Homonymous hemianopia in multiple sclerosis

With report of bilateral case

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Optic neuritis is common in multiple sclerosis, occurring in 22–83 per cent of patients (Carter, Sciarra, and Merritt, 1950; Chamlin, 1953; Leinfelder, 1950; Marshall, 1950; Savitsky and Rangell, 1950a; Zeller, 1967). Symptomatic chiasmal and retrochiasmal involvement is infrequent. Complete homonymous hemianopias (Bielschowsky, 1933; Boldt, Haerer, Tourtellotte, Henderson, and DeJong, 1963; Chamlin and Davidoff, 1954; Kahana, Leibowitz, and Alter, 1971; Luhan, 1944; Vedel-Jensen, 1959), quadrantic defects (Bjerrum, 1890; Boldt and others, 1963; Chamlin and Davidoff, 1954; François and Verriest, 1957; Rönne, 1912, 1915; Traquair, 1942), homonymous scotomas (Boldt and others, 1963; Chamlin and Davidoff, 1954; François and Verriest, 1957; Otradovec, 1965; Rönne, 1912, 1915; Traquair, 1942; Vedel-Jensen, 1959), and other unspecified types of homonymous defects (Croll, 1965; Kurtzke, Beebe, Nagler, Auth, Kurland, and Nefzger, 1968; Leinfelder, 1950; Müller, 1949) have been found in up to 3.5 per cent of cases (Boldt and others, 1963; Croll, 1965; Kahana and others, 1971; Kurtzke and others, 1968; Leinfelder, 1950; Müller, 1949; Savitsky and Rangell, 1950b). One of Bielschowsky's cases had bilateral involvement at different times (Bielschowsky, 1933).

We reviewed the records of 344 patients all of whom had been discharged from the Columbia-Presbyterian Medical Center from 1962 to 1970 with a diagnosis of multiple sclerosis. In 300 of these, the diagnosis was considered to be valid on the basis of characteristic clinical presentation and course. Among these patients there were 40 with observed optic neuritis, 83 with history suggestive of optic neuritis, and 51 without such a history but with optic disc pallor. In the entire group of 300 patients, only four had homonymous visual field defects, an incidence of 1.3 per cent.

A patient was recently seen with simultaneous bilateral homonymous hemianopic defects. This unique case is reported in detail and the infrequent

clinical detection of retrochiasmal lesions in multiple sclerosis is discussed.

Case report

A 19-year-old woman was admitted to the New York Neurological Institute in September, 1972. One month before admission, she had awoken with blurred vision; she had noted that her speech was jumbled and that her balance was poor as well as tingling and clumsiness of her hands. The symptoms became more severe and she developed urinary frequency. All functions had returned to normal within 2 weeks except for her vision. One week before admission, however, all symptoms recurred and her vision worsened.

On admission, she was euphoric with an unsteady, ataxic gait. Visual acuity was 20/20 in each eye. On confrontation she had an upper left homonymous visual field defect. Pupillary reactions were slightly sluggish without relative afferent defect. Optic discs were considered to be normal. There was bilateral horizontal gaze-evoked nystagmus, a left lateral rectus paresis, and mild right central facial weakness. Strength was markedly decreased in the right arm and moderately decreased in the right leg. She had increased muscle tone and deep tendon reflexes on the right; there were bilateral extensor plantar responses, and the abdominal reflexes were absent. There was decreased sensation to pin, position, and vibratory sense in the right arm and leg and moderately severe right-sided dysmetria and dysidiadochokinesia.

Laboratory results included normal complete blood count, serology, antinuclear antibody, lupus erythematosus prep, and serum protein electrophoresis. Erythrocyte sedimentation rate was 30 mm/hr. The cerebrospinal fluid contained 29 mononuclear cells and no red blood cells. The protein was 36 mg per cent, 16 per cent of which was gamma globulin (normal, less than 14 per cent) sugar was 70 mg per cent. Fluorescent treponemal antibody absorbed test for syphilis and routine cultures were negative. Skull x rays, mercury brain scan, and electroencephalogram were normal. Electroencephalogram showed non-specific mild slowing bilaterally.

Three days after admission, visual acuity was 20/25 in each eye, and qualitative confrontation testing revealed a congruous right homonymous hemianopic defect, as well as the previously noted upper left homonymous defect: these findings were confirmed by tangent screen examination (Figure).

FIGURE Results of tangent screen examination

Treatment was started with 80 units intramuscular adrenocorticotrophic hormone (ACTH) per day. The patient showed a slight improvement in strength but developed paraesthesias and hypaesthesia bilaterally below the mid-thoracic region. ACTH was increased to 160 units per day. Within 2 weeks motor and sensory functions were markedly improved; visual fields were unchanged.

Steroids were discontinued and she was discharged 6 weeks after admission. Two weeks later when seen in the clinic, her only complaint was mild visual blurring. Visual acuity was 20/20: the visual fields were unchanged. There were no other abnormal neurological signs.

