

## THE EYE-SYMPTOMS IN LOCOMOTOR ATAXIA.

(A CLINICAL LECTURE.)

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GENTLEMEN,—I purpose to-day to explain and illustrate the Eye-symptoms which occur in Locomotor Ataxia. They are very common, as you will perceive when I tell you that they were present, in one form or other, in 41 out of 64 cases tabulated by Eulenburg,<sup>1</sup> and in 18 out of 20 cases of which I have notes. It is mainly upon a study of these 20 cases that the description I am to give you is founded.

There are four sets of symptoms :

- I. Double Vision, Squinting, and Ptosis.
- II. Changes in the Pupil.
- III. Amblyopia and Amaurosis.
- IV. Colour-Blindness.

### *I. Double Vision, Squinting, and Ptosis.*

These temporary paralyses are very common affections, and manifest themselves often by double vision without apparent squint, sometimes by double vision only when the eyes are turned in particular directions, often also by distinct squint and by ptosis. The existence of the last-named symptom forbids the adoption of any hypothesis of spasm or ataxia, and shows clearly that these conditions are manifestations of the well-recognised tendency to temporary paralysis which exists throughout the course of the disease, and especially in its early stages.

<sup>1</sup> 'Lehrbuch d. Nervenkr., 2te Aufl. Bd. ii. s. 448.

Out of the 64 cases recorded by Eulenburg, 25 had strabismus. Of these 25, 19 had divergent strabismus, the third nerve being paralysed; 6 had convergent strabismus, the sixth nerve being affected; and 4 of the divergent cases had, in addition, paralytic ptosis. Of my 20 cases, I find that 8 had paralytic eye-symptoms, 5 having strabismus, 3 ptosis, and 4 diplopia, without manifest squint. I have not seen dilatation of the pupil occur along with the squint in any of these cases, and from this, as well as the other facts, you observe that all the branches of the third nerve are not necessarily affected together.

In illustration of these affections I may recall the case of a man, who was under observation here, suffering from locomotor ataxia in an early stage, and who exhibited no ptosis nor distinct tendency to squint, though he complained of double vision whenever his eyes were directed much to one side, or when an object was held near the eyes. Here you observe that the branch to the levator palpebræ superioris was unaffected, and in ordinary positions of the eye there was no double vision; but when the patient looked very much to one side, the third nerve of the one eye failed to produce an action equal to that of the sixth nerve of the other, and thereby double vision was produced; and again, when the eyes were converged upon a near object, the paresis manifested itself in the same way. In the case of a man, Cowan, who was for many years under my observation, and whom some of you will remember, a similar set of symptoms occasionally existed. In 1873, for example, I find it noted that he was troubled with double vision, when he looked to one side. The same man's history affords good illustration of the more obvious conditions of ptosis and squinting.

Let me mention another case, that of a man named Garrow, who was under my care in 1874. He told us that his earliest symptom was a sudden attack of giddiness, with ptosis and squinting, which occurred in 1863; that he was for thirteen weeks under the care of Mr. Walker for this malady, and only at the end of that time got completely rid of it. It recurred in the following year, and again in 1866, each time without apparent cause. Patients sometimes say that they noticed

nothing wrong, until their friends drew attention to the drooping of the eyelid or the squint.

Now these symptoms are often premonitory of the commencement of the typical symptoms of the disease. These paralyses may, however, occur after the locomotor ataxia is well advanced, and I have known them to reappear with each attack of gastric crisis, and to linger for some time after it had passed away. Von Graefe has remarked that in these cases there is a striking diminution of the tendency to the blending of images, on which binocular vision depends; that is to say, that whereas in cases of slight double vision an effort is instinctively made by movements of the eye to bring the two images into one, in this condition the attempt is scarcely made. He suggests that this points to a central origin of the affection, which view is rendered more probable by the sudden onset and temporary character of the symptom, as well as by its occurrence in connection with gastric crises.

This group of symptoms, then, you will remember is a local manifestation of the general tendency to temporary paralyses.

