

LETTERS TO THE EDITOR

Clinicoradiographic evidence for oculomotor fascicular anatomy

The anatomy of the third nerve fascicle as it courses through the midbrain is a topic of debate. Several cases of "atypical" third nerve palsies from lesions of the mesencephalon have been reported and from this material topographical models have been proposed. We describe a pupil and inferior rectus sparing unilateral third nerve palsy with detailed radiographic correlation to support one of these models.

A 34 year old right handed African-American woman with a history of systemic lupus erythematosus, peripheral vascular disease, HIV seropositivity, hypertension, cigarette smoking, and "crack" cocaine misuse was seen as an outpatient with a one week history of intermittent diplopia, dizziness, and right sided ptosis. Asymptomatic at the time, the only abnormalities on brain MRI were several small white matter periventricular lucencies consistent with small vessel ischaemic disease. She awoke four days later dizzy, unable to adduct or elevate her right eye, and with severe right sided ptosis. Pupils were equal and reactive, both directly and consensually, to light and accommodation. There was a right hemiataxia of gait and finger-nose-finger, which resolved within 12 hours. The contralateral eye was unaffected. Fundi were normal, visual fields were full, and visual acuity was 20/20 in the left eye and 20/30 in the right eye.

Computed tomography with and without contrast and including coronal sections through the cavernous sinus were within normal limits. Magnetic resonance imaging showed increased signal intensity in the right paramedian midbrain, anterior to the periaqueductal grey matter on the long repetition time images (fig 1A). Coronal MRI showed increased signal intensity just inferior to the right red nucleus (fig 1B). The location of the lesion corresponds to the superior aspect of the decussation of the superior cerebellar peduncle (fig 2).

Oculomotor palsies arising from mesencephalic pathology often involve the third nerve nuclear complex and cause pupillary dysfunction or bilateral oculomotor palsies due to the crossed innervation of the superior rectus and bilateral innervation of the levator palpebrae superioris. Midbrain lacunae that involve the third nerve fascicle and cause unilateral partial third nerve palsies are uncommon. In a recent review of 1015 patients presenting with first stroke, 22 had an isolated midbrain infarct (2.3%) and only one patient had evidence for partial fascicular involvement (0.1%).¹ Nevertheless, several cases of radiographically confirmed midbrain lesions causing unilateral partial third nerve palsies have been reported.² These cases have given rise to debate regarding the anatomy of the third nerve fascicle as it courses through the midbrain. Castro *et al*³ originally proposed a two dimensional model in which the fibres

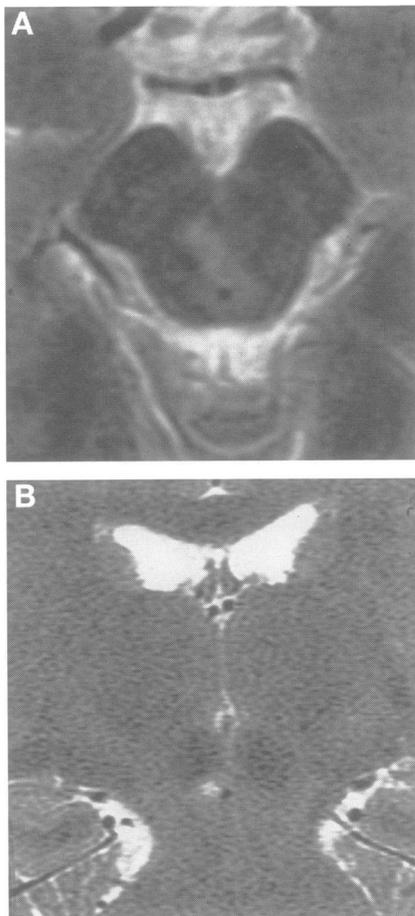


Figure 1 Axial (A) and coronal (B) T2 weighted MRI, 4500/102 (repetition time/echo time), through the midbrain showing increased signal intensity in the right paramedian midbrain, anterior to the periaqueductal grey and inferior to the red nucleus

for the pupil, inferior rectus, levator palpebrae superioris, medial rectus, superior rectus and inferior oblique muscles lie from medial to lateral. This model was later revised,⁴ and the levator palpebrae superioris fibres were moved to a more lateral position between the superior rectus and medial rectus to account for reported cases of isolated superior and inferior branch paresis from fascicular lacunae.⁵

The rostrocaudal organisation of the fascicle has been less clearly worked out, although Ksiazek *et al*² have recently proposed a three dimensional model based on clinical and experimental evidence. Their schema includes the medial to lateral organisation described above as well as a rostrocaudal dimension in which the pupil is most superior followed by fibres for the inferior rectus, inferior oblique, medial rectus, superior rectus, and levator palpebrae superioris (fig 2).

