

RETROBULBAR NEURITIS.

BY WILLIAM GEORGE SYM, M.D., F.R.C.S.E.,

SENIOR ASSISTANT OPHTHALMIC SURGEON, ROYAL INFIRMARY, EDINBURGH, AND OPHTHALMIC SURGEON TO LEITH HOSPITAL.

THERE is much in the subject of retrobulbar neuritis to engage the attention and the interest of the intelligent practitioner. It is a disease, in the first place, which merits attention by the fact that our information and knowledge regarding it are still decidedly insufficient, and, indeed, we sometimes commit ourselves to a diagnosis of its presence solely on the strength of certain subjective symptoms when the patient presents to us no objective signs which would confirm our opinion.

Another important feature about retrobulbar neuritis is the marked—indeed, startling—contrast which it presents, in regard to symptoms, signs, etiology, prognosis, and treatment, with the form of neuritis with which one is more familiar in connection with nervous diseases.

A third point is that under the head of this name we group a number of diseased conditions which have, as we shall see, certain elements in common, but which differ widely in mode of origin, in cause, in appropriate treatment, in prognosis as regards life and as regards vision.

Some time ago a very interesting discussion took place at the Ophthalmological Society of the United Kingdom, introduced by a valuable paper read by Mr. Marcus Gunn, upon this very subject; but, able and instructive as both paper and discussion were, there was manifested a tendency to limit the field of view to one which, in my opinion, was too narrow for the real range of the subject.

The name of retrobulbar or retro-ocular neuritis was introduced by von Graefe to indicate such inflammatory diseases of the optic nerve as arose by continuity of tissue with other inflamed parts, or which took their origin not from alteration of intracranial pressure, but which began primarily in the optic nerve just as they might do in any other motor or (common) sensory nerve, to be followed by loss of function. Since in a certain proportion of cases no ophthalmoscopic changes were seen, it was natural to refer the seat of lesion to a portion of the nerve behind what could possibly be examined, while the occurrence in other cases of evident inflammatory alterations, followed by marked atrophy, gave one the key to their true nature, and enabled one to be confident that a stage of inflammation had been passed through, even in the cases which merely exhibited in the ophthalmoscopic picture the appearance of atrophy.

The definition of the disease suggested by Mr. Gunn, while it tends in my judgment to exclude some of the group, is certainly very valuable and will repay careful study. He says, "Rapid failure of vision, usually in one eye only, often accompanied by pain and tenderness in the neigh-

horhood, absence of early ophthalmoscopic changes, and a tendency to recovery, are the usual prominent features." I propose to discuss these features one by one in order that we may obtain a clear idea of the disease, while at the same time I shall indicate certain facts regarding the disease which seem to me not to be strictly in accordance with Mr. Gunn's definition.

1. *Rapid Failure of Vision.* The failure of vision which occurs is of a peculiar type, inasmuch as it affects principally or solely the central region of the field. The degree of defect varies from a mere loss of definition of the outlines of objects, especially under certain particular circumstances, as we shall see immediately, to a complete though circumscribed loss of perception of light. Thus, the patient may have 6/vi of vision or absolutely none in the central area. In the less severe cases there are two methods of testing in particular which are of much practical value, viz., testing with colors and with Bjerrum's types. In regard to colors we find that the patient will display an inability to perceive accurately with that portion of his retina which is in the immediate neighborhood of the papilla and macula the tint of a small object of from 3 to 5 millimetres in diameter held at a distance of a couple of feet. Such an object held at such a distance from that part of the normal retina is quite capable of arousing a precise estimate of the color which it possesses, but not in such patients as we are now considering. The patient is made to look fixedly at a small white spot on a blackboard, while one places in the near neighborhood of the white spot, but to its outer side, a suitably sized colored spot painted on, or affixed to the end of, a black rod, when it will be found that in a moderate case blue and yellow are fairly correctly seen, while red and green are lost, and appear as yellow and white respectively. It is on account of the perception being retained of the yellow which is always mixed with our commercial reds, as well as to loss of saturation, that red is pronounced to be "yellow" or "brass-colored;" and we frequently notice, too, that a patient will call green "blue" because of the presence of blue in the green employed. It is only in rather bad cases that perception of all colors is lost at the centre; in the majority perception of blue and yellow is retained—of those colors usually which seem, by the fact of their having a larger field of visibility in normal conditions, to have a greater hold upon the retina, so to speak. That the perception of red and of green is less intense in the normal individual, is less a part of our nature, if one might use the phrase, seems also to be indicated by the fact that where congenital color-blindness is present, the probabilities are greatly in favor of its being of this type rather than of the blue-yellow type. In fact, it is only very rarely that persons have difficulty with those latter colors, confusion of which, however, is sometimes seen as a hysterical condition. It has been stated by some authorities that the periph-

