together with prominent cortical veins. Haemostasis was achieved with some difficulty, and without the benefit of angiography no attempt was made to remove the lesion. Nevertheless small biopsies were taken.

After operation, the patient was ventilated, sedated, and paralysed. A subsequent chest radiograph showed that there was a well-circumscribed 4 × 4 cm lesion in the upper left lobe.

Histology of the clot and tissue taken showed that the brain lesion was a metastatic deposit (fig 2A). The primary was initially assumed to be a carcinoma of the lung, which had metastasised to the brain. The patient was a heavy smoker and the prognosis was initially thought to be extremely poor. Ensuing examination, however, showed a firm nodular right testicle. Direct questioning of his wife later confirmed that this had been present for at least three months.

Grossly raised serum HCG was detected (13 815 IU) and immunocytological examination of the original cerebral tissue confirmed the presence of a choriocarcinoma (fig 2B). On the basis of this the patient was started on intravenous etoposide and cis-platinum. At review two months later his condition had improved. Although he had a residual left hemiparesis and left homonymous hemianopia, he had retained full higher faculties. Tumour markers have since declined to 4 IU.

This case serves to illustrate several points: firstly, an intracerebral haemorrhage in a young person may represent an underlying neoplasm; secondly, a careful history from relatives and a meticulous examination of an unconscious patient may produce information of great relevance; thirdly, the importance of biopsy in vascular lesions of undetermined aetiology; and fourthly, the prognosis of a choriocarcinoma metastasising to the brain is relatively good.

All tumours metastasising to the brain have the potential to bleed, especially malignant melanomas. In a similar case report to this one, where two patients with known brain metastases from a testicular tumour were diagnosed, intratumoural bleeding led to a right hemiparesis with sensory involvement and eventual death in one patient, and a left hemiparesis and eventual death in the other. These deaths occurred despite the early commencement of chemotherapy.

Another case study reported complete remission of the growth of a metastatic teratoma from malignant testicular tumour, using salvage chemotherapy, despite an intratumoural haemorrhage. Pulmonary metastases in a patient with a known germ cell tumour should stimulate a search for further metastases in the CNS. Several studies show that nearly all patients diagnosed with brain metastasis from a testicular germ cell carcinoma already had radiologically detectable pulmonary metastatic disease.

One problem highlighted from the literature is that the efficacy of chemotherapy against brain metastasis is restricted by poor penetration of the blood-brain barrier. Radiation therapy is also limited and cannot eradicate a tumour. This has prompted some authors to advocate surgery in the management of a brain metastasis before chemotherapy, or if no response to chemotherapy was achieved. Elective surgical removal of accessible brain metastases larger than 1 cm, as indicated by Jelsma and Carroll, would avoid the possibility of a spontaneous intratumoural haemorrhage, or massive tumour lysis after chemotherapy associated with a haemorrhage, which can lead to such devastating results in an otherwise treatable condition.

We acknowledge the Department of Medical Illustration, Greenwich District Hospital, London.

J TIMOTHY A SOPRA M SHARR B DODHI
Regional Neurosurgical Unit, Brook General Hospital, Woolwich, London SE18 4LW, UK.

Correspondence to: Dr Jake Timothy, Department of Neurosurgery, Brook General Hospital, Shooter’s Hill Road, Woolwich, London SE18 4LW, UK.

Intracranial fusiform aneurysms in von Recklinghausen’s disease: case report and literature review

The patient was a 27 year old man who presented to our hospital with symptoms of left sided weakness—initially involving only the arm—but subsequently affecting the leg during the preceding two years. Three months before admission he began to experience spontaneous clonus of his left arm and leg that impaired his ability to work and thus precipitated his presentation. He also complained of an occipital headache (sometimes associated with vertigo) as well as vertical diplopia. He denied any seizure activity or symptoms attributable to his neurofibromatosis. There was no family history of note.

On examination he was a well looking man with prominent axillary freckling as well as multiple (more than 10) large (many larger than 3–4 cm in diameter) café au lait spots distributed over his trunk and limbs. He had features in keeping with an upper motor neuron weakness of his left arm and leg as well as impaired sensation to all modalities over the same area. He also had a sensory deficit over the area supplied by the trigeminal nerve on the left side. His left pupil was 2 mm larger than his right and he was unable to raise his left eye in the abducted position, suggesting weakness of the superior rectus muscle. He had an ataxic nystagmus as well as incoordination of his left arm and leg. His gait was ataxic. His blood pressure was 130/80 mm Hg and examination of his cardiovascular, respiratory and other systems was entirely normal. He had no Lisch nodules.

