneous, caused by reperfusion of a damaged capillary bed.

Because of the AC treatment, a normal clot was not formed and there was sedimentation of erythrocytes at the bottom of the haematoma. This image may be characteristic of ICH under AC treatment,⁷ or else of very recent ICH.⁸ Clotting of extravasated blood is delayed in case of AC-treatment and may not be completed if a CT scan is performed soon after a stroke. In general, extravasated blood has greater density than normal brain tissue. On CT scanning with a haemoglobin level below 5 mmol/l the density of extravasated blood can be even less dense than normal brain tissue.^{9,10} In case of low haemoglobin the density may later increase by haemoconcentration.⁹ In our patient, the haematoma was immediately visible as a hyperdense lesion on the CT scan, probably because of the concentration of erythrocytes by sedimentation.

The unique pictures of this "ongoing haemorrhage" confirm that haemorrhagic infarction occurring during anticoagulant therapy may show multifoci and can be recognised on CT-scanning, even in anaemic patients.

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Internuclear ophthalmoplegia in giant-cell arteritis

Patients with giant cell arteritis (GCA) occasionally complain of double vision.^{1,2} Arteritic involvement of the ophthalmic artery within the orbit is usually the cause.^{3,4} We describe two patients with GCA proven at biopsy in whom double vision was due to internuclear ophthalmoplegia (INO). Involvement of the medial longitudinal fasciculus (MLF) in GCA has not been previously reported.

The first patient was an 81 year old woman who presented with a three month history of anorexia and 14 kg weight loss. She was confused. One month before admission she had developed diplopia on looking to the left. She also complained of head pains over her vertex "as if gnomes were chipping away at my brain", and she had some jaw stiffness. Examination revealed a visual acuity in her right eye of 6/7.5, N6 and in her left eye of 6/12, N6. Colour vision was normal in both eyes and visual fields were full. Fundoscopy revealed moderate lens opacities and myopic discs. Pupillary responses were normal. There was a right INO. She was able to converge normally. The temporal arteries were variably tender though pulsated normally. Her erythrocyte sedimentation rate (ESR) was 84 mm/hour and haemoglobin (Hb) 9 gm/100 ml. Temporal artery biopsy showed transmural inflammation with disruption of both the internal and external elastic laminae. Treatment with steroids led to resolution of her behavioural state and head pains within two days. When she left

hospital five days later, her eye movements had improved considerably though the internuclear ophthalmoplegia persisted.

The second patient was an 85 year old man who gave a four month history of persistent left periorbital head pains, drooping of the left upper lid, 7 kg weight loss and some scalp tenderness. Two weeks before admission he developed diplopia with horizontal image separation on looking to his right. His double vision developed whilst playing bridge and he accused his friend of having too many cards and cheating. Examination of his eyes revealed visual acuities of 6/9, N5 on the right and 6/6, N5 on the left. Colour vision was normal. A small right choroidal haemorrhage was seen in the right eye. The left fundus was normal. His visual fields were normal. A left Horner's syndrome was present. There was a left INO with associated vertical nystagmus. He was able to converge. The temporal arteries were clinically normal. His ESR was 100 mm/hour, Hb 10 gm/100 ml and the alkaline phosphatase was raised. Temporal artery biopsy revealed near complete occlusion with intimal proliferation and destruction of media and elastica. A prominent chronic inflammatory infiltrate with giant cells was present throughout the vessel wall. His head pains and scalp tenderness responded overnight to steroid treatment and at the time of discharge the Horner's syndrome had resolved and the internuclear ophthalmoplegia was improving.

Ten to 15% of patients with GCA may complain of double vision.^{1,2} Eye movement abnormalities in this condition are due either to arteritic involvement of the vasa nervorum of the oculomotor nerves,³ or to ischaemic necrosis of the extraocular muscles themselves.⁴ Disease affecting the ophthalmic artery within the orbit is the cause in both cases.

However, in our patients INO was the cause of diplopia. The lesion is in the MLF which runs from the pons to the superior colliculus, and the blood supply to this area is from perforating branches of the basilar artery. As the extradural portions of the vertebral arteries are known to be involved in the arteritic process in almost all cases of GCA,³ we presume that the perforating branches to the MLF became involved on the basis of embolisation. It is perhaps surprising that INO has not been reported before in GCA although presumably this is due to the rich anastomotic supply to the pons and midbrain.

The Horner's syndrome in patient two proved pharmacologically to be post-ganglionic and was probably due to a lesion in the "carotid" sympathetic fibre. Many patients with GCA are reported with ptosis and miotic pupils although verification of sympathetic denervation is rare.⁵

These cases highlight the diversity of clinical presentations of GCA and support the maxim that all elderly patients presenting with neurological disease of possible vascular origin deserve an ESR and consideration of temporal artery biopsy and perhaps a therapeutic trial of steroids.

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Periodic fever: an unusual manifestation of a recurrent Rathke's cleft

Rathke's cleft cysts are generally asymptomatic intrasellar cysts. Occasionally they grow and compress surrounding structures and this can lead to various neurological, neuro-ophthalmological and endocrinological symptoms. The report deals with a recurrent Rathke's cleft cyst which was associated with periodic fever; permanent drainage gave remarkable relief of symptoms.

