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CLASSIFICATION OF DISEASES OF THE CHOROID*

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THE nomenclature in relation to diseases of the choroid has always appeared to me most unsatisfactory, and judging from the literature both in well-known text books and in published papers, as well as from verbal descriptions, there appears to be no settled opinion as to the aetiology and course of the various pathological processes which attack the choroid.

The classification and description of the different conditions are still based on the picture displayed to ophthalmoscopic examination—surely a relic of the past—a fact which casts a distinct slur on all the admirable pathological work which everyone acknowledges as one of the most brilliant advances in the science of ophthalmology during the last twenty or thirty years.

I think the time has come when fundus diseases ought to be classified according to the structure in which they occur, either retina or choroid, and, as both these structures are present in all parts of the fundus, there is no justification for describing diseases of the fundus in terms of position, e.g., macular diseases, choroiditis juxta-papillaris, etc.

The results of classification by position alone are that the terms

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retino-choroiditis, myopic choroiditis, choroidal atrophy, degenerations, holes, etc., are applied indiscriminately to all sorts of diseases, both old and recent, regardless of their pathological meaning, and of whether they actually originate in the choroid or in the retina.

In Rayner Batten's papers in 1921 and 1923 read before the Oxford Congress, containing a most remarkable display of ophthalmoscopic drawings, this confusion of thought is very obvious, for we find Tay's choroiditis, retinitis circinata, choroidoretinitis, and amaurotic family idiocy, all put into a group together under the heading "Macular Diseases."

I propose in this paper only to deal with diseases of the choroid and to omit all reference to the retina except in so far as it is secondarily affected. At the outset it is well to bear in mind that all such conditions are associated with pigmentary disturbance, either proliferation or migration, whereas in solely retinal affections no such changes occur. This pigmentary proliferation or migration varies very much in different cases, and is only a question of degree; but it is almost always present to indicate the structure in which the initial process has taken place.

An enormous variety of choroidal affections are described in some text books, and in "The American Encyclopaedia of Ophthalmology," Vol. III, I find no less than 26 different forms of choroidal inflammation alone which seem to require separate headings. No wonder there is confusion in the mind of the reader. The fact that the picture of a definite pathological condition differs slightly in its ophthalmoscopic and clinical details does not justify such an elaborate classification.

The classification I wish to put forward both from a pathological and clinical standpoint falls into five groups:—

1. Inflammatory.
2. Vascular.
3. Degenerative.
4. Congenital.
5. New Growths.

Both inflammatory and vascular types exhibit characteristic clinical appearances according to whether they undergo a process of resolution or not, and therefore we have in each the acute or recent stage as well as the old or scarred stage.

We see a great deal more of the latter than the former, and many of them have never been observed in the recent stage at all, but the scarred stage has been discovered by accident.

The need for discriminating between these first two groups is that in the recent stage the treatment is entirely different.

Whereas in the inflammatory form we must search for the offending organism or toxin and promote the absorption of the inflammatory exudate as soon as possible, in order to minimize the

pressure effects and also to prevent such organism from attacking the central region if it has not done so already, in the vascular type

FIG. 1.

A typical inflammatory scar, showing yellowish-white fibrous tissue with pigment proliferation. An additional proof of the inflammatory origin are the fibrous tissue bands leading from the scar to the optic disc; such bands are only rarely seen in these cases.

we must treat the vascular system in the hope of improving the choroidal circulation and thus influencing the blood supply of the affected part.

In the old or scarred stage no treatment is of any avail in either form since the retinal elements are involved in the scar tissue and have become completely destroyed and functionless.

At present I notice that the term retino-choroiditis is used to describe both acute and scarred forms, whereas if it means anything it implies an acute inflammation.

Take a typical example of an inflammatory disease of the choroid. Here we have the localized oedematous yellowish-grey swelling in

FIG. 2.

A scar produced by the affection of the choroidal vessels. Note the sclerosis of the vessels; the small amount and granular nature of the pigment, considering the size of the area involved.

any part of the fundus with an ill defined border, subsequently either breaking down and causing panophthalmitis or undergoing resolution with the formation of a scar. In actual origin these inflammatory foci are all the same although differing in size, number, and position, and the larger they are the greater is the chance that they will fail to undergo resolution. The cause is to be found in some organism or toxin which finds its way to the choroid through the blood stream from some other part of the body, and may be due to syphilis, tubercle, sympathetic disease, pyorrhoea, or any septic cause. If resolution takes place, as it generally does, pigment proliferation occurs in the affected part giving rise to the black masses

irregularly distributed over and around the original site, with which we are all familiar.

