

## CHRONIC RETROBULBAR AND CHIASMAL NEURITIS\*†

BY

BRYAN ASHWORTH

*From the Department of Medicine, University of Bristol,  
and the United Bristol Hospitals*

ACUTE retrobulbar neuritis is a common disorder, easily recognized if the characteristic features are known, and usually followed by a good recovery of vision. In contrast to this, chronic retrobulbar neuritis is relatively rare, presents problems in differential diagnosis and is associated with progressive visual disturbance which is usually permanent and sometimes severe.

This paper is based on a clinical study of patients under observation for at least 2 years. It will be emphasized that progressive visual failure does occur in multiple sclerosis and may contribute to the disability in that disease.

### Material

Fifteen patients with chronic retrobulbar neuritis (9 bilateral and 6 unilateral) and one patient with chronic chiasmal neuritis were studied. The bilateral cases were all males and the unilateral cases all females. The age range was 18 to 54 years (mean 35.5).

The predominance of bilateral cases in this series probably reflects the tendency for patients with bilateral visual symptoms to attend an eye hospital.

*History.*—In some patients there was a clear history of acute retrobulbar neuritis at the onset of symptoms. This was usually followed by improvement in the vision as the acute attack resolved but later there was gradual deterioration. In other subjects recurrent acute episodes affected the same or the other eye. When these were severe the details could often be recalled but in milder cases it was not easy to differentiate between recurrent acute episodes and chronic insidious progression on the basis of the clinical history. However, in some subjects who seemed to be reliable witnesses, there was no history of an acute episode and the presenting symptom was failing vision of insidious onset and progressive course.

### Clinical Reports

These are divided into five groups and representative case histories are described to illustrate each group.

#### *Group 1. Acute retrobulbar neuritis with recovery followed by gradual deterioration (two cases)*

**Case 1, a girl aged 18 years**, was first seen in 1962, complaining of blurred vision in the left eye for one day. The left globe was tender and the pupil reacted sluggishly to light. Corrected visual acuity was 6/4 right, and 6/18 on the left with central scotoma. During the course of the next week vision in the left eye deteriorated to perception of light, and 2 months from the onset it had improved to 3/60. Later the vision in the left eye deteriorated and in 1965 was reduced to counting fingers at 2 m. There was marked pallor of the left optic disc.

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† Address for reprints: Department of Neurology, Royal Infirmary, Manchester 13.

*Group 2. Acute retrobulbar neuritis with progression (two cases)*

**Case 4, a woman aged 36 years**, was first seen in 1958 when she complained of sudden loss of vision in the right eye. The right globe was tender to touch and there was slight swelling of the right optic disc. Corrected visual acuity was 2/60 on the right with central scotoma, and 6/5 left. In 1962 there was marked pallor of the right disc and gross impairment of colour vision on that side. The corrected acuity was 6/12 right and 6/6 left.

*Group 3. Acute retrobulbar neuritis—recurrence—progression (two cases)*

**Case 6, a woman aged 52 years**, was first seen in 1950 with a history of blurred vision in the left eye for 10 days. She also described a similar previous episode in 1949 which had cleared in 2 days. There was slight swelling of the left disc. One month later the symptoms and signs had cleared and the visual acuity was normal.

In 1958 she complained that vision in the right eye had been blurred for a year. The corrected visual acuity was 6/12 right and 6/5 left and no other abnormality was found. General examination revealed nystagmus and slight cerebellar ataxia affecting the left upper limb. The cerebrospinal fluid was normal.

In 1962 the corrected visual acuity was still 6/12 right and 6/6 left; both discs showed abnormal pallor.

In 1963 the neurological disability had increased and the cerebrospinal fluid showed a paretic Lange curve 5544332210 with negative Wassermann reaction. She died with advanced multiple sclerosis in 1964.

*Group 4. Insidious and progressive visual failure (nine cases)*

**Case 7, a man aged 22 years**, was seen in 1964 when he complained of poor vision in the left eye for a year and blurred vision of the right eye for 2 months. Vision on the left side was reduced to perception of light with marked pallor of the left disc. The visual acuity, fundus, and field on the right side were all normal. Investigations included normal cerebrospinal fluid, normal electroencephalogram, and normal air encephalogram with negative Wassermann reaction in both blood and CSF. 3 months later he complained that the vision in the right eye had become worse: the corrected acuity was then 2/60 right, with slight oedema of the right disc, and a large blind spot with central scotoma for red and white.

In 1965 this patient was investigated at another hospital. The optic nerves were explored at operation but there was no evidence of compression.

**Case 9, a man aged 28 years**, was seen in 1962, when he complained of blurred vision in both eyes of a few weeks' duration; the date of onset could not be accurately determined.

In 1955 he had been admitted to another hospital with acute vertigo, the symptoms had settled in a few weeks and lumbar puncture at that time had revealed normal fluid.

