the sternum and spinal column are common and include scoliosis and spina bifida occulta. 2  Cervical neuralaxial malformations have been reported only rarely, 3-5 and it has been questioned whether this association is causal or a chance occurrence. 6

A 21 year old woman with Noonan’s syndrome presented with three days of anterior tongue numbness, then spreading right facial weakness and numbness. The family history was of a maternal cousin with a similar phenotype and a heart condition (unavailable for examination). Examination disclosed a short girl (148 cm) with mild cognitive impairment and the characteristics of Noonan’s syndrome. Bilateral papillitis, pronounced down beat nystagmus in the primary position accentuated by lateral gaze, diminished right corneal and facial sensation, right facial weakness, and impaired hearing on the right were present. Diminished sensation of the anterior two thirds of the tongue bilaterally and the right palate was also found. The remainder of the neurological examination was normal.

A metrizamide myelogram with cervical and cranial CT showed a Chiari type I malformation with extension of the cerebellar tonsils to the level of C2 and inferior displacement of the medulla. The third and lateral ventricles were very dilated, consistent with hydrocephalus. Neither syringomyelia nor C1 spina bifida occulta were detectable on these films. The patient underwent a decompressive suboccipital craniotomy and C1/2 laminectomy because of the onset of early morning headaches. She made a good recovery, albeit with persistence of down beat nystagmus. A follow up cervical CT myelogram, and more recently, MRI (figure) have shown a small cyst in the upper cervical region, which has enlarged over seven years but without clinical change.

Our experience adds to the single case reports of Noonan’s syndrome with Chiari I malformation plus syringomyelia, 7 Chiari I malformation, 8 and syringomyelia alone. 9 Although small cysts are not usually considered syringomyelic, they may be of a similar nature. 10 These abnormalities are probably part of the range of malformations associated with Noonan’s syndrome. Given that such dysraphic abnormalities may be asymptomatic, their occurrence in Noonan’s syndrome may be underestimated.


Primary progressive hemiparesis

Focal deficits such as motor aphasia or visual defects can be 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. 2 We report a patient who had a slowly progressive hemiparesis, and later 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 of the right 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 intervals of the skull 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, 6 showed a decrease in blood flow in the left fronto-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 brain 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...
stained with haematoxylin and eosin, periodic acid Schiff’s, Weil’s myelin, Klüver’s myelin, Nissl-staining and Bodian, and Congo red. On microscopic examination loss of neurons was most conspicuous present in layer 5 of the left precentral gyrus, most prominent in Betz cells. Neuritis spongiosa (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, for example 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 deposits, and senile changes 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 MRI, as in our patient. The extensive involvement shown by SPECT was comparable with that in previously reported patients with isolated aphasia.

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 our patient. The isolated motor 


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. 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 MRI. 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 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 sport 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 vertebrobasilar transient ischaemic episode. Migraine was also a possibility. Blood tests, including blood count, glucose, cholesterol, lupus anticoagulant, antiphospholipid antibody, 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 neursymal 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.