THE BRITISH JOURNAL
OF
OPHTHALMOLOGY
OCTOBER, 1928

COMMUNICATIONS

A CASE OF NEUROFIBROMATOSIS OF THE
RIGHT ORBIT*

BY

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NEUROFIBROMATOSIS affecting the eyelids is a rare condition, affecting the orbit a very rare condition, so that, although a very good account of the condition with illustrative cases can be found in the Transactions of the Ophthalmological Society of the United Kingdom for 1903 and 1905, in papers by Snell and Treacher Collins, the present case has been thought to be sufficiently interesting to report fully.

A. W., a male, aged 26, came to the Royal Westminster Ophthalmic Hospital in March, 1926, as his right eye was worrying him. He had not been to any hospital since he was taken to Moorfields in 1911, when Mr. Hancock excised a portion of the right upper lid for diagnostic purposes. He stated that the lid became enlarged when he was about eight years old. Mr. Whiting kindly sent us the pathological report from Moorfields, proving the diagnosis.

As shown in the photograph (Fig. 2) the right upper lid was enormously thickened and overhanging. Besides producing great

* A paper read at the meeting of the Section of Ophthalmology, Royal Society of Medicine, June 8, 1928.

deformity it also kept the skin of the lower lid sodden, and frequently caused ulceration in this situation. There was a similar condition of thickening of the skin and underlying tissue in the right temporal region, and a small patch in the right occipital region. There was no affection of any other part of the body. The right eye was blind, the cornea being quite opaque with much surface vascularisation. The tension was normal, and there was no buphthalmos. The left eye and lids were healthy. L.V. 6/9.

There was a considerable amount of conjunctival discharge, and, in spite of regular treatment for some months, including a period of daily attendance at the hospital for irrigation of the eye, it was found impossible to cure the conjunctivitis that existed or to prevent the constant recurrence of ulceration of the lower lid. It was thought that a partial operation consisting of excision of a portion of the upper lid and underlying growth would not sufficiently relieve the condition, and that as the right eye was blind, it would save the patient's time and money if the eye were removed and the socket obliterated. The patient agreed to this procedure, and in November, 1926, the eyeball was removed with a large part of the right upper lid, the edge of the lower lid, and the conjunctiva of the socket, and the cut lid margins were sutured together. Recovery was uneventful and the photograph taken two months later shows the improvement in the patient's appearance. He has had no further trouble with ulceration or discharge.

The pathological report by Dr. Hervey Wyatt and the accompanying microphotographs are of considerable interest owing to the rarity of the condition.

PATHOLOGICAL NOTES BY R. B. HERVEY WYATT

The specimen consists of a portion of the right upper and lower lids, the right eyeball, and surrounding orbital tissue. These structures were bisected vertically. The temporal half was mounted as a naked-eye specimen and the remainder has been used for making microscopical sections.

Gross appearances

The upper lid, which is enlarged in every direction and much thickened, is slightly everted at the inner side of its free border. It completely covers the palpebral aperture and the lower lid and hangs down over the cheek. It is tough and nodular to the touch. The skin covering it is healthy in colour, but several dilated veins are to be seen on the surface. The skin between the upper lid and the eyebrow bulges forwards and is stretched downwards so that

it covers the eye. The skin to the outer side shows an irregular subcutaneous swelling. The eyeball is depressed downwards towards the floor of the orbit. The under surface of the lid shows chronic inflammation and this is communicated by constant contact to the lower lid which is ulcerated. In the temporal region there is an irregular nodular subcutaneous swelling extending from the outer angle of the orbit towards the ear and upwards over the frontal bone in the area of the temporal artery.

Microscopical appearances

The tissue was fixed in formalin, embedded in paraffin wax, and the sections were stained with haematoxylin and eosin.

Sections of the Upper Lid.—The overlying epithelium is slightly thickened and the papillae are prominent especially at the margin of the lid. There is extensive hypertrophy and thickening of the fibrous tissue of the corium. Beneath the skin and in the neighbourhood of the Meibomian glands are extensive areas of small round-celled infiltration and a few giant cells. The blood vessels are numerous and very congested and dilated.

In the subcutaneous tissue are numerous well-defined areas of varying shape and size which are composed of thickened fibrous tissue from the nerve sheaths.

The Meibomian glands which are widely separated by hypertrophied fibrous tissue are irregular in shape and size and contain degenerated and vacuolated cells. (Fig. 1).

The Lower Lid.—This shows an increase of fibrous tissue and an inflammatory reaction beneath the epithelium. Thickened nerve fibres were not seen.

The Eyeball.—Measurements : antero-posterior 25 mm., vertical 23 mm., lateral 24 mm. The cornea measures vertically 9.5 mm., and laterally 10.4 mm.

The optic nerve is embedded in a mass of fibrous tissue. After section the disc is seen to be cupped.

Cornea.—The epithelium is regular and thin with rather flattened cells. The fibres of the substantia propria are also rather atrophied and thin. Immediately beneath the anterior limiting membrane are groups of elongated cells forming an almost complete layer. At the limbus these cells become much more distinct, and numerous thickened nerves are present. I think these cells are hypertrophied nerve-supporting tissue. Descemet's membrane is well defined.

Iris and Ciliary Body.—The iris is short and thick. There is a suggestion of ectropion of uveal pigment. The stroma is oedematous and the vessels somewhat dilated. The ciliary body

FIG. 1.

FIG. 2.

FIG. 3.

FIG. 4.

shows dilatation and engorgement of the vessels, otherwise it is normal. (Fig. 3).

The Retina.—Rod and cone layer much degenerated. The other layers are distinguishable.

The Choroid.—Rather thick and densely pigmented. The blood vessels are dilated and engorged. I was not able to see, however, any hypertrophied end organs as described by Treacher

FIG. 5.

Collins in his paper on this subject in the Transactions of the Ophthalmological Society, Vol. XXV, 1905.

Optic Nerve.—The disc is cupped (Fig. 4). The fibrous tissue of the nerve sheath is increased while the nerve elements are decreased with a kind of myxomatous degeneration. Mingled with the optic nerve is tissue similar to that found affecting the other nerves. Outside the dural sheath many sections of the ciliary nerves show this same fibromatosis. This is also seen where the nerves perforate the sclerotic.

The orbital tissue (Fig. 5) contains large numbers of affected nerve twigs.