

IFN in MS patients will have to be determined. As any beneficial effect of the induced cortisol would be expected to be short-lived, it seems unlikely that it would have much effect in a protocol in which IFN doses are given infrequently [4], but it might be of significance when IFN is given daily [3, 11]. Similarly, it would be difficult to attribute any persistent beneficial effect of IFN to cortisol induction [4]. Monitoring of cortisol levels in MS patients receiving IFN should clarify these issues.

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Acquired Ocular Motor Apraxia Due to Bilateral Frontoparietal Infarcts

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A case of acquired ocular motor apraxia in a 78-year-old man is reported. Disordered saccades and pursuit movements in the horizontal and vertical planes were recorded electro-oculographically. Magnetic resonance imaging showed bilateral border-zone infarcts, involving the frontal lobes and the superior part of the inferior parietal lobules.

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Acquired "ocular motor apraxia" is characterized by great difficulty in initiating saccades to command and to visual targets, usually in all directions [1]. This rare syndrome results from bilateral frontoparietal lesions and must be distinguished from various, and at times severe, saccade abnormalities present in several degenerative diseases involving the basal ganglia. It is also different from the congenital form of ocular motor apraxia, in which saccade disturbance is restricted to the horizontal plane and usually decreases with age [2]. We report a case of acquired ocular motor apraxia which may add to our knowledge of the control of purposive saccades and pursuit movements by the cerebral cortex.

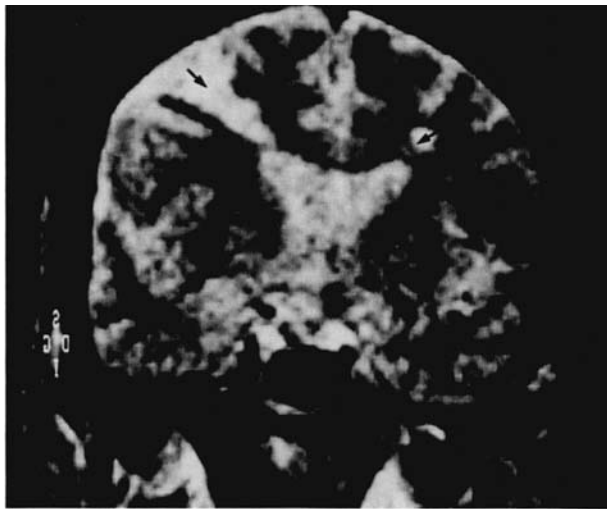
Case Report

A 78-year-old righthanded man was admitted for right hand weakness that began suddenly a few hours before. He had a history of peripheral arterial insufficiency in the legs, first degree atrioventricular block, and untreated hypertension. The day after admission, he had paresis of both arms, predominantly distal and more severe on the right side, and complex oculomotor deficits (see text following). Pupils and visual fields were normal. There was no visual extinction and no obvious visual neglect: the patient saw and easily named objects placed in any part of his peripheral visual field

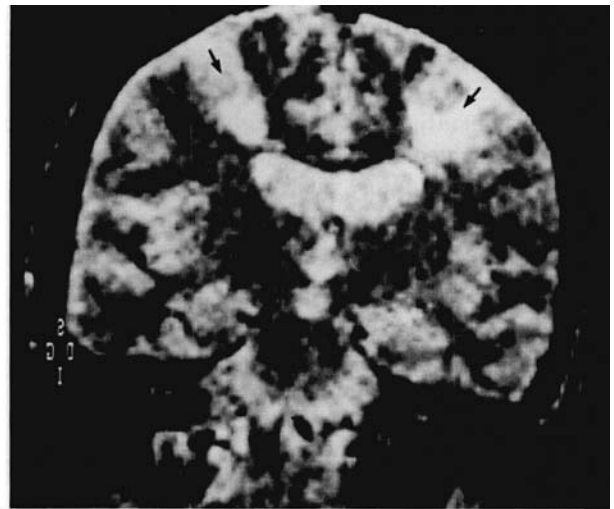
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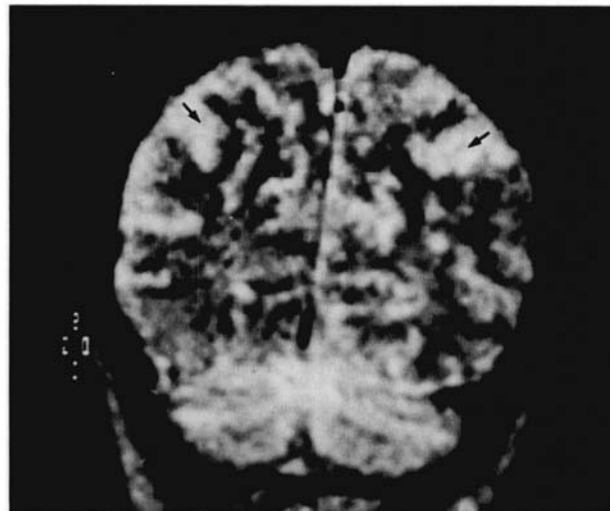
A



B



C



D

Fig 1. T2-weighted magnetic resonance imaging. Coronal sections through the posterior part of the frontal lobes (A), the anterior part of the parietal lobes (B), the posterior part of the parietal lobes (C), and near the parietooccipital junction (D). Lesions (arrows) involved the junctional territories of the middle and anterior cerebral arteries.

without glancing at them. General physical examination was normal except for the presence of atrial fibrillation. Doppler studies showed mainly an occlusion of the right internal carotid artery with reversed flow through the right ophthalmic artery and tight stenosis of the left internal carotid artery at its origin, with left ophthalmic artery flow in the normal direction.

