there was wasting and weakness of deltoid, biceps, and brachioradialis bilaterally; biceps and supinator reflexes were absent bilaterally but the triceps reflexes were present and there were no sensory symptoms or signs. Tone, power, and reflexes in the lower limbs were normal and both plantars were flexor. Abdominal wall movements with respiration were paradoxical and vital capacity fell from 3·2 litres sitting to 1·9 litres lying. The ESR was now 90 mm/hour. Electrical studies showed normal nerve conduction with evidence of acute C5 and C6 denervation bilaterally. An MRI scan of the cervical region and cerebrospinal fluid examination were normal.

A gradual improvement in the patient's condition occurred but the ESR remained high and he continued to complain of occasional headaches. Eleven weeks after presentation a temporal artery biopsy was performed. Microscopy showed features typical of temporal arteritis with numerous giant cells within the wall of the artery, intimal fibrosis, and fragmentation of the internal elastic lamina. Steroid treatment was started immediately with prompt relief of the remaining headache and malaise; the strength in both arms and the exertional dyspnoea continued to improve.

Observation over the next 12 months failed to find any other cause for his symptoms and signs. MRI (including MR angiography) of the brain showed that the pituitary tumour and the aneurysm of the ophthalmic artery had not increased in size; both are being managed conservatively. He remains well on a gradually reducing course of steroids and clinical examination of the eyes and upper limbs is now normal.

In this case temporal arteritis was associated with a third nerve palsy, a bilateral classical internuclear ophthalmoplegia, and a radiculopathy affecting the fifth and sixth anterior cervical roots bilaterally. On clinical grounds it seemed that the patient had associated impairment of diaphragm function but this was not assessed formally by transdiaphragmatic pressure measurements. Although other vascular risk factors were present, and a pituitary tumour, these could not be implicated in the pathogenesis of all the neurological deficits seen.

The third nerve palsy was probably due to involvement of branches of the intraorbital ophthalmic artery as there were no features to suggest a nuclear or a fascicular third nerve palsy. A bilateral classical internuclear ophthalmoplegia is a rarely reported complication of temporal arteritis[1 2] and is probably due to emboli from the vertebral artery occluding perforating branches of the basilar artery. Both the vertebral and ophthalmic arteries are recognised as being frequently and severely affected in temporal arteritis.[3]

A cervical radiculopathy is an unusual feature of temporal arteritis and the aetiology is less clear. Five cases have been reported[4-7] and in all cases the fifth anterior cervical root has been involved, unilaterally or bilaterally, and in only one case were there sensory signs; in one case there was also evidence of unilateral involvement of the sixth and seventh anterior cervical roots. Constitutional symptoms but no other neurological signs were present in the four cases for which details are available and in all of these patients considerable improvement occurred with prednisolone treatment. Two other cases of upper limb paresis occuring in patients with temporal arteritis have been reported but the aetiology of the weakness was not established.[8]

A review of the blood supply of the cervical spinal cord and roots provides a possible explanation for the pattern of root involvement seen. The main supply comes from the anterior and posterior spinal arteries, branches of the vertebral arteries. These vessels anastomose with anterior and posterior radicular arteries that enter the spinal canal alongside their corresponding roots. There is great variation in the number, size, and location of the radicular arteries,[9] but if present those to the upper six cervical roots also originate from the vertebral arteries whereas those for C7, C8, and T1 originate from other vessels in the vicinity including the thyrocervical trunk and the costocervical trunk. Therefore the first six cervical segments and roots are dependent solely on a blood supply originating from the vertebral arteries.

We think that the most likely explanation for the radiculopathy is arteritic or embolic occlusion of the relevant radicular artery due to involvement of the parent vertebral artery. A myelopathy has not been seen in association with the radiculopathies suggesting that essential downwards flow in the spinal arteries is maintained. This downwards flow may be able to compensate via anastomotic channels for involvement of radicular arteries to the upper cervical roots but not to the lowest "vertebral artery dependent" roots, C5 and C6; this may explain why these roots are so commonly affected. It has been suggested that the impairments seen are due to damage to the cords of the brachial plexus caused by a vasculitis in the neighbouring subclavian and axillary arteries[7] but this seems unlikely as sensory involvement has been a feature in only one case and was restricted to a single dermatome.

This case serves as a reminder of the many neurological signs that may occur in temporal arteritis and of their natural history. The diagnosis of temporal arteritis should always be considered in patients with multiple or fluctuating neurological signs involving the vertebrobasilar circulation, even if the ESR is not grossly raised at presentation.