ery of the fields as regards color in those patients of whom we have been speaking is apt to be restricted; from my own experience I cannot confirm this assertion, and do not consider it to be accurate.

Bjerrum's types, just mentioned, consist of a series of letters printed on a board much as the ordinary test-types are, but the contrast between the print and the background is reduced by printing the letters in dark gray upon a background of lighter gray (or *vice versa*) in place of black upon white. They may also be printed in green on a faintly green background. Thus the ability to distinguish minor degrees of contrast is tested. These types are made in various ways, but the system which seems to me best is to have the letters from the top of the board to the bottom all of one size, but the contrast between letter and surface diminishing toward the foot. It will be found on examining a patient with retrobulbar neuritis that his vision with these types is bad out of proportion to the degree of his defect with the black-on-white types; he has difficulty in perceiving differences where the contrast is slight. The patient often tells us the same facts about himself spontaneously, for he will say that he has great difficulty in recognizing the faces of his friends. We recognize faces largely by our knowledge and perception of the small differences in illumination, shade, tint, etc., arising partly from the prominence and recession of features, and if these are blurred and blotted, if only coarse and harsh contrasts can be appreciated, a face will not be recognized. Our patient knows the figure, perhaps the peculiar walk, of an acquaintance, but the features he cannot be sure of except close at hand.

Now, the point in regard to this failure of vision which next merits attention is the influence exerted by variations in the amount of illumination. Many of such patients may tell us (though here again, of course, they are little liable to notice the fact when one eye only is affected) that, while vision is bad during the bright hours of the day, it improves in the evening or in dull light. It is not at all probable that there is any actual improvement in the acuity of vision, at least I am not aware that any such has been proved, but at all events it is true relatively that the patient sees better in a subdued light. Why this should be is not easy to say, but it is possible that it may be due to the exhaustion by the brighter light of the affected fibres of the nerve. In this connection, too, it has been observed that vision is better after a night's rest, worse on fatigue or prolonged want of food.

The patient is apt to complain of a fog or mist before the eye, usually of but slight density; and this not in proportion to the degree of defect of vision. Thus a patient whose vision is nearly normal will complain much of this fog, while another whose vision is very much lowered will admit perception only of a delicate, luminous, perhaps faintly colored haze. Sometimes this haze moves or shimmers like the heat haze we see

on looking at a landscape on a hot summer's day, or like the appearance seen on looking over the top of a burning lamp or gas-jet. This appearance is probably due to the insufficient "insulating" of the nerve fibres as a result of the inflammation, the nervous stimulation thus passing from one nerve fibre to another contiguous one.

In retrobulbar neuritis one never meets with a certain symptom present in ordinary neuritis in a considerable proportion of cases, and which was first described by Hughlings Jackson, the occurrence, namely, of temporary, almost momentary attacks of blindness.

