Sands (H.B.)

A CASE

OF

SUDDEN MONOCULAR AMAUROSIS,

PRESENTING

UNUSUAL DIFFICULTIES IN DIAGNOSIS.

Bound cover in front

BY

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It is now fifteen years since the invention of the ophthalmoscope. Few persons not practically familiar with its use can realize the influence which its employment, during this brief period, has had in modifying previously existing views regarding the pathology and treatment of the diseases of the eye. By enabling the surgeon to examine the crystalline lens and vitreous with a degree of minuteness hitherto unattainable, and by rendering distinctly visible the parts constituting the fundus oculi, which had previously been concealed from direct observation during life, it naturally led to the adoption of an entirely new system of classification of the more deeply seated ocular diseases—one in which vague conjecture was replaced by scientific precision. By the joint labors of a multitude of earnest investigators, a great variety of morbid appearances were soon detected and described, and their true nature made evident in the light of pathological anatomy. In the description and diagnosis of disease, refinements so nice as to excite a feeling of ridicule in the minds of the uninitiated found their justification in facts capable of easy and convincing proof. The gain to science was immense, and few persons at the present day will be found willing to dispute it.

As has happened before, however, in the case of the stethoscope, the microscope, and other valuable inventions, the new

instrument failed to afford all the information that was demanded of it, and we are yet compelled to retain the unsatisfactory and unmeaning term "amaurosis" in the nosology of ophthalmic medicine. This is not owing to any defect in the instrument itself, but simply to the fact that its application is restricted to the interior of the eyeball; consequently important lesions affecting the sense of sight may exist in the optic nerve, or in the brain, which the ophthalmoscope is unable to reveal. Yet even here the more deeply seated disease generally leads to ultimate changes in the optic papilla and the retina, the observation of which, by the aid of the ophthalmoscope, is often sufficient, if taken in connection with the history of the case, to establish a correct diagnosis.

These remarks may appear trite, but they seem naturally to precede the narration of a case in which the evidence afforded by the ophthalmoscope, although valuable, proved insufficient to explain all the symptoms presented by the patient under observation.

On the 13th of November, 1865, I was consulted by a medical friend, who told me that while dressing himself a few hours previously he had suddenly felt his left eye "dazzled" by the light of the sun. Presently he noticed that the sight of that eye was obscured, but supposing that the dimness would soon pass off, he immediately bathed his eyes in tepid water. Within fifteen minutes, however, from the time he first discovered the imperfection of his sight, vision of the left eye was entirely abolished. No pain attended the attack. He was certain that both eyes had been normal until that morning, as during the two or three days immediately preceding he had used the affected eye in looking through the microscope. The patient was twenty-seven years of age, and, with the exception of an attack of hæmoptysis, which had occurred eight years previously, he had enjoyed uninterrupted good health. He had never suffered from rheumatism.

On examination I found as follows: *Right eye*, normal in external appearance; vision perfect. *Left eye*, pupil considerably dilated when both eyes are exposed to the light, and greatly so when the right eye is closed; feeble contraction takes place only when light is admitted into the sound eye.

Perception of light almost entirely absent, although patient can just discern the concentrated light thrown into the eye by means of the ophthalmoscope; tension normal.

Ophthalmoscopic Examination, 12, M., five hours after the attack. *Right eye*, normal. *Left eye*, media transparent; branches of central artery very small, being scarcely traceable beyond the papilla; veins hardly, if at all, diminished in size; at centre of macula lutea a circular disk, of a deep red color, sharply outlined, having about one-third the diameter of the optic papilla; the latter very pale and anæmic, contrasting strongly with that of the healthy eye; between the optic papilla and the macula lutea another deep red spot, resembling the first, but of nearly double the size, and having an irregularly oval shape, the long diameter situated in the horizontal median plane. The appearances thus described were also seen by my friends, Drs. Noyes and Althof. Careful auscultation of the chest failed to discover the least sign of cardiac disease, the presence of which, indeed, there was no good reason to suspect. The patient was directed to remain quiet, in a dark room, and to have six leeches applied to the left temple.

At 7 P.M. I saw him again, in consultation with Prof. Schweigger, of Berlin. Meanwhile, and, according to the patient's statement, before the application of the leeches, a decided improvement in vision had taken place, and he was able to count fingers at 2′ in several parts of the visual field. The latter, however, showed various interruptions, and central vision was absent. *Ophthalmoscopic examination:* Arterial vessels of the retina restored to their natural size; veins slightly turgid; retina gray and cloudy, especially around the macula lutea, where the red spot formerly mentioned still exists. The larger red spot near the nerve also remains the same. Patient ordered to have the eyes bandaged.

Nov. 15. Hardly any change. The visual field exhibits, beside other interruptions, a large central scotoma, about 7″ in diameter, at 1′ from the eye. Latterly, patient counts fingers at 3′, except when held above and to the right side. Ophthalmoscopic appearances nearly the same. The whole retina, however, is more decidedly cloudy, and conceals the choroid from view. The cloudiness is most marked in the

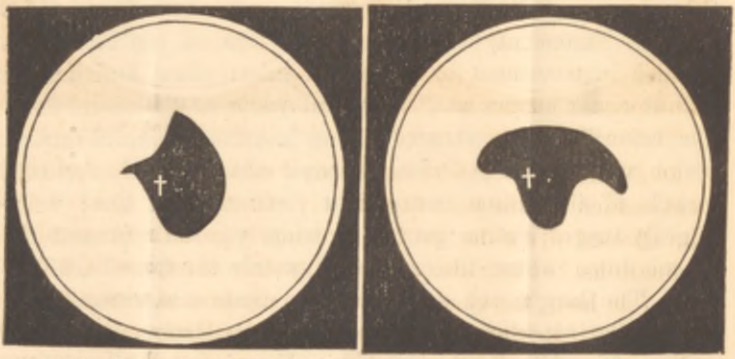
neighborhood of the macula lutea, toward which numerous fine vessels are seen to converge. The two red spots still quite distinct.

Nov. 17. No change. Ordered potass. iodid. gr. v., ter in die.

Nov. 21. Vision a little improved, the central scotoma being smaller. The ophthalmoscope shows a nearly transparent retina, and also, for the first time, a swollen, reddish, infiltrated and dirty looking optic nerve, the contour of which is hazy and ill defined. Removed bandage from the eyes, and substituted smoke glasses.

Nov. 24. Still slight improvement in vision. Central scotoma a little smaller, extending principally to the right side of the centre of the visual field. *Ophthalmoscopic examination:* Retina nearly transparent; red spots less distinct; optic papilla still red and indistinct. Allowed patient to walk out, wearing the glasses. Iodide of potassium to be continued.

Dec. 5. Signs of neuritis disappearing, the nerve looking abnormally white, with irregularly notched margin. It is needless to give further dates of examination, except such as relate to the projection of the visual field. The accompanying

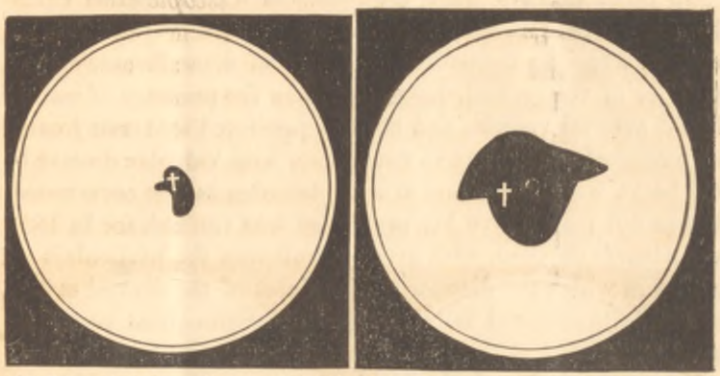


Limit of central obscuration, Dec. 5, 1865. Limit of central obscuration, Dec. 18, 1865.

figures represent the form and relative size of the central obscuration, which, it will be observed, diminished up to December 26; subsequently it again grew larger, and now has nearly its original form and dimensions. Its longest diameter, when measured upon a blackboard held at the distance of twelve inches from the eye, is seven inches.

