Although small malformation, and plus without clinical which of reports of Down syndrome are made of ventricles in the primary position pronounced. The spontaneous or non-specific spongiform degeneration in the primary area or only loss of neurons. We report a patient who had a slowly progressive hemiparesis, and then mild dysphasia, with only atrophy and loss of neurons on postmortem study.

A right handed woman, born in 1921, had been in excellent health until 1987 when she noticed weakness of the right hand. In the next six months all movements in the left side of the body, especially the left arm and right leg became affected. In 1988 her speech became hesitant. Examination at that time showed slight difficulties in word finding, and a right sided facial weakness and a slurred speech. She had an upper motor neuron type weakness of the right arm and to a lesser extent of the right leg with increased tendon reflexes and an extensor plantar response. There was no atrophy, fasciculation, or sensory abnormality. Laboratory examination of blood, urine, and CSF was unremarkable. Serial CT scans of the brain showed slightly enlarged ventricles and cortical atrophy, especially at the convexity of the left hemisphere. Magnetic resonance imaging confirmed these findings and showed shrinkage of the left side of the pons. Single photon emission computed tomography with the tracer 99m-Tc-HMPAO showed a decrease in blood flow in the left frontal-temporal region; PET showed hypometabolism predominantly in the left perisylvian region.

Neuropsychological assessment indicated a slight dyscalculia, dysgraphia, dysphasia, constructive dyspraxia, and a verbal memory deficit. Serial examinations in the subsequent two years showed a gradual increase of the right sided hemiparesis; because of the difficulties in walking she was admitted to a nursing home in 1989. Neuropsychological re-examination in 1989 showed no abnormalities in appearance and behaviour. She had slight problems with spatial orientation. A striking feature was pronounced slowing of both motor and cognitive functions, with slight difficulties in word finding. Selective and sustained attention were unimpaired. The aphasia screening test (Halstead-Reitan) and the token test indicated motor dysphasia, dysgraphia, and dyscalculia. Performance on the Stroop colour test and word test was below average and showed signs of interference in cognitive functioning and in attention. Memory deficits were found on the Benton visual retention test, a word learning task, and the symbol digit modalities test. The patient's Wechsler adult intelligence scale (WAIS, Dutch version) overall IQ score was 74 (verbal IQ 79, performance IQ 74). She scored 100 on Raven's standard progressive matrices, in accordance with the level estimated from education and occupation. On the whole, the neuropsychological assessment showed few changes compared with the first examination. She died in November 1990, at the age of 69, from pneumonia.

At postmortem, her brain weighed 1290 g. It showed leptomeningeal thickening at the convexity, and slight cortical atrophy in the precentral and postcentral gyri, predominantly on the left side and in both superior temporal gyri. The arteries of the circle of Willis were normal, except for minimal atherosclerosis. After two weeks of fixation (4% phosphate buffered formaldehyde), the education was sliced. Atrophy of the left pyramidal at the level of the medulla oblongata was evident. Serial 6 mm paraffin slides were prepared from multiple blocks from the brain stem, cerebellum, basal ganglia, and cerebral hemispheres, including precentral and postcentral gyri. The sections were

Primary progressive hemiparesis

Focal deficits such as motor aphasia or visual defects were the first sign of Alzheimer's or Pick's disease, but sometimes the initial deficit remains isolated even after many years; postmortem examination of the affected area in the cortex has shown non-specific spongiform degeneration or only loss of neurons. We report a patient who had a slowly progressive hemiparesis, and later mild dysphasia, with only atrophy and loss of neurons on postmortem study.

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Cervical MRI nine years after surgery showing a cyst about 7 mm in diameter in the upper cervical cord.

Microscopic view of the left precentral gyrus showing neurophagia (arrow: neurons undergoing phagocytosis; small arrowhead: satellite cell, large arrowhead: phagocytosis by microglia cell) magnification × 400, Klüver myelin stain.
stained with haematoxylin and eosin, periodic acid Schiff’s, Weil’s myelin, Klüver’s myelin, Nissl-counterstained violet, Bodian, and Congo red. On microscopic examination loss of neurons was most conspicuously present in layer 5 of the left precentral gyrus, most prominently in Betz cells. Neuritophagy (figure) and some proliferation of astrocytes was seen. The underlying white matter showed a decrease in the density of axons and slight gliosis. Other cortical areas, e.g., the hippocampus, amygdala, and Broca’s area showed no evident abnormalities. Although a single senile plaque was present, fibrillary tangles, Hirano bodies, Lewy bodies, amyloid deposition, and senile degeneration were not seen. The basal ganglia showed no abnormalities.