In 1962 the corrected visual acuity was 6/24 right and 6/12 left, optic discs both pale, central scotoma on the right side. There was nystagmus, intention tremor of the right upper limb, satisfactory reflexes and no abnormality of the motor system. The cerebrospinal fluid was normal.

A few months later there was an episode of tingling in the fingers which persisted for 2 weeks, and during the next 3 years he was seen regularly and complained of increasing visual difficulties.

In 1965 the corrected visual acuity was 6/36 N36 right and 6/18 N10 left. The visual fields showed enlargement of the blind spots with central scotomata for red and white.

**Case 10, a man aged 41 years**, was seen in 1964, when he complained of blurred vision in both eyes for one year which had slowly worsened.

In 1960 he had complained of numbness and weakness of the legs and investigation at that time of the cerebrospinal fluid showed 10 lymphocytes/cu.mm. but no other abnormality.

In 1964 the corrected visual acuity was 6/36 right and 6/60 left, with pallor of both optic discs. There was an enlarged blind spot on both sides, but no scotoma was clearly defined. He had ataxic nystagmus, absent abdominal reflexes and extensor plantar responses, ataxia of the lower limbs, and impaired vibration sense at the ankles. The cerebrospinal fluid showed 2 cells/cu.mm., with protein 44 mg. per cent. Lange curve 4321100000. Wassermann reaction negative in blood and CSF. The vision continued to deteriorate.

*Group 5. Chronic chiasmal neuropathy (one case)*

**Case 16, a man aged 42 years,** was seen in 1963 when he complained of blurred vision in both eyes, weakness and clumsiness of the left arm and leg, and tingling in the left hand.

In 1944 he had first complained of blurred vision in both eyes; this had never improved and during the past year it had deteriorated.

Since 1962 he had been aware of tingling in the left hand and clumsiness of movement of the left arm and had observed that both the clumsiness and the blurred vision were worse after exercise. He did not smoke and the family history was not contributory.

The corrected visual acuity was 6/60 right and 6/18 left, pupil reactions to light were not well held, the optic discs and fundi were within normal limits, and the visual fields showed bitemporal hemianopic scotomata (Figure). He had ataxic nystagmus, tendon jerks all increased and extensor plantar responses, bilateral cerebellar signs, and no sensory disturbance. The Wassermann reaction was negative in blood and CSF, and the CSF was normal.

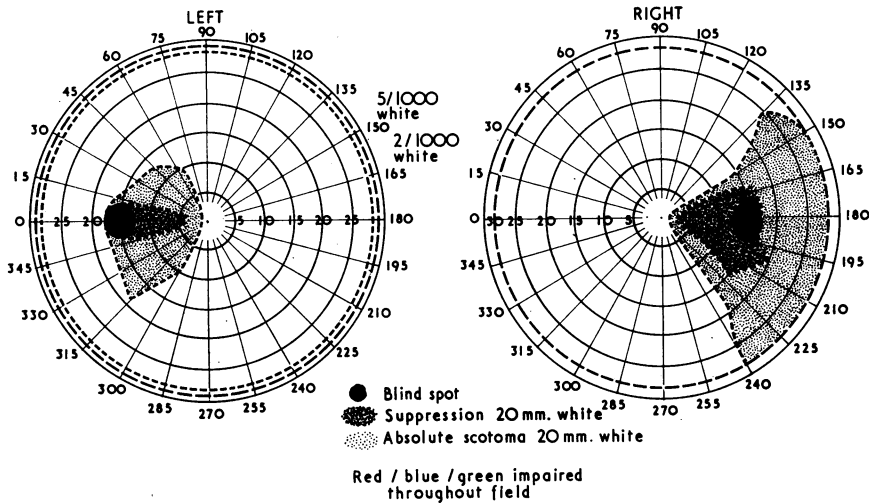


FIGURE.—Bitemporal hemianopic scotomata in Case 16.

### Clinical Findings

#### (A) Ocular

*Visual Acuity.*—Of 24 eyes in fifteen patients with retrobulbar neuritis, the corrected acuity was 6/18 or better in five, between 6/18 and 6/60 in nine, and less than 6/60 in ten. One patient had normal acuity when first seen but complained of pain and the pupil reaction was sluggish; later the vision was reduced to perception of light.

*Pain and Tenderness* of the globe were reported on three occasions. One patient complained of a gritty sensation in the eye.

*Pupil Reaction to Light.*—This was sluggish in some cases and failure to maintain the constriction was an almost constant feature.

*Optic Disc.*—The disc of the affected eye was judged to be abnormally pale in all cases and usually at an early stage. In four cases slight oedema of the disc (papillitis) was observed. In two of these cases oedema of one disc was combined with gross pallor of the other (optic atrophy). This Foster Kennedy syndrome is, of course, entirely different in aetiology from the condition in which a frontal tumour causes optic atrophy from compression of one optic nerve and papilloedema on the other side due to raised intracranial pressure to which this eponym is usually applied (Kennedy, 1911).