Within the dark spot vision is wholly absent; eccentric vision, however, does not seem to be materially impaired. An ophthalmoscopic examination made a few days ago revealed the ordinary signs of atrophy of the optic nerve, which had a whitish, fibrous, glistening appearance, but was slightly, if at all, depressed. Both the arteries and veins were greatly reduced in size, the former being scarcely perceptible. Nothing special was seen at the macula lutea.

The case above detailed is the only one of the kind that has come under my observation, and possesses a singular interest, I think, from the obscurity which attended the diagnosis. I have preferred giving at first a simple statement of facts, reserving until now the reflections which they suggest.



Limit of central obscuration, Dec. 26, 1865. Limit of central obscuration, May 1, 1866.

In the first place, there can be no doubt that the loss of sight was sudden, as the patient not only remembered to have used the affected eye a day or two previously in microscopical examinations, but had also observed, within the space of a quarter of an hour, the change from mere dimness of vision to total blindness. In the next place, it may be fairly assumed that, when one eye is sightless and the other perfect, the lesion must be situated either in the eyeball itself or at some point between the latter and the commissure of the optic nerves. Any lesion involving the commissure, the optic tracts, or the tubercula quadrigemina, would, for anatomical reasons, necessarily affect both eyes at the same time. A similar statement may be made in regard to sudden blindness coming on after sexual excesses, long fasting, exhausting diarrhœa, sudden and copi-

ous hemorrhage, etc.; in all these instances both eyes are simultaneously involved. Among the causes of sudden blindness situated within the eyeball itself, if we exclude cases of injury to the cornea, iris, and crystalline lens, the most frequent are detachment of the retina and hemorrhage into the vitreous, either from the vessels of the ciliary processes, or of the choroid and retina. Here, too, the diagnosis is easy, and the difficulties are at once cleared up by the ophthalmoscope. Finally, we meet with instances in which the refractive media are entirely transparent, and in which the retina shows no structural alteration sufficient to account for the total and almost instantaneous loss of sight. To this category belongs the case now under discussion.

In 1856, Virchow[1] gave the results of a post mortem examination of two women, who had died of puerperal fever, in each of whom he discovered evidences of acute disease of the interior of the eyeball, depending upon the presence of emboli in the arterial vessels; and he then predicted that many cases of amaurosis, occurring in connection with valvular disease of the heart, would find their true explanation in this occurrence. It was not long before his prediction was verified, for in 1859 von Graefe detailed, with great minuteness, the particulars of a case in which the diagnosis of embolus of the central artery of the retina seemed fully established. Subsequent post mortem examination proved the accuracy of this opinion, and some among us have been fortunate enough to see the beautiful specimen prepared by Dr. Schweigger, in which the embolus can be distinctly seen, completely occluding the central artery of the retina, just behind the plane of the lamina cribrosa. Since the date of von Graefe's publication, at least ten[2] additional cases

[1] Gesammelte Abhandlungen, pp. 539, 711.

[2] Blessig. Archiv. für Ophthalmologie, Bd. 8, Ab. 1, S. 216.
Schneller. Ibid., S. 271.
Liebreich. Deutcher Klinik., No. 50, 1861.
Pagenstecher. Mithcilungen aus der Augenheilanstalt zu Wiesbaden, No. 2, p. 275.
Just. Klinische Monatsblätter, Juni, 1863.
Saemisch. Ibid., Janv., 1866.
Hirschmann. Ibid.
Quaglino. Ophthalmic Review, April, 1866.
Quaglino. Ibid.
Fano. Annalés d'Oculistique, t. lii., p. 239.

have been recorded in foreign journals, in which the diagnosis of embolus was pretty clearly made out. The symptoms of the disease are tolerably uniform. Sudden blindness, with subjective luminous sensations, announce the invasion of the disorder. At this period, on ophthalmoscopic examination, the media are found to be transparent, the optic papilla pale, and both the arteries and veins of the retina reduced to an extreme degree of tenuity. After the lapse of some days or weeks, the vessels may partially regain their previous size, especially towards the periphery, probably from the development of a collateral circulation. The retina exhibits an opalescent opacity, especially near and at the macula lutea, the centre of which, however, is usually marked by a deep red spot, which, by some observers, is looked upon as an extravasation of blood, while by others it is ascribed to the natural color of the choroid, seen through the retinal substance (which, in this situation, is exceedingly thin), and contrasting with the grayish opacity of the surrounding infiltrated retinal tissue. The final result is usually atrophy of the optic nerve and retina, with nearly total loss of sight; and when vision is partially restored, it is generally limited to the eccentric parts of the visual field.

