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 palsy 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 sinuses 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 region of the right third nerve fascicle at the superior aspect of the decusation 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 palsy 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%). Nevertheless, several cases of radiographically confirmed midbrain lesions causing unilateral partial third nerve palsies have been reported. These cases have given rise to debate regarding the anatomy of the third nerve fascicle as it courses through the midbrain. Castro et al originally proposed a two dimensional model in which the fibres 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, 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. The rostrocaudal organisation of the fascicle has been less clearly worked out, although Kaisake et al 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.

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 Kaisake et al, such a lesion would impinge 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 testes, and delayed puberty. Deformities of...
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