In the subsequent months there were episodes of pyramidal, sensory, and cerebellar symptoms. The mild visual blurring persisted. In June 1973, 10 months after the onset of the illness, the visual acuity was 20/20 in each eye with visual fields unchanged except that the left upper homonymous defect appeared less dense. Pupillary responses and disc appearance remained normal. Horizontal and vertical gaze-evoked nystagmus persisted.

Discussion

As early as 1890, Uhthoff remarked on the frequency of pathological lesions in the posterior visual pathways in multiple sclerosis in the absence of clinical manifestation. This has been found to be generally true of cerebral plaques (Kahana and others, 1971) which commonly involve the paraventricular white matter traversed by the optic radiations (Zimmerman and Netsky, 1950). Savitsky and Rangell (1950a)

found asymptomatic lesions in the optic radiations in 23 of 50 necropsies, and no homonymous visual field defects in 415 personally observed patients (Savitsky and Rangell, 1950b). Optic nerve involvement is, in contrast, frequently symptomatic, although recent data have documented the extent to which asymptomatic progressive demyelination and axonal loss also occur in the optic nerve (Kahana, Leibowitz, Fishback, and Alter, 1973). This has been suggested by pathological studies (Gartner, 1953; Lumsden, 1970) as well as analysis (Feinsod, Abramsky, and Auerbach, 1973; Halliday, McDonald, and Mushin, 1973; Richey, Kooi, and Tourtellotte, 1971) of visually evoked responses (VER) and is confirmed in this series (51/300 cases).

What might explain the greater disparity between pathological and clinical manifestation in the posterior visual pathways? Clinical and experimental evidence indicate that small lesions of the occipital radiations and cortex may exist without demonstrable visual field defect (Walsh and Hoyt, 1969). Anatomically fibres in the optic radiations are relatively widespread. A small plaque in the radiations would involve fewer visual fibres than in the optic tract or in the optic nerve; the compact arrangement of fibres within the optic nerve sheaths must increase vulnerability to swelling of a neighbouring plaque. Physiological differences between retrogeniculate and pregeniculate pathways might make defects from

small lesions in the radiations less detectable. Such factors might include differences between receptive fields at various levels of the visual pathways, as well as feedback inhibition and integration at the geniculate level (Hubel and Wiesel, 1961; Weiskrantz and Cowey, 1967; Weiskrantz, 1972). Further speculation might include greater redundancy and plasticity in the posterior pathways as well as the possibility that demyelination without detectable axonal conduction deficit (Gledhill, Harrison, and McDonald, 1973) might conceivably occur there more readily.

Optic atrophy in multiple sclerosis, whether asymptomatic or following optic neuritis, may be unassociated with demonstrable scotoma. This is attributable to a diffuse loss of axons. Similarly a diffuse loss of retrochiasmal axonal function may occur without visual field defect by ordinary perimetry, particularly if bilateral, although it might be detected by qualitative confrontation testing (Frisén, 1973) if asymmetric or unilateral and corroborated by VER analysis.

One can also speculate as to factors which might make demonstrable retrochiasmal visual field defects asymptomatic. Fibres in the radiations closest to the ventricles have been found to represent the most peripheral and perhaps unioocular portion of the visual field (Spalding, 1952); plaques in this area may give rise to minimal, easily ignored symptoms. Many of the retrochiasmal defects reported have been congruous homonymous scotomas, presumably of occipital origin; such defects, without impairing visual acuity or the periphery of vision, may also be asymptomatic. A small plaque may not affect corresponding fibres from both eyes and result in a deficit masked by the intact visual field of the other eye.

If retrochiasmal involvement of the visual pathways occurs late in the disease, it may be, as suggested

by Chamlin and Davidoff (1954), that 'other manifestations of the disease may be severe and numerous enough to cloud a mere hemianopic defect and that perhaps such field defects therefore go unnoticed.'

The simultaneous bilateral involvement of retrochiasmal visual pathways in our case and the frequency of homonymous scotomas among such defects previously reported raise the possibility that occasionally some patients with 'bilateral optic neuritis' may have involvement not of the optic nerves but of the posterior pathways. With pregeniculate involvement, such patients may have afferent pupillary defects and develop optic atrophy.

Summary

A patient with multiple sclerosis and bilateral retrochiasmal visual field defects is reported. Homonymous field defects are rare in multiple sclerosis despite the frequency of pathological involvement of the retrochiasmal visual pathways. A higher incidence might be found with a higher index of suspicion and careful visual field testing with qualitative confrontation technique.

Other reasons for the infrequent clinical detection of retrochiasmal lesions are considered. Such lesions may exist without demonstrable defect. This may be explained by anatomical factors, for example, fibre arrangement, or physiological factors, such as, geniculate or retrogeniculate integration. Lesions producing demonstrable defects may be asymptomatic because they:

- affect only the peripheral field,
- are small scotomas that do not impair visual acuity,
- affect only one eye, or
- occur late in the course of disease when masked by optic nerve involvement.

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