It was considered that a lesion in the upper midbrain, involving the spinal, medial, and trigeminal lemnisci (carrying crossed sensory fibres from the body and face), the corticospinal tracts (carrying motor fibres), and the superior cerebellar peduncle could account for most of the neurological signs shown by this

References
Summary of the literature concerning intracranial fusiform aneurysms

**Clinical details** | **Site of aneurysm** | **Treatment** | **Reference**
--- | --- | --- | ---
19 year old man. Right retro-orbital headache and diplopia (VP nerve palsy) Symptoms developed over night | Petrous apex near the cavernous sinus | Seleverstone clamp of right internal carotid artery with gradual occlusion | Muhohen et al.
36 year old man. Right temporal headaches of 3 months duration | Saccular aneurysm and multiple fusiform aneurysms at right middle carotid artery trifurcation | Aneurysmal clip of saccular aneurysm and coating of fusiform aneurysms with methyl methacrylate | Muhohen et al.
18 year old woman Asymptomatic 27 year old man Left sided weakness (arm and leg); left hemisensory disturbance (arm, leg, and face); left sided incoordination and left superior rectus weakness. Symptoms of 1 year duration | Right petrous carotid artery | Nil | Frank et al.
| Right petrous and cavernous internal carotid artery | Aspirin and β-blocker |

MRI angiogram. Two intracranial fusiform aneurysms are demonstrated: of the internal carotid on the right and of the basilar artery in the centre. In each case, one arrow points to blood flowing through the aneurysm and the other arrow to the region of slow flow or clot within the aneurysm.

Mills’ syndrome: ascending (or descending) progressive hemiplegia: a hemiplegic form of primary lateral sclerosis?

We describe two patients with slowly progressive hemiplegia. These two cases bring to mind a rare clinical syndrome described in 1900 by Mills—namely, ascending (or, less often, descending) progressive hemiplegia. 1 Mills claimed that this disorder was a new form of degenerative disease characterised by progressive disturbance of the corticospinal pyramidal pathways. Despite its age, the concept of Mills’ syndrome is still controversial. Indeed, a number of pathogenetic factors can cause such clinical findings, and the cases that remain isolated and can be considered as primary are rare.

Case 1, a 49 year old right handed woman with no previous personal or family medical history, complained in 1975 of motor deficiency on the right side of the body. The initial symptoms were weakness of the right foot and leg, which slowly progressed to the thigh. The patient was admitted to hospital in 1981 at the age of 55. Physical examination showed right Babinski’s and Hoffmann’s signs. Tendon reflexes were very pronounced on the right side. The motor deficiency was strictly limited to the right lower limb. There was no sensory loss. An EEG, CSF examination (cytology), EMG, brain CT, and contrast myelography were normal. Re-examination in 1987 showed that the disability had increased: there was a pyramidal gait and a distal motor deficit of the right arm associated with a moderate hypertonia. Facial mobility was normal. No sensory deficit was noted. Routine laboratory tests were normal. Serological tests for syphilis were negative. Examination of CSF showed a slight increase in protein content (0·60 g/l) without pleocytosis; immunological tests did not disclose intrathecal synthesis of immunoglobulin or oligoclonal IgG bands. Visual, auditory, and somatosensory evoked potentials were normal. EMG was performed a year later with normal findings. MRI of the brain and spinal cord did not show any lesions. Spinal angiography showed no evidence of vascular abnormality. We have not had the opportunity of re-examining this patient since 1987 but the referring physician has kept us informed. In April 1993 after 18 years of evolution he noted a persistent right hemiparesis with hyper-reflexia, Babinski’s and Hoffmann’s signs, and ankle clonus; there was urinary urgency but no sensory loss, fasciculations, or atomyotrophy. Brain and spinal cord MRI and a further EMG were still normal.

Case 2, a 25 year old right handed female nurse first noticed weakness in her left hand in 1976. In ensuing years the weakness progressively spread to the whole arm. In 1985 weakness appeared in her left leg and she began to have trouble walking in the patient had no personal or family history of neurological disorders. In 1989 physical examination revealed a global weakness of the left upper and lower extremities with pyramidal gait and no sensory deficit. Tendon reflexes were increased in all four limbs with Babinski’s and Hoffmann’s signs on the left. A facial asymmetry was noted on her left side when the patient was asked to make a face. Mild wasting without fasciculation was noted in the left leg. Laboratory findings, including assessment of the