ANGIOMA OF THE RETINA*

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ANGIOMATOSIS retinae (von Hippel's disease) is not uncommon and angiomatous formations form a part of its clinical and pathological picture. The following case is of interest in demonstrating a retinal angioma unassociated with the excessive gliosis which is characteristic of the condition described by von Hippel.

Case Report.—Mrs. A. B., aged 29 years, was seen by an ophthalmic surgeon on April 2, 1940, complaining that her glasses were not satisfactory, and that she wished to have them changed. She said her eyes ached, and easily became tired. On examination

FIG. 1.

the right vision was 6/9 with +0.50 D.Sph. + 3.00 D.Cyl. axis 75° and the left vision 6/9 with +3.50 D.Cyl. axis 75°. The right fundus was normal. The left fundus showed a grey swelling adjoining the nasal margin of the disc. Several dilated veins were present on this tumour, one being particularly distended, but the source of these vessels was not visible. Above and to the nasal side of the swelling there were about half a dozen red spots, which appeared to be retinal haemorrhages. It was diagnosed as a "congenital malformation," but when seen a week later, it had increased in size and the disc was almost hidden. The appearance

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at this time was so suggestive of a malignant growth that the eye was removed.

No observations are available to indicate any abnormality of the nervous system.

Pathological Report.—A vascular formation at the junction of the retina and optic disc, invading the optic nerve, but not the choroid. Endothelial lined channels, containing red blood corpuscles, varying in size from capillaries to vessels as big as the main branches of the central artery and vein; the supporting framework contains some glial cells.

Discussion

Angiomatosis retinae, though cases had been previously described, was established as a clinical entity by von Hippel in 1904. It shows hereditary characteristics, and is considered to originate as a developmental error. It appears "with disconcerting polymorphism" (Duke-Elder). Gourfein-Welt, Carr and Stallard, Niccol and Foster Moore, and Feig have described cases characterised by tumours localised in the region of the posterior pole of the eye, and the case described appears to belong to this group. The tumour was so close to the disc that the vessels supplying it, probably greatly swollen, were not visible. Treacher Collins and von Hippel considered that angiomatosis of the retina was, primarily, an overgrowth of blood vessels, and that retinal gliosis was a secondary manifestation, though Meller, Guzman and others believed that the condition commenced as a gliosis and that the vascularisation followed. It is generally accepted at the present time that an angiomatous growth is the initial abnormality and this case confirms the belief. It may be considered as a very early state of the condition, in which an uncomplicated angioma is present. It is probable that, if the eye had not been removed, retinal gliosis would have occurred, followed by the complete degeneration of the eye, which is, so often, the final result of angiomatosis retinae.

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