The case we present provides excellent clinicoradiographic confirmation for this model. Symptomatically, the patient presented with a transient hemiataxia secondary to involvement of the ipsilateral brachium conjunctivum, a structure situated inferior to the third nerve fascicle (fig 2). Axial and coronal MRI confirm the lesion's position just inferior to the red nucleus in a parasagittal plane (fig 1A and B). In accordance with the model of Ksiazek *et al*,² such a lesion would impinge

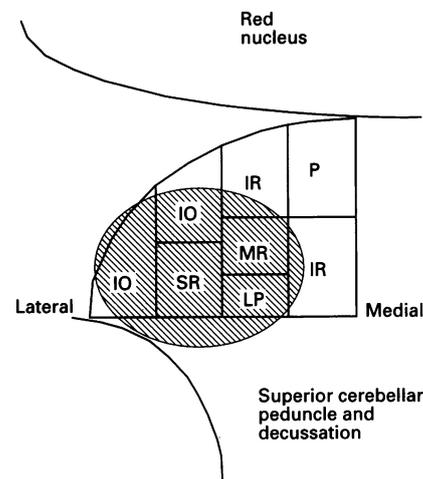


Figure 2 Diagram showing the intrafascicular anatomy of the third nerve in the coronal plane. Shading represents the site of the lesion seen on MRI. IO = inferior oblique, IR = inferior rectus; SR = superior rectus; MF = medial rectus; P = pupil; LP = levator palpebrae.

on the lateral, caudal aspect of the oculomotor fascicle, which would spare the pupil and the inferior rectus. Ultimate acceptance of this model awaits further clinicopathological confirmation.

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Noonan's syndrome with hydrocephalus, hindbrain herniation, and upper cervical intracord cyst

Noonan's syndrome is an inherited disorder characterised by mental retardation, short stature, hypertelorism, ptosis, low set ears, small mandible, a short neck, and congenital pulmonary stenosis. Further abnormalities may be elbow valgus, hepatosplenomegaly, coagulation disturbances, hypopituitarism, undescended testis, and delayed puberty. Deformities of



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

the sternum and spinal column are common and include scoliosis and spina bifida occulta.^{1,2} Cervical neuraxial malformations have been reported only rarely,³⁻⁵ and it has been questioned whether this association is causal or a chance occurrence.³

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 proptosis, 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 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,³ Chiari I malformation,⁵ and syringomyelia alone.⁴ Although small cysts are not usually considered syringomyelic, they may be of a similar nature.⁶ 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.

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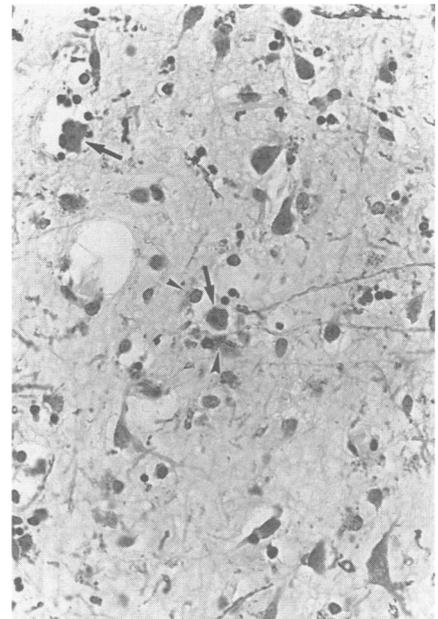
Primary progressive hemiparesis

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.^{1,2} 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 six months 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 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



Microscopic view of the left precentral gyrus showing neuronophagia (arrows: neurons undergoing phagocytosis; small arrowhead: satellite cell, large arrowhead: phagocytosis by microglia cell) magnification $\times 400$, Klüver myelin stain).

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 pyramid at the level of the medulla oblongata was evident. Serial 6 μ m 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