The clinical features of a right-sided hemiparesis, slowly progressive in the course of three years, with only slight disturbances of intellect, in combination with postmortem evidence of non-specific loss of neurons in the motor cortex are indicative of local degenerative disease of the cerebral cortex. Alzheimer’s and Pick’s disease were ruled out on clinical and pathological grounds. In a series of six patients with progressive aphasia PET showed hypometabolism that was more extensive than the lesions visible on CT or MRL,5 as in our patient. The extensive involvement shown by SPECT was comparable with that in previously reported patients with isolated aphasia.2

Our patient with an isolated hemiparesis of cortical origin, later accompanied by dysphasia, may represent a separate variety of a localised loss of cortical neurons, comparable to that with isolated motor cortex.6 The mild disturbances of intellectual functions on neuropsychological examination are largely explained by the hemiparesis and dysphasia.7 Mesulam has also pointed out that such disturbances may result from the influence of the original lesion on the function of other regions of the brain.1

Patients with localised cortical atrophy in normal-pressure hydrocephalus and without dementia have been described in several groups—namely, visuoperceptual disorder;8 generalised apraxia;9 perceptuomotor deficits, often combined with hemiparesis10 and behavioural changes11—and progressive aphasia with unilateral extrapyramidal signs.12 The underlying pathological process in these localised cortical diseases is incompletely understood and not uniform. One patient with a slowly progressive aphasia showed (on biopsy) only non-specific and slight degenerative changes.1 One patient with a neuropsychiatric syndrome showed spongiform changes and mild gliosis in superficial layers of the prefrontal cortex.7 Three other patients with aphasia showed atrophy, spongiform changes, and astrocystosis, mainly in one or two areas of the cortex, without Pick or Lewy-type inclusion bodies.1

It seems that primary progressive hemiparesis is comparable with primary aphasia or one type of degeneration syndrome of the cerebral cortex. This variety of disorders follows a more protracted course than the more generalised dementias, and has no morphological characteristics to link it with Alzheimer’s or Pick’s disease.

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4 Poock K, Luzzati C. Slowly progressive aphasia in three patients, the problem of accompanying neuropsychological deficit. Brain 1988;111:511-68.

**Vertebral artery dissection mimicking migraine**

We agree with Giroud et al that internal carotid artery dissection is a major cause of cerebral infarction in those under 50 years of age.11 We also suggest that spontaneous dissection of the vertebral artery is an important cause of ischaemic stroke in this age group, increasingly recognised with the advent of effective, non-invasive methods of diagnosis such as colour duplex ultrasound and MRL.2,3 We report a patient presenting with symptoms that would not necessarily have justified investigation by conventional angiography but who was found, by non-invasive means, to have had a vertebral artery dissection.

A 35 year old woman, previously well, suddenly experienced distorted vision while doing the housework. She described wavy, flickering lines in both eyes and hazy vision that persisted for about an hour. As the flickering faded she noticed more that she could only see the right half of her husband’s face. This field defect persisted for about 30 minutes before her vision returned to normal. The next day she was aware of mild, diffuse headache and a stiff neck, which gradually improved over the next two weeks. There were no other associated symptoms and no history of trauma to the neck, although she had been playing in a competitive netball team earlier in the day. In the past, she had had hypertension while pregnant but there was no history of prior cerebrovascular events or of migraine. She did not smoke.

Examination two weeks after the onset of symptoms was normal other than slightly hyperextensible little fingers on both hands. Cardiovascular examination was normal; blood pressure was 130/80 mm Hg and no bruits were audible in the neck. Skin and other joints were normal and there were no focal neurological signs.

It was considered that her symptoms could have been caused by a vertebralbasilar transient ischaemic episode. Migraine was also a possibility. Blood tests, including blood count, glucose, cholesterol, lupus anticoagulant, anticardiolipin antibody concentrations, and syphilis serology were normal. Magnetic resonance imaging of the neck with axial, spin echo, T1 weighted images showed bright intramural thrombus with a very small residual lumen in the right vertebral artery (figure A). The abnormality was localised to a 2 cm portion of extracranial vertebral artery at the level of the C2 vertebra. Magnetic resonance angiography (MRA) showed disturbed flow within the vertebral artery at this level. The changes were diagnostic of vertebral artery dissection.

The patient was advised to continue aspirin for six months and has had no further symptoms. Repeat MRI, four months after the episode, showed no evidence of a persisting lesion (figure B).

Extracranial vertebral artery dissection, with occlusion or distal embolisation, is an important cause of transient ischaemic attack and stroke in patients under 50 years of age. Often there is no history of trauma to the neck or, when a history of trauma is present, it may be trivial. Epidemiological studies may have underestimated the true incidence of both carotid and vertebral artery dissection because of the requirement in the past for invasive angiography, with its attendant risk, to make the diagnosis. With the introduction of routinely available ultrasound and MR techniques, allowing non-invasive diagnosis, there is little doubt that more cases will come to light. It is interesting to note that the incidence of carotid dissection seems to be similar to that of aneurysmal subarachnoid haemorrhage.

This woman’s symptoms were “trivial” and the episode could easily have been diagnosed as an attack of atypical migraine with no further investigation performed.
Primary progressive hemiparesis.

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