## II. *Alterations of the Pupil.*

a. *Mydriasis.* This condition was observed by Eulenburg in 9 out of his 64 cases; in 3 the dilatation being double, in 4 single, and in 2 accompanied by myosis in the other eye. I have not noted this condition in any of my cases. Eulenburg states that it appears to occur mostly in the later stages of locomotor ataxia. Had it been an early symptom one might readily have explained its occurrence by one or other of two hypotheses, viz. either a paralysis of the third nerve, or an irritation of the cilio-spinal fibres. The latter is made the more likely, as Eulenburg suggests, by the circumstance that there is no defect of the power of accommodation, and that during accommodation the pupil acts, both of which facts indicate that the third nerve is unaffected. If, however, it be true that mydriasis occurs mainly in the later stages, it is difficult to understand how irritation of the cilio-spinal nerves should arise, unless in cases which have had no affection of the cilio-spinal region till a late period, and that then irritation has

preceded the paralysis. Here again is a difficulty, for were it the case that irritation always preceded paralysis, we should expect mydriasis to be a frequent early symptom of locomotor ataxia, seeing that myosis is common. Perhaps this is actually the case, and the condition is overlooked on account of the absence of concomitant symptoms.

*β.* Myosis has long been well known as a frequent eye-symptom in locomotor ataxia. It is referred to by Romberg, Remak, Duchenne and Trouseau, as well as by recent writers. Sometimes it is so marked as at once to attract the observer's attention. Eulenburg found it in 28 of his 64 cases, 21 showing double, 7 single myosis. Of my 20 cases, 7 had distinct myosis, and of these 4 had one pupil smaller than the other. The two pupils are seldom exactly of the same size. The degree of contraction varies greatly in different cases; it also varies from time to time in the same case, becoming, however, as a rule more marked as the disease advances, although not necessarily in a regular ratio. It may become very intense, and then again diminish. Thus I have seen it increase during gastric crises; and Charcot,<sup>1</sup> in his admirable lecture on the subject, states that during the attacks of lightning-pain the pupil dilates. He has also remarked that in an early stage of ataxia, it is commonly observed in inequality of the pupils, and on the side on which contraction is more marked there are sometimes evidences of vasomotor paralysis, in the reddened cheek, the congested conjunctiva, and the local elevation of temperature.

In the patient whom I now present to you, you observe extreme myosis. You notice that his ataxia is now far advanced. He often suffers from lightning-pains, and has almost constantly a broad girdle pain around the lower part of his throat. His sensibility is much diminished, patellar tendon-reflex is lost, and he has the characteristic ataxic gait, and sways about and staggers when he attempts to stand with his eyes closed. You observe that his pupils are like pin-holes, and they have been so for years. The myosis was in his case an early symptom; indeed, it was on account of dimness of

<sup>1</sup> 'Leçons sur les Maladies du Système Nerveux, Anomalies de l'Ataxie Locomotrice,' p. 56.

sight that he first sought medical advice. You will understand how such a condition necessarily makes the vision dim, even if there be no change in the optic nerve. If you will hold in front of your eyes a metal plate, or a card, with a very small aperture in it, such as might correspond in size to the pupil of our patient, you will find that although objects may be well enough defined, they appear dim from defective illumination.

Observe also in the left eye of the patient the result of the instillation of atropine. Instead of the complete dilatation produced in the normal eye, we have induced in our patient something less than medium dilatation. This modified effect has long been known as one of the characteristics of spinal myosis.

Myosis is due to disease of the cilio-spinal region of the cord. From that region there proceed nerve-fibres which join the sympathetic, and ultimately supply the radiating muscular fibres of the iris. When from affection of these cilio-spinal nerve filaments the radiating fibres of the iris are paralysed, there is nothing to oppose the action of the circular fibres, which by their contraction produce myosis. The pupil-symptoms of certain cases of thoracic aneurism are explained by pressure on the cilio-spinal fibres.

γ. Associated with the myosis, but sometimes occurring independently of it, is another curious feature, which was first described in 1869 by Dr. Argyll Robertson,<sup>1</sup> and which for convenience we are accustomed at the bedside to call "the Argyll-Robertson symptom." It consists in the absence of any contraction of pupil on exposure of the eye to light, while movement with accommodation is normally retained. This circumstance had not attracted attention until it was described by Dr. Argyll Robertson; and even now it is less known than its importance deserves. It may be well that I quote to you the description as it was originally given.

"That although the retina be quite sensitive, and the pupil contract during the act of accommodation for near objects, yet an alteration in the amount of light admitted to the eye does not influence the size of the pupil."

<sup>1</sup> 'Edin. Med. Journ.' Dec. 1869.