No doubt both retinal and choroidal pigment take part in this formation and the pigment is deposited anywhere in the choroidal and retinal tissues wherever there happens to be any space left free which is not obliterated by scar tissue. The original inflammatory material undergoes partial absorption or fibrous tissue formation and in this way is produced the yellowish white scar surrounded by

FIG. 3.

A degenerative lesion. Note the yellowish-white mass with perfectly even pigmented border and the absence of proliferation. Although situated at the macula, the vision of the eye was 6/6.

masses of pigment, which gives rise to the typical ophthalmoscopic picture.

The white areas are sometimes called atrophic, meaning presumably that the scar tissue has contracted, thus leaving a gap in the choroid which results in exposure of the sclera.

This is the only sense in which the term choroidal atrophy can be used, and no doubt it does sometimes occur, as when vessels are seen crossing the floor of the scar, but it is certainly by no means always the case, as is shown by many pathological sections, and is probably far less common than the continual use of the term "atrophy" justifies. I believe that in the majority of cases the white appearance we see with the ophthalmoscope is due to fibrous tissue.

The clinical features associated with inflammatory diseases of the choroid are vitreous opacities, and sometimes keratic deposits in the acute stage, with scotomata in the visual field of different shapes and extent which remain permanent in the scarred stage.

The acuity of vision is suddenly reduced to considerably below normal, but may recover completely when the inflammatory exudate has become absorbed, unless the macula happens to be the part involved, when there is less likelihood of a lasting or permanent improvement.

FIG. 4.

A coloboma. Notice the pearly white appearance of the scar, with a narrow border of fine pigment; vessels are at the edge of the gap only.

Contrast this with a typical case of choroidal affection due to vascular changes—primary pigmentary degeneration of the retina.

I am quite aware that I am here treading on controversial ground as all observers do not hold this view, but it must be remembered that any theory based on a neurological origin must become a weak one unless it can account for the migration of pigment which in the large majority of cases accompanies this condition. The pigment epithelial layer, being originally an epiblastic structure, at first belongs to the retina, but in post-natal

life it becomes so intimately connected with the choroid that it practically forms part of this structure; and therefore is not affected in diseases of the retina alone. Personally I cannot quite follow the argument that perfectly healthy pigment and pigment epithelial cells should wander away from their normal position merely because their anatomical boundaries in front are destroyed, as suggested by Collins in his theory of abiotrophy (*Trans. Ophthalm. Soc. U.K.*, 1919), unless it is shaken out by concussion or washed out by haemorrhage; otherwise we should expect to find it in other forms of atrophy of the retinal elements.

Now in primary pigmentary degeneration of the retina we have the typical bone-corporuscle-shaped arrangement of pigment in the mid-peripheral region scattered over an otherwise perfectly normal looking fundus, and the cause has been generally acknowledged on experimental and pathological grounds to be found in the altered condition of the choriocapillaris.

We meet with cases where the bone-corporuscle-shaped pigment is scattered over a fundus which is not normal, and where evident changes in the choroid have taken place previously, resulting in the larger vessels becoming affected. In this way the choroidal blood supply is cut off, and the same effect is produced on the retina as in primary pigmentary degeneration. These are cases of pigmentary degeneration secondary to disease of the choroid. No matter in what way, or in what part, the choroidal vessels are constricted, the results are the same. The term retino-choroiditis is commonly applied to this condition. There is no pigment proliferation, nor excess of pigment in these vascular cases, but the normal amount of pigment wanders into abnormal positions and assumes different formations according to the part of the retinal tissue in which it is found and the channels through which it is carried.

The clinical signs and symptoms of this condition are familiar to all, but in these cases vitreous opacities are uncommon, no keratic precipitates are ever seen and the effect on vision is extremely variable, depending upon the amount of degeneration in the neuro-epithelial layer which cannot be seen ophthalmoscopically but can only be discovered by the effect on the visual field.

A haemorrhage from the choroidal blood vessels will sometimes be so extensive as to wash out the pigment mechanically from the pigment epithelial layer, when it is seen indistinctly mixed up with the blood. When the latter becomes absorbed these small masses of pigment are left high and dry, giving a granular appearance to the disturbed area, but there is no proliferation.