A 30 year old woman was referred in December 1983 with a 17 month history of visual impairment, occasional headaches, and irregular menses. She was slow of thought, otherwise physical examination was normal. Perimetry showed bitemporal hemianopia. Routine blood and urine analysis gave normal results. Endocrine evaluation revealed hypocortisolaemia and hypogonadotropinism of pituitary origin. Moderate hyperprolactinaemia was also found. A contrast-enhanced CT scan of the brain showed a well-circumscribed intra- and suprasellar homogeneous low density lesion (15 × 15 × 18 mm) with capsular enhancement. No evidence of calcification was found.

In May 1984 a craniotomy was performed. This showed a cystic tumour with compression of the optic chiasm. Needle aspiration yielded 4 ml sterile fluid containing leukocytes. Fenestration of the cyst was performed. The histological features were consistent with a Rathke's cleft cyst accompanied by signs of chronic inflammatory reaction. Postoperatively she remained well and her visual fields improved, but panhypopituitarism made replacement therapy necessary.

From April 1986 she suffered from attacks of fever, which recurred at two week intervals and lasted six hours. These attacks occurred during her sleep and were associated with feelings of cold and discomfort, pilo-erection, and frequent chills. The rectal temperature rose to 39°C. This was followed by a three hour period of excessive sweating during which the body temperature gradually returned to normal.

In December 1986 she was admitted to hospital because of severe bifrontal headache and vomiting. Routine laboratory investigation revealed no abnormalities. A CT scan of the brain showed a homogeneous low-density lesion (20 × 20 × 20 mm), situated in the midline above the sella turcica. Both lateral ventricles were dilated. A bifrontal ventriculoatrial shunt with a Holter-Hausner normal pressure valve was installed. The cerebrospinal fluid (CSF) pressure was considerably elevated (50 cm H₂O); the constituents were normal.

This operation produced only slight improvement of the headaches and had no effect on the fever attacks, which now recurred about once a week at random times during the day. Magnetic resonance imaging (MRI) of the brain was performed on a Gyroscan S5 operating with a field strength of 0.5 Tesla. Coronal and sagittal T1 and T2 weighted spin echo pulse sequences were used (TR/TE 400-600/30 and 2000/50-100, respectively). This clearly demonstrated suprasellar extension of the cyst, which reached to the level of the foramen of Monro and caused compression of hypothalamic structures. The cystic lesion was non-homogeneous and isointense to slightly hyperintense on the T1-weighted image compared with the signal intensity of CSF and slightly hyperintense on the T2-weighted image. Adequate drainage of the ventricles was established (fig). Under CT guidance, drainage of the cyst was accomplished by stereotactic introduction of a drain tip into the cyst. Aspiration yielded 4 ml of fluid.

Microscopically, this fluid contained necrotic material. Cultures were again negative. A subgaleal reservoir was mounted and connected to the ventriculoatrial system. MRI of the brain performed one month later showed that the cyst had almost disappeared. Almost two years after this operation the

patient is still doing well. The headaches ceased immediately and the fever attacks became much less frequent and less severe within one month after drainage of the cyst. About once or twice a month she wakes up for a few minutes during the night feeling cold and shivering slightly without any change in body temperature.

In the few reports of Rathke's cleft cysts that have produced symptoms, the most common features are headache, hypopituitarism, visual symptoms, and hydrocephalus.^{1,2} Fever has rarely been reported in association with Rathke's cleft cyst.^{3,4} In our patient, the periodic attacks of fever developed more than one year after the first operation, and eight months before the obstructive hydrocephalus became symptomatic. Because large oscillations of intracranial pressure occur during the paradoxical stage of sleep, the initial nocturnal occurrence of fever attacks might represent related phenomena of elevated intracranial pressure.⁵ However, after ventriculoatrial shunting the fever attacks persisted and only permanent drainage of the cyst resulted in rapid clinical improvement, suggesting that local compression of the hypothalamus by the cyst is more essential than elevated intracranial pressure in general. The initial two week cycle of fever attacks and suprasellar extension of the cyst further suggest a hypothalamic disturbance.

Two other possible causes of fever in Rathke's cleft cyst should be considered. In the first place, the signs of chronic inflammatory reaction in the cyst wall suggest the presence of cytokines, which may act on the hypothalamus to induce fever. This is, however, contradicted by the frequently documented microscopic signs of inflammation in the cyst wall without the development of fever.⁶ Second, the contents of the cyst may leak into the subarachnoid space and cause aseptic meningitis. This seems unlikely because our patient did not show signs of meningitis and the CSF was normal.

In conclusion, suprasellar extension of a Rathke's cleft cyst may cause signs and symptoms of hydrocephalus and hypothalamic dysfunction. This case illustrates the occurrence of periodic fever as a hypothalamic feature of suprasellar extension of a Rathke's cleft cyst.

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Figure Coronal T1 and T2 weighted images demonstrate the suprasellar component. Note the flow void phenomenon in the ventriculoatrial system bifrontally.

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