2. The second point in Mr. Gunn's description of the disease is that the condition is *usually present in one eye only*. It may be that this is true, but the truth of the statement will depend almost completely upon whether one regards tobacco (and other toxic) amblyopia as a variety of this disease or not. It is difficult clinically to make a hard-and-fast line of distinction, and it is probable that the conditions differ rather in degree than in nature. I do not wish to discuss the matter at present; I may recur to it on another occasion. I would only say now that many points are in favor of the opinion that toxic amblyopia, if it be not actually neuritic, if there be lacking some of the real inflammatory element, is at the least so very closely allied to neuritis, and is so apt if untreated to go on to permanent alteration of tissue in a manner so similar to that brought about by a neuritis, that I for one cannot put the two conditions into separate categories as altogether distinct and non-related entities, as some do. Looking to the comparative frequency of toxic amblyopia, I can only with reservation admit that the condition is usually present in one eye only. But even leaving out of account for the meantime the cases of toxic amblyopia, it is not such a very infrequent occurrence that a patient recovers from an attack of retrobulbar neuritis in one eye only to be similarly affected in the other. I have seen this occur in a patient apparently in other respects in perfect health, except that she—a strong young woman about thirty years of age—was rather rheumatic in temperament.

3. The lesion is often accompanied by *pain and tenderness* in the neighborhood, in those cases at least in which there is inflammation of neighboring structure. It is an interesting question whether if the inflammation be limited to the nerve itself any pain will be experienced. Does the occurrence of pain not depend upon neighboring tissues and the nerve sheath being involved? We do not know whether the optic nerve has any "common" sensation or not; there are records to show that on section of it a sensation of very bright light is experienced, and there are also observations directly contradictory of this. Even if a sensation of light be experienced, that would not prove the question. Pain is but rarely complained of spontaneously in cases of retrobulbar neuritis, but the patients very often admit a sensation of weariness,

aching, or discomfort on forcibly moving the eyes in one direction—in my experience this direction is usually upward. Some have attempted to make out that there is a certain relation between the painful direction and the area of the field chiefly involved—e. g., that if pain is felt on looking upward, the upper region of the field will be found to be the most seriously affected; for my part I have no evidence of the truth of this assertion, which seems to me, indeed, to be unsound, and not to be substantiated. The pain on movement is no doubt to be attributed to the tension of the sheath of the nerve (itself probably inflamed) on exaggerated movements. Pain, or at least some sense of discomfort, is apt to be experienced too when the eye is even very gently pressed back into the orbit by the fingers placed over the upper lid, but this is by no means constant and may be due solely to pressure on an inflamed periorbital tissue. There is in some cases spontaneous complaint of a dull ache in the forehead, not amounting to pain.

4. *Absence of early ophthalmic signs* is a feature of great importance, and, since some cases run their course from beginning to end with practically no change in the aspect of the disk whatever, one ought, perhaps, rather to say that ophthalmoscopic signs are usually absent in the early stages, and may be absent altogether. However, it is more usual to find certain manifestations becoming obvious as the disease progresses. And it is most certainly true that in the great proportion of cases with which we meet, the ophthalmoscopic appearances are trifling in proportion to the alteration of vision, are very slight relatively to the gravity of the lesion as regards sight; and herein these cases form a marked and striking contrast with those of ordinary optic neuritis, for in this latter condition the ophthalmoscopic changes may be very great while vision remains perfectly unaltered as regards acuity, field, color, perception, etc., where in fact the visual symptoms are *nil*. The changes which are usually to be found in the later stages, and which may be present even in the early, are (1) slight general increase in the vascularity of the disk, which acquires a grayish-pink color, this being more marked in some districts of the nerve-head than in others; (2) fulness and tortuosity of the larger veins, which acquire a more distinctly marked double contour and light-line along with their darker color, on account of their more decided cylindrical shape and circular form in section; (3) a faint blur or haze hanging like a fine cloud or mist over the disk, suggesting an infiltration of that portion of the vitreous humor immediately overlying the nerve-head; (4) slight indistinctness and lack of definition of the margin of the disk, which fades almost imperceptibly into the surrounding fundus. Such are the signs, all nearly invariably being but slight in degree, which are most usually to be seen when first there is any manifestation; but in a certain number of cases even in the earlier stages, and in a very large number in later stages, there is on the con-

trary marked pallor of the disk, at all events, of its outer quadrant. This pallor is usually simple—that is, without any other change such as evidence of perivasculitis or of deposit in the physiological cup, or any material alteration in the calibre of the vessels, or hemorrhage in the neighboring parts of the fundus. Diminution in the size of the retinal arteries is, however, sometimes present quite early, and in most cases appears sooner or later, and there may be some blurring and disturbance of pigment about the margins of the disk, especially perhaps to the outer side.