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Methylenetetrahydrofolate reductase deficiency revealed by a neuropathy in a psychotic adult

5,10-Methylenetetrahydrofolate reductase (MTHFR; EC 1.1.1.68) deficiency, although rare, may be suspected when homocystinuria is detected. Homocystinuria is also present in other inherited disorders such as cystathionine β synthase deficiency or some inborn errors of cobalamin metabolism.[1] Deficiency of MTHFR, an autosomal recessively inherited error of folate metabolism, leads to an inability to synthesise 5-methyltetrahydrofolate, the major form of folate in fluids and tissues, in amounts sufficient for remethylation of homocysteine to methionine. Therefore homocysteine accumulates in plasma whereas methionine concentrations are normal or decreased. Deficiency of MTHFR is associated with a wide spectrum of neurological abnormalities, most often convulsions and mental retardation[1]; rarely psychotic symptoms.[2] Age of diagnosis has ranged from the first days of life to adolescence.[2] Recently MTHFR deficiency was detected in a young adult of a family with premature vascular disease.[3] We report a case of MTHFR deficiency diagnosed in a psychotic adult with neuropathy.

A 45 year old Chinese woman was referred to the hospital for a gait disturbance over a week. She had lived in France for 15 years but spoke very poor French. The first of four siblings of unrelated healthy parents, her early growth and development had been normal. A schizophreniform disorder appeared at age 19. She had four children and no abortions. No relapse occurred during her pregnancies, but several psychotic episodes happened 10 years later and were treated with perphenazine enanthate. Since then, she was almost socially adapted despite probable progressive cognitive impairment with increasing difficulties in speaking and writing French and eventually speaking Chinese. Withdrawal, hallucinations, delusions, and anorexia led to psychiatric consultation. She was treated with chlorpromazine. Paranoid symptoms decreased in three weeks, and then she developed a weakness of the lower limbs with dysaesthesia.

On admission she had a flaccid symmetrical paraplegia and severely reduced sensation, predominant distally. Tendon reflexes were absent in the legs and weak in the arms. Cranial nerves were normal. She had no sphincter problems. She had a mild bilateral genu recurvatum but no noteworthy morphological abnormalities. She was oriented in space and time but was very slow, and timid. She refused food, had difficulties in sleeping, collected food wastes, and was suspicious. Cognitive evaluation was impossible and only part of the investigations required were done because of the patient's negative attitude.

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Cerebrospinal fluid was normal (leucocytes 0·6 μl; protein 0·43 g/l). Electromyography showed decreased compound muscle action potentials in tibial nerves and signs of degeneration of motor axons (fibrillation potentials). Motor nerve conduction velocities, and distal and F wave latencies were normal. Compound muscle action potential amplitudes were reduced in tibial nerves (right 1·5 mV; left 0·2 mV). Sensory nerve conduction velocity was normal in all four limbs but potential amplitudes were slightly reduced in the sural nerves (right: 6 μV left: 8 μV). Blood chemistry and cell count were normal except for a mild macrocytosis (mean corpuscular volume 99 fl; haemoglobin 11·6 g/dl). Serum cobalamin concentration was 240 pg/ml (normal 200–1000 pg/ml), but serum and red cell folate were lowered, respectively 2·4 ng/ml (normal >3 ng/ml) and 146 ng/ml (normal > 150 ng/ml). Serum thyroxine, creatine, phosphokinase, lactic dehydrogenase and aldolase were normal. Tests for heavy metals and coproporphyrins in urine were negative. Amino acid chromatography detected homocystinuria (16 μmol/1 per 24 hours) whereas no homocystinuria was found in controls. Because of the clinical features, homocystinuria, and lowered folates, a disturbance of folate metabolism was suspected. Activity of MTHFR was assayed on skin fibroblast according to the method described by Kutzbach and Stokstad.[4] MTHFR in the patient was 1 nmol/h/mg protein (mean control value in 20 normal subjects 3·8 nmol/h/mg protein with a range of 2·6 to 5). An artefactual decrease of this enzyme activity due to folate deficiency was excluded because cells were grown in a complete RPMI medium containing folic acid.

She was treated with infusion of thiamine, pyridoxine, cyanocobalamin, and intramuscular folinic acid for nine days; these were then given by mouth. Gait and behaviour improved in two weeks. Homocystinuria and delusions disappeared within 15 days, neuroleptic drugs were stopped, and contact improved progressively. Six weeks after discharge, a new paranoid episode occurred, without neurogenous relapse. She was admitted to a hospital psychiatry department and treated with loxapine with success. Folic acid was continued. Five months later, mean corpuscular volume was 93 fl, haemoglobin 13 g/dl. Her gait was normal, tendon reflexes were present except for the Achilles tendon. She refused further examinations. One year later, her state was unchanged, comparable with that before the neurological episode.