It is much more difficult to find a satisfactory explanation of this fact than of the myosis. Dr. Argyll Robertson suggested an hypothesis that the contraction of the pupil to light is not entirely due, as has been generally supposed, to contraction of the circular fibres of the iris produced through the third nerve but in part, at least, to temporary arrest of action in the radiating fibres of the iris, effected through the cilio-spinal filament, in the sympathetic. This hypothesis demanded that a function be assigned to these last-named fibres which is not known to exist in any other nerve. The considerations which induced him to hazard the hypothesis were the circumstance that, in the cases of spinal myosis which he recorded, the patients retained good vision, indicating a sensitive condition of the retina, normal conducting power in the optic nerve, and healthy state of the brain centre connected with visual impressions; further, that in them the contraction of the pupil associated with the accommodation indicated a healthy state of the third nerve and circular fibres of the iris, and yet, with an apparently complete chain of connection between the sensitive retina on the one hand, and the contractile fibres of the iris on the others light did not affect the pupil. The one pathological condition which was evidently present was paralysis of the cilio-spinal filaments in the cord, to which, therefore, he ascribed a share in the mechanism of pupil contraction under light. According to this view, one lesion seems to explain both the myosis and the Argyll-Robertson symptom.

But it was at once apparent that if this hypothesis were correct, myosis and the Argyll-Robertson phenomenon must always coexist, and I looked anxiously for evidence as to whether this was so or not. The first case in which I found them dissociated was one recorded by Wernicke, in Virchow's Archiv for 1872, vol. lvi. The patient was the subject of chronic alcoholism, and his pupils were of normal size, the right larger than the left; the former measuring 2 lines in diameter, the latter  $1\frac{1}{2}$ . The right reacted to light, coming down to  $1\frac{1}{4}$  lines; the left did not react to light, but both acted with accommodation. Now it is true that in this case the pupil which manifested the phenomenon was smaller than the other, but it was not by any means very small. Since that time I

have met with at least one well-marked instance in my own practice in which the pupils were of normal size and were equal, but the phenomenon was distinctly present. It thus appears that this suggestion does not afford the desired explanation.

Wernicke and Hempel adopt the view that the nervous connection between the optic and oculomotor nerves is interrupted,<sup>1</sup> and consequently the reflex irritation of the oculomotor can no longer occur, but the centre of the third is not destroyed. It appears, on the whole, most probable that the movements of the iris on exposure to light, and those for accommodation, are controlled by different nerve centres, and that in this disease the one centre and not the other is affected. Further anatomical and physiological investigations of course may be expected to clear up this question; and I have some hope that certain preparations in my possession may contribute to this end.

As to the frequency of the occurrence of this phenomenon, I know of no statistical statement, but of my 20 cases it was present in 8. In 7 of these it was associated with myosis; in 1 not, the pupils being in that case of normal size. It is not present in every case of myosis. It seems not improbable that its absence in such cases may depend upon its being a later symptom in the order of development.

### III. *Amblyopia and Amaurosis.*

Dimness of sight is also a very common symptom, but it rarely goes on to complete blindness. Besides the degree of blindness arising from the cutting off of rays by the contracting iris already referred to, we find in some cases another form of dimness, temporary and apparently functional, and corresponding to the temporary paralyses so often seen in the earlier stages of the disease. I found, for example, that a man who was under my care in 1872 had suffered from an attack of dimness of vision eight years before, and another eighteen months before I saw him, both attacks being temporary and unexplained. In 4 out of my 20 cases similar facts are noted.

<sup>1</sup> Graefe's 'Archiv,' 1876, vol. xxi., Part. I., p. 22.

It seems safe to conclude that such temporary seizures depend, as do the temporary muscular paralyses, on some undetermined condition.

But far more important is the permanent and slowly advancing amblyopia. Eulenburg found it in 31 of his 64 cases, and it was present in 14 of my 20 cases. In 7 of the 14 it appeared to be one of the earliest symptoms; in 5 of the cases there was distinct atrophy of the optic disc.

This condition is well illustrated in the patient before you. The sight of his right eye has been gradually diminishing, and is now quite lost, so that he cannot distinguish light from darkness. The sight of the left eye is also impaired. The ophthalmoscope reveals in both of the discs marked atrophic change, most advanced, of course, in the right. The features of this condition are admirably described by Charcot and sketched by Erb. I quote the description given by the latter (Ziemssen's *Cyclopedia*, Eng. Trans. vol. xiii. p. 579).