5. The *tendency to recovery* is also an important feature in the unilateral type of retrobulbar neuritis. In the majority of such cases the prognosis is decidedly good, and one is justified in expecting distinct improvement after the lapse of a very few weeks or even not many days. This amelioration is in fact sometimes synchronous with the appearance of visible ophthalmoscopic changes, if the injection has spread down to the disk, so that we have the curious state of affairs that a patient is improving in symptoms while the signs are becoming worse. In a large proportion of the cases the recovery of vision seems to be absolutely complete, but this is a matter somewhat difficult of proof, because the final stages are apt to be slow, and patients are thus apt to be neglectful to return for examination; they feel quite well, have returned to their ordinary habits and duties, and tend to postpone the final examination *sine die*. In a number of the “recoveries,” however, it is more than probable that careful perimetric testing would elicit the existence of a small paracentral area of still deficient vision with imperfection of the color sense. All cases, too, do not recover, and in a certain percentage a decided and perhaps an absolute scotoma remains permanently. Here again we often see a marked disagreement between the signs presented and the symptoms complained of, for a patient may have optic disks almost dead-white in color, and retinal arteries small in calibre, and yet possess capital vision, or, on the other hand, good-looking disks with only a slight degree of pallor in the outer quadrant, hardly more than is often present under physiological conditions, and yet a distinct and distressing scotoma.

I pass by the subject of the microscopic appearances of retrobulbar neuritis, since so careful and able an account of these appeared so recently in this JOURNAL (September, 1897) from the hand of Dr. de Schweinitz, whose observations practically corroborate those made in the first instance by Samelsohn.

But to turn to the larger sense of the subject of retrobulbar neuritis, and to include in one view all those cases and types which I believe to be of one and the same character, though manifested somewhat differently under varying circumstances, and to include under one heading all cases with a similar “local” pathology (if the phrase may be per-

mitted), I should divide the cases somewhat in the following manner: Retrobulbar neuritis may be unilateral or bilateral.

(a) A unilateral neuritis may be due:

1. To the spreading to the nerve of local inflammation.
2. To syphilis.
3. To rheumatism.
4. To gout.
5. To malaria.

It is of these unilateral cases that we have chiefly been speaking hitherto; but I fail to see any essential feature, save in the facts that they are unilateral and tend to recovery, wherein they differ from the (b) bilateral variety, of which I recognize the following forms:

1. Those occurring along with diseases of the spinal cord, especially locomotor ataxia and disseminated sclerosis, as well as with general paralysis of the insane.

In this class the inflammatory element may be almost completely eliminated, and the cases may be and very often are regarded as optic atrophy.

2. Those progressive cases occurring simply—that is, without any apparent relation to cord lesions, just as the ordinary type of optic atrophy occurs sometimes. It is true of this retrobulbar atrophy (for in this class also the atrophic character is the more obvious) as it is true of the ordinary optic atrophy that in a certain number of these patients spinal or head symptoms may eventually be manifested, although only after the lapse of a long time; but in a certain proportion also no such symptoms are ever displayed.

3. Cases of what has been called stationary scotomatous atrophy. This is not a good name for the condition, for one associates the idea of a progressive deterioration with the word atrophy, but there is none here, and we have a contradiction in terms. In these cases, most frequently in young persons, we find a central scotoma, usually small and generally, but not always, absolute, along with a very white and even chalky looking nerve-head, and with vessels, especially the arteries, somewhat diminished in calibre. In some of these cases there is a history pointing to a meningitis or possibly a periostitis at the apices of the orbits, or in the neighborhood of the foramina some time previously, but this is not the case in all.