Three out of her four asymptomatic children were investigated. Their total serum homocysteine concentrations were normal and no homocystinuria was detected. In one of them, a slight decrease of MTHFR activity was found (2·48 nmol/h/mg protein). Normal values (4·96 and 5·19 nmol/h/mg protein) were found in the two others.

In this case, enzyme deficiency, relatively well tolerated, was diagnosed in adulthood. Her poor feeding in relation to her psychotic symptoms first led us to suspect a folate deficiency. Because of homocystinuria, a disturbance of folate metabolism was suspected, confirmed by a decrease of MTHFR activity to 25% of the mean control value. Freeman and colleagues[2] described the case of a mildly retarded 15 year old girl with a schizophrenia like behaviour and axonal neuropathy responsive to folic acid. This patient also stopped eating, had low serum folate, later related to an MTHFR deficiency. Botez *et al*, who reported many cases of neurological disorders associated with folic acid deficiency responsive to folate treatment, proposed the hypothesis of a vicious circle between neurological disorders and nutritional folate deficiency.[5] One pathogenic hypothesis for the role of folate in the nervous system is that reduced methionine biosynthesis[3] causes reduced biosynthesis of S-adenosyl-L-methionine, a methyl donor involved in the production of phosphatidylcholine, and consequently in myelin synthesis. A relation between homocystinuria and schizophrenia had been noted. An abnormality in folate metabolism may predispose to schizophrenia.[2]

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Transcallosal intraventricular tumour excision, alcohol abuse, and amnestic syndrome: a case study

Amnesia is defined as short-term and long-term memory impairment occurring in a normal state of consciousness and due to a specific organic factor (DSM-III-R). The most frequent alteration in mental activity after manipulation of midline basal cerebral structures is thought to be a transient amnesic syndrome that usually resolves within several weeks. Our patient had an intraventricular tumour (oligodendroglioma of the lateral ventricles) surgically removed with a transcallosal approach. Neuropsychological testing included evaluations completed just before the operation and one and two months later.

Case history

A 32-year-old white, right-handed male factory worker was admitted to an urban hospital with a headache, blurred vision, and ataxia. An MRI scan revealed an interventricular mass (6 × 5 cm) larger in size on the left than the right. Prior head injuries or seizures were not reported although the patient had a significant history of alcohol abuse. He had completed 13 years of schooling.

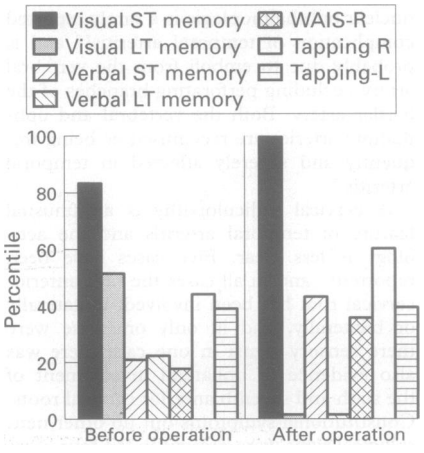
The operation involved a right frontal craniotomy and an interhemispheric, corpus callosal approach. The right frontal lobe was immobilised and a small incision was made in the corpus callosum. The tumour was removed with gentle suction from the right lateral and third ventricles. The left intraventricular tumour was removed with the same technique. The postoperative course was complicated by increased intracranial pressure for which shunts were placed. Over 90% of the tumour was removed and radiation therapy followed surgery.

BEFORE OPERATION

During his first neuropsychological assessment the patient was cooperative and attentive. Initial neuropsychological testing revealed cognitive functioning within the low average range of intelligence with significantly better performance scores than verbal scores. Fine motor skills in both hands were mildly depressed. Verbal memory with a standardised battery was within the 20th percentile (low average) and visual memory skills were at least average (figure). The discrepancy between the verbal and visual skills was consistent with the slightly better visual perceptual skills demonstrated on the intelligence test.

AFTER OPERATION

The patient's behaviour after surgery and transfer to the rehabilitation unit was striking. The patient was not generally orientated to date and was unable to give his correct age. He was found to confabulate and remote memory for significant events was inaccurate. His affect was flat and initiation of conversation limited. The patient was able to learn the location of his room with clues and reminders during his four-week stay but easily became lost in unfamiliar territory. Although the patient's physical



Testing before and after operation showed equal or superior test performance on all measures except long-term (LT) verbal and visual memory. ST = short term.

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