In degeneration, the most typical one being the ophthalmoscopic pictures described as Tay's choroiditis, the pigment is merely pushed aside by the hyaline masses. We see no proliferation of pigment but only small white or yellowish-white patches with a

perfectly even pigmented border. The regular circular arrangement of this pigmented margin is to my mind very characteristic. In these cases again the degenerative patches may be of different sizes, and greater or less in number, presenting a variety of ophthalmoscopic pictures.

We know now that the so called Tay's choroiditis is hyaline degeneration of the membrane of Bruch. Usually this choroidal disease does not seriously interfere with vision, exactly as we might expect.

In congenital defects, of which the ordinary coloboma is a typical example, the characteristic feature is the pearly white appearance of the exposed sclera with some vessels passing near the edge of the gap, and again the even border of pigment round the margin which is similar to the ordinary heaping of the choroidal pigment on the temporal side of the disc, but there is no pigment proliferation. If it were possible to imagine this white area being a mass of scar tissue from inflammation, such a large inflammatory deposit would have hardly undergone resolution, and, had it done so, there would have been a far greater pigmentary development than occurs in these cases.

The above principles can be applied to colobomata occurring anywhere in the fundus, though it is naturally extremely rare, for embryological reasons, to find them in any other situation.

From these preliminary considerations, it seems to me that we may draw the following general conclusions.

1. Where there is a large or small oedematous patch accompanied by sudden lowering of the visual acuity, vitreous opacities and keratic precipitates, followed by the formation of fibrous tissue of varying amount, with many coarse masses of gross pigmentation, and sometimes vessels crossing the floor of the scar, the pathological condition producing it is inflammatory in origin.
2. Where the pigmentary changes are of a fine granular type and the choroidal disturbance, though decided, is of a somewhat indefinite character, accompanied by variable visual acuity, with no vitreous opacities and no keratic precipitates, the pathological condition producing it is of vascular origin, either complete or partial cutting off of the blood supply, an old haemorrhage, etc.
3. When we find white or whitish-yellow areas, never very large and sometimes quite minute, surrounded by a perfectly even, well-defined border of pigment which is not excessive, with no choroidal disturbance around it, the condition is one of hyaline degeneration, probably of the membrane of Bruch,
4. When there is a pearly white patch with absence of vessels crossing the floor of it, and a narrow fringe of pigment round a

well-defined margin with no choroidal disturbance beyond the actual defect, the probability is that the condition is congenital in origin.

5. The characteristic features of a new growth are the raised choroidal swelling, often with a well-defined non-oedematous border, and whatever pigment there is, is indefinitely mixed up with the main mass. There is frequently an ordinary detachment, namely, separation between the neuro-epithelial layer and the pigment epithelial layer, somewhere in the neighbourhood of the growth, and, if the case can be safely watched, a gradual increase in the size of the swelling is noticed. The presence of new vessels helps the diagnosis.

I now propose to support this classification, so far as I can, by drawings illustrative of the points I have endeavoured to bring out; but it is to be noted that there are considerable differences in the actual ophthalmoscopic picture.

There is one form of choroidal affection, occurring generally at the macula, which is difficult at first to classify according to any of the above descriptions, although it belongs to the vascular type. In the early stage there is a raised oedematous swelling occupying the whole of the macular region with a good deal of outlying exudate, and possibly some haemorrhage in places. There is hardly any pigmentary disturbance, and the scarred stage shows a puckered mass of fibrous tissue at the macula with a complete or incomplete ring of organised exudate round it (so-called retinitis circinata). The characteristic features of this form are its sudden appearance and the fact that it attains its maximum size almost at once and then remains stationary for a long time before the scarred stage makes its appearance. I have ventured to call this an infarct of the macular choroidal blood vessels.*

Dr. Batten has attempted to connect many diseases of the macula with affections of the brain, which produce various mental disorders; he associates the two together as a disease of the nervous system, analogous to such cases as amaurotic family idiocy. I think this is stretching the analogy too far. Surely the vascular changes are similar in the two places and lead to similar results without any direct neurological relationship between the two.

*See *Trans. Ophthalm. Soc. U.K.*, Vol. XXXIII, p. 86, acute stage, and *Trans. Ophthalm. Soc. U.K.*, Vol. XXXI, p. 266, scarred stage.