Now, in the case I have recited, although many of the symptoms above mentioned were present, I think we must reject the diagnosis of embolus for the following reasons: 1st. The absence of any evidence of cardiac disease. This alone, however, would not be decisive, provided all the other symptoms were present. 2d, and principally, because the phenomena of the retinal circulation, as revealed by the ophthalmoscope, cannot be reconciled with the theory of complete and permanent arterial obstruction, such as would be caused by an embolus. In every reported example of this affection, it has been noticed that both the arteries and the veins of the retina were greatly reduced in size. This is what we should expect, *à priori*, seeing that an embolus, while it would prevent the blood from entering the central artery and its branches, would in no way interfere with the return of the blood through the veins, which would, consequently, also be empty. But in the present case, the *arterial branches alone* were anæmic, while the veins were filled with blood. Furthermore, if the two red

spots above described are to be regarded as extravasations, it may be inferred that the hemorrhages were the result of increased pressure on the walls of the veins, due to their congestion. That the red spot at the macula was hemorrhagic in character is, to my mind, evident, not only from its appearance, but also from the fact that throughout the whole course of the disease the total absence of central vision pointed strongly to the existence of a lesion at the yellow spot. The other and larger red spot had also exactly the appearance of an extravasation, and its situation was such that it could hardly be regarded as the effect of contrast. Finally, it is to be observed that the symptoms of arterial obstruction were of but few hours' duration. The arteries which, at noon of the day of attack, were almost invisible, at 7 o'clock in the evening of the same day were seen by Dr. Schweigger and myself to be of their normal size. This fact would be difficult to explain upon the supposition of embolism.

The theory which appears to me to explain most satisfactorily the morbid phenomena in question, is that which assumes the primary lesion to have been an extravasation of blood into the sheath of the optic nerve, between the globe and the optic commissure. Such a lesion has not, so far as I am aware, been demonstrated by dissection, but several instances have been recorded in which the symptoms rendered its existence highly probable.[1] Hemorrhages in the substance of the retina and at the base of the brain are by no means rare, and it is quite possible that the same lesion may occur in that commissure which we call the optic nerve. My own solution of the present case would be as follows: at first, hemorrhage within the sheath of the optic nerve, compressing suddenly the nerve fibres, the central artery and vein. Hence, the empty state of the arteries, the fullness of the veins, and the general œdematous infiltration of the retina. Hence, also, total blindness, due partly to insufficient arterial supply, and partly to a diminution in the conducting power of the optic nerve fibres. Next, hemorrhages into the retina, owing

[1] Hutchinson. Ophthalmic Hosp. Reports, vol. iv., p. 237. Pagenstecher. Op. cit., H. 1, p. 54.

to mechanical congestion of its veins. Later in the day, diffusion of extravasated blood, in consequence of which, the pressure being lessened, the arteries again filled with blood, and the retina again resumed its function, except centrally, where vision remained absent in consequence of hemorrhage at the macula lutea. Lastly, inflammation of the optic nerve, with its usual termination in atrophy. During the progress of the neuritis, I thought that the red discoloration of the optic papilla might be owing to the presence of extravasated blood, which had found its way along the sheath to the surface; but I was not certain that I saw any thing more than the simple redness of hyperæmia.

I have spoken of hemorrhage *within* the sheath of the optic nerve, because if blood had been effused external to the sheath in sufficient quantity to have caused the symptoms mentioned, it would necessarily, at the same time, have exerted pressure upon the other nervous filaments traversing the orbit, and would have given rise to pain, strabismus, and other morbid phenomena, which were not present.