*Visual Fields.*—The usual features were central scotoma, enlargement of the blind spot and a striking impairment of colour vision. Constriction of the peripheral field might occur later but was not a prominent feature.

### (B) Neurological

Signs of organic neurological disease were present in seven patients, in three of whom the cerebrospinal fluid was also abnormal, showing a paretic Lange curve in two and a slight pleocytosis in two, with a negative Wassermann reaction. In two further cases the history suggested neurological disease but there were no confirmatory signs. In all of these the most likely diagnosis was multiple sclerosis and in six the findings and clinical course were typical. Occasionally, the clinical history at once suggested multiple sclerosis; for example, one subject complained of an episode of numbness of the right upper lip combined with blurred vision in the left eye.

One other feature is of interest. The patient with chiasmal neuritis stated that he had noticed a deterioration in his vision after exertion. This has been noted in retrobulbar neuritis by Earl (1964) but was not sought in the other cases in this series.

### Prognosis

Within a year or two of the onset of symptoms in retrobulbar neuritis a striking improvement in vision may occur, particularly in children. After 2 years, as in the cases described here, improvement was not observed and further deterioration may occur.

### Discussion

The concept of chronic retrobulbar neuritis is an old one but it has tended to be regarded as the end-result of an acute process rather than as an insidious and progressive disorder. Parinaud (1884) described three types of amblyopia in patients with *sclérose en plaques*:

- (a) Slow development with disturbance of red and green vision.
- (b) Rapid onset, pallor of the disc which persists as the vision improves, and variable field defect.
- (c) Unilateral, gross, and persistent (rare).

Schlossman and Phillips (1954), in a clinical study, reported 72 cases of optic neuritis; 69 per cent. of the patients had multiple sclerosis and visual disturbance was the first symptom in seventeen. In seven there was a history of gradual visual loss.

Hierons and Lyle (1959) reported 47 cases of bilateral retrobulbar neuritis. The follow-up was from 2 to 15 years (average 6) and the series included thirteen children in whom the visual recovery was generally good.

In the present series of sixteen patients, the associated neurological findings suggested multiple sclerosis in seven and there was a probability of this disease in two others. It is generally accepted that acute retrobulbar neuritis is commonly associated with multiple sclerosis and is often a presenting feature of the disease. There may be a long remission before other evidence of the disease declares itself and in a few cases this may never occur (McAlpine, 1965).

The differential diagnosis of progressive visual failure due to optic nerve disease may be considered under three headings:

1. Compression of the optic nerve.
2. Systemic disease with neurological involvement.
3. Intrinsic neurological disease.

Optic nerve compression demands consideration in any progressive failure of vision not due to disease of the globe. The features have been discussed by Meadows (1949) and Hughes (1954). No firm rules can be laid down and each case presents a special problem.

A central scotoma which breaks through to the periphery of the field suggests a compressive lesion, while a marked impairment of colour vision favours retrobulbar neuritis. Special radiological techniques may be helpful, such as views of the optic foramina, air encephalography, and arteriography. If doubt remains, surgical exploration may be indicated. If this is done exposure must be complete, as is well illustrated by a case reported by Sanders and Falconer (1964); their patient had a centrocaecal scotoma with a 7-month history of progressive visual loss. Investigations were not contributory and when the orbit was explored a minute meningioma was found to be compressing the intracanalicular part of the optic nerve.

Among systemic diseases, sarcoidosis may closely mimic retrobulbar neuritis and multiple sclerosis. This disease might be suspected from the appearance of hilar adenopathy in a chest radiograph, a high protein level in the cerebrospinal fluid (over 100 mg. per cent.), or raised serum globulin where the other clinical features are absent. Syphilitic optic neuritis is rarely seen but may be arrested if treatment is prompt: it is usually bilateral. The recognition of vitamin B<sub>12</sub> deficiency is important, but it is rare in patients under 40 years of age and is readily diagnosed from the serum level. The family history is always important and a history of exposure to toxic chemicals or drugs must be sought.

Of the neurological diseases, familial conditions are usually easy to recognize. In Leber's disease there may be neurological features other than optic atrophy and it has been suggested that the process may have a metabolic basis in disordered cyanide metabolism which might be aggravated by smoking (Adams, Blackwood, and Wilson, 1966). Neuromyelitis optica is probably closely related to multiple sclerosis.

### Summary

The diagnosis of chronic retrobulbar and chiasmal neuritis is reviewed in a series of sixteen cases. Multiple sclerosis is thought to be the commonest cause. The occurrence of progressive visual failure in multiple sclerosis is emphasized. Important conditions in differential diagnosis are discussed.

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