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CONGENITAL CHOROIDEREMIA

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This case of absence of the entire choroid except in the macular area, observed by Captain Connor during his recent service in the Medical Corps in the United States Army, is of special interest because of the extremely small number of such cases hitherto reported. The circumstances under which this case was discovered indicate that they may be less rare than the reports seem to show.

On February 16th, 1918, Corp. C. B., Bat. B. 78 F. Art., came to the eye department of the Base Hospital at Camp Logan, Texas; complaining that he could not see to get about at night. Recently while on guard duty at night he had failed to recognize and salute an officer. As the bridge where the guards were stationed was over a deep drainage ditch, the soldier feared he would walk off some night and seriously injure himself.

Vision was tested with the regulation test type and found to be 20/20 in each eye. When told that his vision was normal, he replied, "Oh yes, I can see as well as anyone in the day time, but I cannot see as well as other people when it begins to get dark." This condition has existed as long as soldier could remember, eyes had never been sore and did not trouble him when he went to school. He had formed the habit at home of not going out much at night, unless some relative or friend went with him.

Patient twenty-five years old, a strong, well-developed soldier. Before entering the Army he did clerical work without trouble to his eyes. Personal history negative, not sick since childhood, thinks he had measles when six or seven years old, not very sick and does not think eyes were affected, denies venereal infection.

Parents both living and well, not related. One sister and one brother,

living and well, none dead. No other case of night blindness in family that he knew of, and none have any form of eye trouble except one uncle on mother's side had repeated attacks of sore eyes. Says uncle had been ordered to wear glasses, but he does not think eyes were benefited by their use.

Examination shows lids and conjunctiva normal—pupils equal and respond to light and accommodation, iris normal and dark brown color.

Ophthalmoscopic examination shows cornea, lens and vitreous clear, but the picture presented by the fundus is most striking. Instead of the red fundus reflex usually seen, the examining eye encounters a glistening white fundus over which the larger retinal vessels appear about normal in size and distribution, (as shown in Plate XII). The sclerotic is exposed to view over the entire fundus, except a circular area about twice the size of the disc in the macular region. In this macular area the appearance of the fundus is normal, having the normal yellowish red color. The macula shows as a slightly darker spot in about the center of this patch of normal fundus. Scattered about the fundus are a few small ciliary vessels, and a few pigment spots. Both eyes are affected in the same way and present practically the same ophthalmoscopic picture, except that the left macular area is less regular in outline than the right, the latter

having choroidal vessels extending out from it, toward the disc and below.

Altho the retinal vessels appear entirely normal the retina itself is not properly developed over the white area, as shown by the markedly contracted, field of vision (Fig. 1) which accounts for the symptom "night blindness," of which patient complains. The color fields are correspondingly contracted, the red and green having their normal relation. Both eyes are one-half diopter hypermetropic.

In looking up the literature on anomalies of the choroid, the writer was able to find only one allusion to this peculiar choroidal defect recorded—

whether this condition called choroideremia is in reality a condition separate and distinct from the more common defect of the choroid, coloboma. Is the former dependent upon different etiologic factors, or is it simply an unusual form of coloboma?

Fuchs, in discussing the cause of coloboma, attributes the defects in the choroid to nonunion of the edges of the retinal cleft.

Wm. Lang calls attention to the fact that a failure of the retinal cleft to close, would not account for those colobomata which are covered with retina. According to Lang the explanation of

Fig. 1. Boundaries of Contracted Fields of Vision in a Case of Choroideremia (Connor).

that by the English ophthalmologist, the late Nettleship. A copy of Nettleship's original article was not available, but in de Schweinitz' book "Diseases of the Eye," we find: "A striking developmental abnormality is one in which the entire choroid except a small area in the region of the macula is absent. To this condition the name choroideremia, has been given. Both eyes are affected. The patients are night blind." (Nettleship.)

From the brief description from Nettleship's paper it will be seen that the lesion and symptoms presented by other cases were identical with those of our soldier.

The question naturally arises as to

all congenital defects of the choroid, wherever situated, is the same, an adhesion forms between the developing retina and mesoblast, which latter consequently fails to become differentiated into choroid and sclerotic. If the adhesion occurs before the closure of the retinal cleft, the coloboma is devoid of retinal covering; if it occurs after the retinal cleft has closed the coloboma is covered with retina.

The retinal fissure begins to close at its posterior extremity, the closure extending forward along the under surface of the optic stalk and optic vesicle, until the cleft should be completely closed by the second month of fetal life. This development may be arrested at any stage, hence the shape

of the typical coloboma is more or less triangular, situated in the lower segment of the fundus with the apex toward the optic disc, and broadening out as it extends forward toward the ciliary body. It may involve the ciliary body, iris and lens in this region, producing coloboma of these structures as an associated condition.

Whether or not colobomata of the choroid are due to adhesions between the blastodermic membranes as considered by Lang, it is certainly true that they usually occur along the line of the optic cleft and correspond to the general shape of this cleft during the various stages of its closure, so that we must consider their etiology as at least associated with the development and closure of this fissure.

The exception to this rule is found in those rare forms of coloboma which occur in areas of the fundus, remote from the region of the cleft, of which macular colobomata are examples.

In macular coloboma there is an absence of the choroid in the region of the macula, while in choroideremia,

just the opposite is found, the macular area being the only part of the fundus which is supplied with choroid.

From the foregoing it will be seen that the lesion in choroideremia differs widely in location and extent from coloboma of the choroid; and it is not unreasonable to suppose that it has a different etiologic foundation.

The analog in embryonic life of what is later the macular area of the fundus is a spot on the outer surface of the secondary optic vesicle, where the first signs of differentiation of the mesoblast into sclera and choroid are seen, the first appearance of the choroid being manifest by a tiny plexus of capillaries at this point. Hence, in seeking an explanation of the cause of choroideremia, we must consider that differentiation of the mesoblast, of the secondary optic vesicles, into sclerotic and choroid, began in a normal way and at the normal point, but for some reason was arrested when only the macular area was supplied with choroid. [For references see p. 620.]

THE EYES OF THE SIGNALMAN

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This paper, by a scientific investigator who has had practical experience as a signal officer, is especially valuable for its coordination of the scientific and practical aspects of the subject. It was read before the American Ophthalmological Society, June 14th, 1919.

These observations are based on a two years' experience in charge of the signal bridge on the U. S. S. Granite State. During this time I had the opportunity of testing practically the ability of some two hundred men in making and reading signals. A hundred of these served on the bridge under my direct supervision, so that I could judge, by actual observation of their work and of the behavior of their eyes under active service conditions.

The relation of ophthalmology to naval signalling may be studied under two aspects.

1. What are the visual qualities required in order to carry on the delicate and important work of signalling at sea?

2. What is the reaction of the eye to work of this sort, i. e. what pathologic effects may result from the visual strain involved?

The first question alone will be considered here, since with rather extensive opportunities for observation, I have had no occasion to note any untoward effects from naval signalling.

To answer either question intelligently, we must consider: