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Eye Changes in Disseminated Sclerosis

A. D. Chalmers

Abstract

What is probably the first written account of the clinical features of disseminated sclerosis is contained in the diary of Augustus D'Este, the illegitimate son of Prince Augustus Frederick, and grandson of George III. The first manifestations of the disease in his case, as m some 30% of all such patients, were referable to the eyes. In 1822, we read, he journeyed to visit a much-loved relative; On my arrival I found him dead. I attended his funeral-there being many persons present I struggled violently not to weep, I was however unable to prevent myself so doing; shortly after I was obliged to have my letters read to me and their answers written for me, as my eyes were so attacked that when fixed upon minute objects, indistinctness of vision was the consequence. Until I attempted to read or cut my pen, I was not aware of my eyes being in the least attacked. Soon after I went to Ireland, and without anything having been done to my eyes they completely recovered their strength, and distinctness of vision."3 It is interesting to note the temporal relationship of the onset of symptoms in this case, to a period of emotional stress, a relationship which has been much discussed in connection with disseminated sclerosis, but which has little statistical evidence in its support.

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EYE CHANGES IN DISSEMINATED SCLEROSIS

By A. D. CHALMERS

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What is probably the first written account of the clinical features of disseminated sclerosis is contained in the diary of Augustus D'Esté, the illegitimate son of Prince Augustus Frederick, and grandson of George III. The first manifestations of the disease in his case, as in some 30% of all such patients, were referable to the eyes. In 1822, we read, he journeyed to visit a much-loved relative; "On my arrival I found him dead. I attended his funeral—there being many persons present I struggled violently not to weep, I was however unable to prevent myself so doing; shortly after I was obliged to have my letters read to me and their answers written for me, as my eyes were so attacked that when fixed upon minute objects, indistinctness of vision was the consequence. Until I attempted to read or cut my pen, I was not aware of my eyes being in the least attacked. Soon after I went to Ireland, and without anything having been done to my eyes they completely recovered their strength, and distinctness of vision."[3] It is interesting to note the temporal relationship of the onset of symptoms in this case, to a period of emotional stress, a relationship which has been much discussed in connection with disseminated sclerosis, but which has little statistical evidence in its support.

As indicated above, there is general agreement on the approximate incidence of eye-disease early in the course of disseminated sclerosis. On the overall incidence of eye changes there is less agreement, but it does appear that a high proportion of sufferers from disseminated sclerosis do, at some time during its course, exhibit signs or symptoms of eye-disease. For example the report of one series of investigations puts the incidence of ocular signs in disseminated sclerosis as high as 83%.

The three main mechanisms by which the disease process affects the eyes are by the production of retrobulbar neuritis, nystagmus and oculomotor disorders. Other effects on the eyes, less useful diagnostically, are alterations in pupillary characteristics, and changes in the appearance of the retinal veins.

RETROBULBAR NEURITIS

Just as the cervical spinal cord appears to be particularly vulnerable to the disease in its early stages, so the optic nerve suffers early in a significant number of cases. It is difficult, even in any one series, to establish a definite figure for the incidence of retrobulbar neuritis as an early symptom, because in many cases a number of years elapse between the first attack and the appearance of any other manifestations of the underlying disease e.g. Adams found a latent period of over 15 years in 8% of 389 cases. However, by careful and specific history-taking, several authors have estimated that an attack of

retrobulbar neuritis heralds the disease in 13-23% of cases. By similar means the overall incidence of retrobulbar neuritis in the whole course of the disease was shown to be only slightly higher (up to 30%). It also would appear that all the workers in this field are agreed that this particular manifestation always occurs early in the natural history if it appears at all.

In view of the numerous theories which have tried to connect the onset of disseminated sclerosis with local stress of various kinds, it is interesting to note that Adams et al. investigated the connection between occupation and the form of the presenting symptoms, and that the only positive correlation they were able to obtain was between the clearly defined group of brain workers and the incidence of retrobulbar neuritis as an initial symptom.

The effects of retrobulbar neuritis may be considered under the headings of (a) acute and (b) persistant.

(a) Acute. In the acute stage symptoms are more marked than signs. The commonest complaint is of mistiness or blurring of vision, in some cases progressing to complete blindness in the affected eye. This was probably the mechanism by which D'Esté's initial symptoms were produced. In most cases the affection is unilateral, but in a few instances both eyes have been reported to be involved together.[8] However it does appear that an eye is never affected twice.

If the eyes are examined during the acute attack, the common finding in the visual fields is a central scotoma of a size which depends, among other things, on the proximity of the lesion to the nerve head. The pupillary reaction to light is sluggish and ill-maintained in some cases, while the appearance of the nerve head may be surprisingly normal, being only a little paler and less well-defined than in the normal eye. In the severe case with total blindness, a mild papillitis may be seen with a few small haemorrhages. Relatively few patients complain of pain during the retro-bulbar neuritis of disseminated sclerosis, but in those who do, the pain is usually increased by eye-movement.

(b) Persistant. After the acute attack subjective recovery is usually good. On examination, however, persistant defects of vision may be found in quite a high percentage of cases. Wybar found that visual acuity and performance in Ishihara's colour discrimination test were significantly diminished in about one half of the eyes previously affected by retrobulbar neuritis, and that of 33 such eyes, 11 showed central or paracentral scotoma and 13 showed peripheral field contraction. The incidence of pallor of the optic disc varies quite widely in different series and this is hardly surprising since it is subject to individual interpretation by the observer. However it does seem that about one-third of all cases of disseminated sclerosis show pallor of one or other optic disc. Wybar has shown that there is no exact correlation of this finding with a history of retrobulbar neuritis. It is interesting to note that the stigma of temporal disc pallor, widely revered as a diagnostic sign, is not enthusiastically reported by most authorities.

NYSTAGMUS

The frequency of nystagmus was well recognised by the early writers on the subject, and this sign is included in the most famous of the many "triads" set out as aids to diagnosis i.e. Charcot's Triad of nystagmus, scanning speech and intention tremor. More modern writers are also agreed on its frequency, and it is now generally accepted as occurring in from 20 to 40% of early cases, and in from 40 to 70% of late cases. Thus far agreement is general, but on

the topic of the precise nature of the nystagmus of disseminated sclerosis less unanimity is noted. Both vertical and horizontal types are reported; the monocular variety is not uncommon; and Wybar has stressed the diagnostic importance of ataxic nystagmus, in which on lateral fixation oscillations are most prominent in the abducted eye. Subjective awareness of nystagmus (oscillopia) is also reported in a small number of cases. Because of the prolonged natural history of the disease very little work has been done on the correlation of nystagmus with anatomical location of lesions in the central nervous system. However, Savitsky and Ranzell, in a series of well-documented and autopsied cases, found plaques of demyelination in the medial longitudinal bundles of 2 patients who had showed nystagmus.

OCULOMOTOR DISORDERS

Ocular imbalance, manifest to the patient as diplopia, usually without apparent squint, is the presenting symptom of the disease in about 15% of cases, and its overall incidence in the disease course is usually about 30%. Palaysis of the sixth cranial nerve is the commonest cause of this symptom in disseminated sclerosis; third nerve palsies occur much less frequently and all are agreed that the fourth cranial nerve is spared in the great majority of cases. Because of the relative sparing of the third nerve, ptosis is not common but may occur. Physical examination may fail to reveal any demonstrable occular palsy, but in about 10% of cases some abnormality is discovered, usually in the form of simple limitation of adduction or abduction.

Apart from diplopia, supra-nuclear optic palsies may be discovered on examination in some 15% of those suffering from this disease. In these cases there is no diplopia and the clinical abnormality consists of paralysis of conjugate movement with persistence of reflex movement in the paralysed muscles. Thus, for example, a sudden loud noise may cause the patient's eyes to deviate in the direction of the paralysis, because of the persistence of connections from the auditory cortex to the optic nuclei.

PUPILLARY DISORDERS

One pupillary abnormality often described in disseminated sclerosis is the phenomenon of hippus, in which continued variations occur in the size of the pupil in sufficient amplitude to be definable by the eye of the observer. It is difficult to be definite about the usefulness of this sign as an aid to diagnosis, since no two series have produced comparable results for its incidence. For example, in one series of 264 cases collected from the literature hippus was not mentioned once, while in another series of 91 cases observed by an ophthalmic surgeon bilateral hippus was present in 37% of these patients. Here, as so often in medicine, the sign will not be detected unless it is specifically looked for. Again there is disagreement about changes in the pupillary reflexes, the incidence of which is, in any case, small. The consesus of opinion seems to be that the light reflex is absent in a small number of cases (less than 5%), and that in a very few of these cases, the fully developed Argyll-Robertson is present, in the absence of cerebro-vascular syphillis.[6] Changes in pupillary size are not now believed to be of any diagnostic importance in disseminated sclerosis, and the old view that myotic pupils were common in this disease now finds no favour.

RETINAL VEIN CHANGES

The changes which occur in the retinal veins as a result of disseminated sclerosis are described as sheathing. In this condition a thin white line is

observed on either side of the column of blood in one or more retinal veins. The affected veins are always more than two disc-diameters away from the optic disc, and the arterioles are not affected. Vitreous opacities may also be present in the region of the affected vein. Similar changes may occur in the retinal veins and arterioles as the result of chorioretinitis, diabetis mellitus or hypertension, but in all these cases other associated ophthalmoscopic changes serve to indicate the primary cause of the abnormality. A reliability of about 80% can be claimed for this sign in the diagnosis of disseminated sclerosis,[5] but unfortunately (from the diagnostic viewpoint) of all patients with disseminated sclerosis, only 10% demonstrate this sign.

These are the principal changes which occur in the eyes of a patient suffering from disseminated sclerosis. Taken as a whole, and in conjunction with an adequate history and complete physical examination, they may be of considerable help. But it is worth emphasising that, neither these eye changes nor any other isolated sign, is pathognomonic of the disease. For example, nystagmus was observed in a patient with progressive paraplegia, due, not to disseminated sclerosis, but to an operable spinal tumor. At present the diagnosis of disseminated sclerosis is important from the point of view of exclusion of any potentially tractable condition, but it is equally important to bear in mind that some advance in treatment, comparable in effectiveness to that of antobiotics in infection, or insulin therapy in diabetes may at any time be made. It is with this possibility constantly in view that the early diagnosis of disseminated sclerosis should be made.

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