"But the atrophy of the optic nerve, which is unfortunately very frequent in tabes, is of much more importance than the disturbances of the visual apparatus thus far considered, and is much harder for the patient to endure. It belongs to the saddest complications of this already melancholy disease; the helplessness caused by the ataxy is aggravated beyond all measure by the blindness added thereto, and the sources from which the patients can obtain comfort and oblivion amid their suffering are materially diminished.

"The trouble begins with slowly or rapidly advancing diminution in the sharpness of vision, which soon increases to amblyopia, and finally to amaurosis. The field of vision, which is at first slightly veiled and cloudy, becomes increasingly narrowed, generally from without inward, sometimes from without and above, sometimes more from below. Examination shows that this narrowing of the field of vision does not take place in a uniform manner, but with entering angles. Finally, the blind area involves all but a spot towards the inside, which allows the patient but a limited and insufficient amount of vision. . . ."

"Ophthalmoscopic examination reveals the signs of so-called white atrophy of the optic nerve. At first there is a slight



grayish discoloration of the papilla, which gradually grows paler, and finally appears quite white and sharply outlined. The arteries at the same time show a progressive narrowing, but otherwise the retina remains quite unaltered. These changes are easily to be distinguished from the ophthalmoscopic picture of neuritis optica and congested papilla, and, according to French authors, they are so characteristic that the existence on the approach of tabes can be recognised from them alone.

“Atrophy of the optic nerve in tabes is always—with quite rare exceptions—a decidedly progressive malady, and leads, without interruption, to complete amaurosis. In different cases, it is true, this occurs with very variable rapidity; sometimes it requires only weeks, sometimes months or even years, for its accomplishment. Occasionally the malady comes to a standstill, even after it has lasted for a comparatively long time. Sometimes the trouble is limited for a considerable time to one eye, but it is far more common for both to be attacked and to become blind simultaneously, or within a short time. Modifications between these two extremes are of course manifold.”

Such dimness of sight and white atrophy of the nerve is by no means confined to locomotor ataxia, for it sometimes occurs without any central lesion, sometimes with cerebral disease, and sometimes with other spinal affections.

In illustration of this, I show you a patient suffering from antero-lateral sclerosis, who has for upwards of a year had well-marked amblyopia, with white atrophy of the optic nerve. In all these cases the anatomical change is essentially the same with that which exists in the spinal cord.

#### IV. *Colour-Blindness.*

This is another symptom, fully recognised as occurring in locomotor ataxia. You have seen recently, in Ward I., an illustration of this condition, and I shall presently show you a well-marked example. It is not so common as the other eye-symptoms. Of my 20 cases it was noted only in 3. One of them is the patient now before you. His colour-blindness has

existed for a long time, and it appears to vary from time to time. Thus, in February 1877 he could recognise green, white, pink, and black; but scarlet he called yellow. A year later he called green, dark-brown; red, black; yellow, he called light, but not white; and in February 1879 he could discern blue, yellow, and red. I now show him a series of sheets of coloured paper; the blue, he says, is white; the red is like blot-sheet, the green is drab, the pink is white, the yellow is yellow, the orange-brown is yellow, and the white is recognised as white. He is thus considerably worse than he was four months ago.

It is stated by Erb that colour-blindness can usually be demonstrated before any limitation in the field of vision, and he states that generally the perception of green is lost first, then that of red, finally that of yellow and blue.

Charcot states that the achromatopsia is characterised by loss of perception of the secondary colours, and by loss of perception of red and green; perception of yellow and blue persisting in a high degree, and for a long time. The facts I have shown you in our patient do not strictly correspond with these statements.

It is clear that colour-blindness does not necessarily accompany the amblyopia of locomotor ataxia, and we have not at present sufficient evidence to determine the question, whether it is more frequently associated with this form of amblyopia than with others. I have also no knowledge of any case in which colour-blindness alone was present, and am therefore inclined to attribute it to some peculiar distribution of the atrophic condition in the optic nerve.

I have not deemed it necessary to draw your attention specially to other eye-symptoms which have been described, because they are certainly of very rare occurrence, and, so far as I can judge, of but little importance.