4. Cases of what is known as hereditary optic atrophy. In this condition we have another instance of the unfortunate use of terms, for the disease is not primarily—as I believe—an atrophy, but a neuritis, and it certainly is not, strictly speaking, hereditary. “Leher’s atrophy,” as the condition is sometimes called, after the professor in Heidelberg, who has given a classical description of it, is a most interesting malady, but this is not the place to do more than merely indicate its outlines.

It is apt to manifest itself among the males of a family as they attain the age of eighteen to twenty-three years; the parents have usually been quite healthy, but several of the sons are attacked by it one after the other as the age indicated is reached. The daughters of the family (sisters of the patients) are usually quite free of the disease, but may transmit it to their sons. Both sons and daughters of the patients themselves may remain, and usually do remain, healthy, but the daughters may again transmit it to their sons; the liability usually dies out in the third generation. When women are affected by it, the amblyopia usually comes on much later in life, about the menopause, rather than in the early adult period. In the case of males the patients are usually in perfect apparent health, and no cause can be assigned for the onset of the blindness, which may come on with great rapidity, a few days sufficing practically to extinguish vision, while in a certain number of instances even more rapid loss of sight would appear to have occurred. Over the central area of the field of vision the amblyopia may be complete, perception of light even not being retained, and the only portion of the field remaining may be a mere ring round the periphery. All cases are not so bad as this, and even at the centre the scotoma may not be absolute. So far as my experience goes there is no peripheral restriction of the field. It is not always possible to distinguish between this class and that of the stationary scotomatous atrophy.

5. Without prejudice to the question of the precise condition of the nerves and their vascular supply in such cases, I think one may fairly include here the cases of toxic amblyopia, typically represented by "tobacco blindness," as coming under the same category. Whether or not there is in this condition an actual neuritis—and for my part, I think it highly improbable, for many reasons—there can be no doubt that the borderline which separates toxic amblyopia from retrobulbar neuritis and the atrophy which is so apt to follow is very narrow, and is very easily overstepped by a patient who will not take warning as to his state of health, and will not moderate his supply of tobacco, alcohol, or whatever may be his particular "indulgence."

So far as regards the treatment of retrobulbar neuritis generally, it is not necessary to say much. In the unilateral cases one must attack vigorously the vicious diathesis, whether syphilis, gout, or what not, which has led to the condition, and at the same time use derivatives and local depletion. For example, in the distinctly rheumatic cases I place most reliance on purging, salicylates, and blistering applied to the temple. At the same time one must encourage a hearty, vigorous action of skin and kidneys, by warm baths or even Turkish baths, by keeping the patient wrapped in flannel, and if needful by drugging with diuretics and diaphoretics. I believe that subcutaneous injection of pilocarpine will be found as useful in this disease as it is in persistent

scleritis. Some place much faith in iodide of potassium and in mercury, even when syphilis is not present.

In the more long-standing cases, and especially in those which are more nearly allied to atrophy, strychnine is our chief reliance; but even strychnine has but little influence on the course of the disease.

It should not be forgotten that recurrence of an acute attack is rather apt to take place on exposure to conditions similar to those which brought it on before; therefore patients ought to submit for the future to regulations of diet, dress, and habits, since a second attack is more likely to leave permanent and great evil results than a first onslaught.

HEAD-NODDING AND HEAD-ROTATION USUALLY ASSOCIATED WITH NYSTAGMUS IN VERY YOUNG CHILDREN.

WITH REPORT OF TWO CASES.¹

BY CHARLES J. ALDRICH, M.D.,

LECTURER ON CLINICAL NEUROLOGY, COLLEGE OF PHYSICIANS AND SURGEONS; NEUROLOGIST TO CLEVELAND GENERAL HOSPITAL AND DISPENSARY; NEUROLOGIST TO CLEVELAND CITY HOSPITAL.