Three weeks after his admission, a T1-weighted magnetic resonance image (MRI) was normal, but a T2-weighted MRI showed bilateral watershed infarcts in the junctional territories of the middle and anterior cerebral arteries. The lesions involved the posterior part of the left middle frontal gyrus (Fig 1A), which includes the frontal eye fields (FEF) [3]. On the right side, only the white matter underlying the

right FEF appeared to be involved. The lesions extended posteriorly in the paramedian region of both hemispheres (Fig 1B), involving the superior part of the inferior parietal lobule (IPL) and the lateral bank of the adjacent intraparietal sulcus bilaterally (Fig 1C). They ended near the parietooccipital junction (Fig 1D).

Eye Movements

Eye movements were recorded in darkness with direct-current electrooculography. Calibration was carried out using those rare visually guided saccades that were performed with the head immobilized. Saccades were tested to command, then to visual targets (located 25 degrees away from the center), first with the head immobilized, then with the head free. Foveal smooth pursuit was tested by asking the patient to follow movement of the target light as it was driven in triangular waveforms at several constant velocities (starting at 5 degrees/second). Optokinetic nystagmus (OKN) was stimulated with alternate black and white stripes moving on a screen at several constant velocities (starting at 10 degrees/

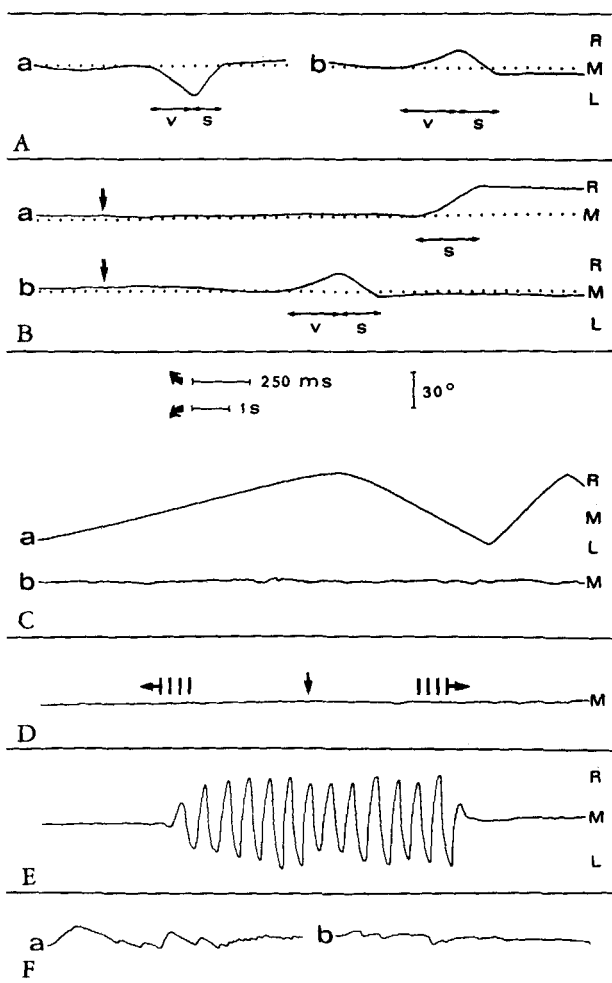


Fig 2. Lateral eye movements. (A) Saccades to command, rightward (a) and leftward (b), preceded by a head movement with a vestibular ocular movement in the opposite direction. (B) Visually guided saccades: rightward, without a head movement (a); leftward, preceded by a head movement (b) (vertical arrows indicate the onset of the visual target). (C) Foveal smooth pursuit: stimulation (a) and eye movement (b); no movement. (D) Optokinetic nystagmus: horizontal arrows indicate the stimulation direction and the vertical arrow the reversal of stimulation; no movement. (E) Oculoccephalic movement: present. (F) Vestibular nystagmus, induced by caloric stimulation of the left ear (a) and of the right ear (b). L = left; M = midline; R = right; s = saccade; v = vestibular ocular reflex.

second) and covering 80 degrees of the visual field. Oculocephalic movements were tested by passive movements of the patient's head performed by the examiner. The head movement was not recorded. All these eye movements were stimulated successively in the horizontal and vertical planes. Horizontal vestibular nystagmus was elicited by successive caloric stimulation of each ear with several milliliters of ice water. Lastly, convergence was stimulated by slowly moving a small object toward the patient's nose and then toward each eye.

The first eye movement recording was carried out 1 week after admission. The patient was alert and cooperative throughout the eye movement examination. At rest, he

looked straight ahead (without strabismus or diplopia), and his eyes were permanently immobile without slow deviation or spontaneous saccades (even in darkness). Blinks were rare and usually absent during eye movements or attempted eye movements.

All eye movement disorders were similar in the lateral and vertical planes. Consequently, only the lateral eye movements will be described in detail (Fig 2). With the head immobilized, rightward and leftward saccades to command were absent; the eyes remained on the midline. With the head free, disorders were similar rightward and leftward. There was first a head movement in the appropriate direction, inducing a slow vestibular eye movement in the opposite direction (Fig 2A, ab). Then a saccade was performed in the appropriate direction, allowing the eyes to return to the midline. This final eye movement usually consisted of one large saccade, although several small hypometric saccades were occasionally observed. When there was only one saccade, velocity was measured and was about 50% less than that of saccades with identical amplitudes performed by normal subjects in our laboratory.

Visually guided saccades with the head immobilized were obtained only five times (2 rightward and 3 leftward) in 30 attempts in each direction. One of the two rightward visually guided saccades is shown in Figure 2B, a. Latency of these saccades was long, between 1,100 and 1,700 msec (i.e., five to eight times the norm). Their velocity was reduced by the same amount as that of saccades to command. With the head free, the same sequence of movements as that described in saccades to command was observed either rightward or leftward (Fig 2B, b).

Foveal smooth pursuit and OKN were absent in all directions, even at the lowest stimulation velocities (Fig 2C,D). Oculocephalic movements were present, laterally (Fig 2E) and vertically. Caloric stimulation of the left ear induced a right-beating vestibular nystagmus, and caloric stimulation of the right ear induced a smaller left-beating vestibular nystagmus (Fig 2F, ab). Quick phases of vestibular nystagmus were slow. Convergence was absent for both eyes.

Course

Six weeks after the onset, paralysis of the right hand persisted. The motor deficit of the left upper limb had completely recovered. Eye movements were recorded again. All saccades (to command and to visual targets, in the horizontal as well as in the vertical plane) were normal, even with the head immobilized. Latency (right = 262 ± 55 ms; left = 256 ± 62 ms) and velocity of lateral visually guided saccades were normal. Pursuit movements, however, were still absent in all directions. Foveal pursuit was saccadic, without any smooth movement, and OKN could not be elicited, the eyes remaining immobile during stimulation. Convergence was still absent. The patient refused to undergo MRI a second time.

Discussion

This severe disturbance of the initiation of purposive saccades in all directions, without actual paralysis, may be considered ocular motor apraxia in its acquired form [1]. At least two parallel excitatory suprareticular

tracts could be involved in saccade initiation [4, 5]. They originate from the FEF and the superior colliculus, respectively, and project directly onto the premotor reticular formations in the brainstem. Such an organization is supported by lesion experiments in the monkey [6]: bilateral damage to the FEF alone, or to the superior colliculus alone, induced only temporary disturbance of saccades, whereas bilateral damage to both structures resulted in the severe and long-lasting impairment of saccades. It has recently been suggested that the superior colliculus could in fact be a relay in an excitatory pathway arising from the posterior portion of the lateral bank of the intraparietal sulcus (located in the IPL) [5, 7].

It is likely that the actions of the frontal and parietal areas overlap somewhat in the normal subject because only bilateral frontoparietal lesions severely impair saccade initiation in the hemispheric cerebral pathology. In 3 clinicopathological patients with such lesions, saccades and pursuit were described as being absent. In 2 of them, lesions were metastatic or vascular and affected extensive areas of both the frontal and parietal lobes bilaterally [8, 9]. In the third case, the lesions were much smaller, ischemic in origin, and located, as in the case of our patient, at the junction of the territories of the middle and anterior cerebral arteries in both hemispheres [10]. They involved the FEF and the superior part of the IPL. Although the abnormal image visible on MRI in the present case involved only the subcortical white matter in the right frontal lobe, this lesion could interrupt the efferent connections of the right FEF. These data in humans, as well as recent experiments with analogous lesions in the monkey [11], confirm that the control of purposive saccades is mainly distributed across the FEF and the IPL in the cerebral cortex. In the present case, the facilitation of saccades by the combined head and vestibular ocular movements could originate from a simple vestibular mechanism or from the action of undamaged cortical areas (such as the supplementary motor area). This case also confirms that cortical lesions can influence saccade velocity [4].

Lastly, foveal smooth pursuit and full-field pursuit were absent in all directions. Bilateral damage to the IPL (visible on MRI) could account for this abnormality because, as has been recently reported, such lesions may result in severe impairment of pursuit movements [7]. Unlike saccades, pursuit movements showed no improvement after 6 weeks. This suggests not only that no significant recovery occurred in the damaged parietal areas but also that there were no adaptive mechanisms from other oculomotor areas, at least within such a period of time.

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Hearing Loss in Myotonic Dystrophy

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Seventeen of 25 patients with myotonic dystrophy had moderate to severe hearing loss, usually sensorineural, that was identified by routine audiometric screening and was treatable in some patients. Further testing failed to reveal a single pathophysiological process.

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Myotonic dystrophy (dystrophia myotonica, DM) involves multiple systems. We noticed a surprisingly

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