OF all the functional nerve-diseases of children few are more rare and none possess greater interest than the rhythmical nodding or horizontal rotary movements of the head, commonly associated with nystagmus. Medical literature makes scant mention of these cases. Some of the books on diseases of children briefly refer to the disorder, but seem to confuse the reader by associating it with *epilepsia nutans* or *tic salaam*, which, without doubt, is a true epilepsy.

In 1851 Romberg and Henoch published the first observations of this peculiar affection that I have been able to find; later Hadden, of London, reported a series of twelve cases, and referred to similar ones; soon after he reported another series of ten cases; lastly, in 1892, Peterson, of New York, detailed five cases. Others, to whom due credit will be given, have reported cases, but the fore-mentioned observers have contributed about all that is known of this curious disorder.

In the very noble address of J. Hughlings Jackson before the Ophthalmological Society of the United Kingdom, in 1889, occurs the following: "Physicians should try to help ophthalmology by bringing forward cases little likely to come under the notice of ophthalmic surgeons. Cases of spasmus nutans with nystagmus (a variety, I think, of spinal, or rather spinal system chorea, a symptomatic condition akin to canine chorea) would as 'living specimens' interest all our members." This was said eight years after Henoch's last published observations, and thirty-

¹ Read before the Ohio State Medical Society, May, 1898.

eight years after Romberg, Hensen, Faher, and Ebert had published cases; yet it remained for Hadden, in 1890, to differentiate clearly head-nodding from *epilepsia nutans* or *tic salaam*. And it is to his clear sight that we are indebted for the observation that "There are cases of nystagmus unassociated with head movements the nature of which is hard to explain, . . . some, at least, of these instances are, in my opinion, fundamentally the same as the class under consideration."

The quotation from Jackson is given not alone for the purpose of calling attention to the confusion then existing in regard to the disease, but to serve also as an apology for reporting the following cases:

CASE I.—E. G., a large, healthy appearing, breast-fed female, sixteen months old; born at term after a short normal labor of a healthy mother. The father is well but absolutely deaf from a large dose of quinine. The patient has a strong-looking brother of ten years that possesses a well-defined rachitic rosary and bowed shins. One sister, two years older than the patient, while under my care, died of tubercular meningitis. Although the child is large and plump, the osteal epiphyses are enlarged, and a well-marked rosary is felt. The anterior fontanelle is nearly closed and twelve strong teeth have erupted. No other *stigmata* are present.

The mother says that this is the first sickness of any kind that the child has suffered. It was noticed for a week preceding my visit that she kept turning her head from right to left in a curiously rapid and regular manner, also that she appeared fretful and vomited frequently. On examination I found that the circulation and temperature were normal; tongue was coated; bowels constipated; appetite good, but without apparent cause she had vomited three or four times in the forepart of the day; to use the mother's expression, "had spewed about a yard." When questioned, the mother admitted that the child had tumbled down and bumped its head, to which she seemed to attach little importance, for she added, "that it got no more than its share of bumps."

The movements of the head were distinctly horizontal and rotary; they stopped during sleep, and were for a few seconds arrested when the attention was strikingly drawn. The sight of my watch stopped them, but in an instant they were resumed, although she continued busily looking at the dial. A sharp whistle had the same evanescent effect. The movements were singularly smooth, easy, regular, and in almost perfect rhythm, and occurred about seventy-two to eighty times a minute. The excursion began with the chin a trifle to the left of the median line and was carried horizontally to the left about 40 degrees, and then brought back to repeat the act. As before said, the movement was smooth, easy, and rhythmical, without any spasmodic or jerky elements whatever.

While observing these gyral movements of the head I discovered a rapid horizontal nystagmus of the left eye, made more distinct or increased when the eyes were directed to the left. From the notes of Dr. William Brunner, who at my instance kindly examined the eyes, I quote the following: "Examination of the eyes of E. G. shows a slight convergent strabismus; fixation with the right eye. In the left is a fine rapid horizontal nystagmus slightly more marked, perhaps, when the