MELANOMA OF THE OPTIC DISK*

BY

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MELANOMA of the optic disk is rare so that the opportunity of making a histological examination seldom occurs.

Historical.—Michail (1934) summarized the literature referring to 42 cases of pigmentation of the optic disk and added reports of four examples. Of these 46 cases, six had been studied histologically. François (1934) also gave a full résumé of the literature. Brief mention may be made of three of these reports, which have points of particular resemblance to the present case.

Liebreich (1863) illustrated a pigmented optic nerve head in which the point of emergence of the retinal vessels was enclosed by intense black streaks arranged in bundles covering almost one-third of the disk. Hirschberg (1883) illustrated the striated appearance of the tumour in his case, which was, however, of a reddish colour. Pálích-Szántó (1915) also remarked on striation comparable to that of opaque nerve fibres.

Few reports include details of the visual fields, but Pálích-Szántó mentions the absence of any defect. Oguchi (1933), on the other hand, found a peripheral field loss, but his case (not included in the reviews mentioned above) was shown to be a melanotic tumour occupying the nasal half of the optic disk itself, which he considered to have arisen on the basis of a congenital melanoma.

Levine (1935), Fry and de Long (1942), Loewenstein (1945), Paulo Filho (1945), Cordes and Hogan (1948), Kreibitz (1949), Scudéri (1949), and Vetter (1950) described pigmented tumours of the optic disk, and Sedan and Halbron (1948) described four cases of pigmentary anomalies of the disk, two of which showed dense black patches with striated edges. Laje Weskamp and Páez Allende (1950) reported a melanoma of the optic disk which they considered benign. Moehle (1945) described complete pigmentation of an optic disk, which did not change over a period of 6 years; he considered the pigmentation to be congenital.

Case Report

T. A., a miner, aged 45, attended the Royal Infirmary, Edinburgh, on September 16, 1950, complaining that for a few weeks he had noticed occasional blurring of vision in the left eye, particularly when reading. The left eye "had always been weaker than the right". He later said that, 2 months before, he had had an injury caused by a piece of coal, but it seems that this had been trivial.

Examination.—He appeared to be in good general health. There was congenital torticollis, for which an operation had been performed at the age of 22, and there was facial asymmetry.

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Left Eye: visual acuity was 6/9 (partly) with +2.5 D sph. and J. 2 with +1.25 D sph. added. The iris was uniformly brown, the pupil reacting normally. The media were clear and there was no evidence of inflammation.

The appearance of the fundus of the left eye was striking. The optic disk was completely obscured by a feathery mass of dark brownish-black pigment which appeared to be in the superficial layers and showed radial striation. The appearance was aptly compared to that of opaque nerve fibres except that the colour was very dark instead of white. Centrally there was a slightly paler triangular depression. Passing outwards from the dark mass, mainly to the temporal side and inferiorly, there was an ill-defined slightly yellowish-grey area, suggestive of a solid detachment of the retina, shading off into the normal background. The retinal vessels passed outwards from the dark mass, being concealed centrally. The summit of the elevation obscuring the disk was estimated to be 3.5 D higher than the surrounding background. Elsewhere the fundus had a normal appearance. The tension of the eye was normal. Perimetry demonstrated a considerable enlargement of the blind spot and reduction of the field for a 10/2,000 red test object with involvement of the fixation area. The optic foramen was radiologically normal.

Right Eye: The eye appeared to be healthy and the visual acuity was 6/5 with +1.5 D sph. and J 1 with +1.0 D sph. added.

In the general examination of the patient no melanotic areas of skin were noted, the liver was not enlarged, and there was no glandular enlargement in neck, axillae, or groins.

FIG. 1.—Appearance of the left fundus on January 17, 1951. (Unfortunately the outline of the choroidal tumour does not show up in this monochrome reproduction of the original coloured drawing of the fundus.)

The urine showed neither albumen nor sugar. The blood Wassermann reaction was found to be positive +++ and the Kahn test +; the patient was therefore referred to Dr. R. C. L. Batchelor who has kindly made available the results of his investigations. He found that one sister of the patient had suffered from congenital lues. Examination of the patient's cerebrospinal fluid showed cells 2 per cu. mm., protein 70 mg. per cent., globulin "a trace", Wassermann reaction negative, Lange gold curve 0000000000. He considered that the patient had been congenitally infected with lues and he carried out treatment with bismuth, Prolophen, and N.A.B. The appearance of the left eye was in no way affected by this treatment.

Progress.—Repeated ophthalmoscopic observations were made and by January 4, 1951, it was decided that there had certainly been some increase in the size of the solid detachment

FIG. 2.—Scotoma on September 20, 1950.
Test objects: 30/2000 and 2/2000 white.
Vision 6/9 partly and J 2.

FIG. 3.—Scotoma on January 17, 1951.
Test objects: 30/2000 and 2/2000 white.
Vision 6/12 (2 letters) and J 2.

of the retina and that there was also a secondary serous detachment of the retina at the inferior periphery. The pigment on the nerve head showed no change. J 2 type could still be read slowly. The fundus drawing (Fig. 1) shows the appearance on January 17,

1951. The visual fields were charted at intervals and the isopters delimiting the scotoma showed some fluctuations (Figs 2 and 3); at no time was there any evidence of a nerve-fibre bundle defect, and this, together with the preservation of the greater part of the visual field, was held to indicate that the head of the optic nerve was not itself invaded by tumour cells.

Operation.—The left eye was enucleated on January 19, 1951, the diagnosis of malignant melanoma adjacent to the optic disk being based on the onset of symptoms, the appearance of a solid detachment of the retina increasing in size, the changes in the visual field, and the secondary detachment of the retina at the periphery. It was considered that the pigment overlying the nerve head was congenital.

Histology.—The eyeball was fixed in formal saline and divided antero-posteriorly through the optic nerve after freezing. The macroscopic appearance of the divided eyeball is shown in Fig. 4.

FIG. 4.—Macroscopic appearance of the divided eyeball. (The detachment of the retina is mostly artefact.)

FIG. 5.—Section of nerve head. Haematoxylin and eosin $\times 15$.

a, a: benign melanoma of the disk. *b*: sub-retinal exudate. *c*: malignant melanoma of the choroid. *d*: extension of *c* into the retina. *e*: extra-ocular extension of *c* into the pial sheath.

Microscopical sections (Figs 5 and 6) show the nerve head to be considerably enlarged; this is due mainly to the presence of a mass of densely black pigmented tissue which lies in the nerve fibre layer itself, with smaller masses lying in the deeper layers as far back as the lamina cribrosa. This aggregation of pigmented cells is interpreted as a congenital benign melanoma of the optic disk which has probably been derived from the lamina choroidalis of the lamina cribrosa. In the juxta-papillary region of the choroid there is a typical malignant melanoma, the cells of which are of the epithelioid type, with a heavy reticulin formation in the tumour mass. The pigment-bearing cells of this tumour have extended into the retina and have also grown around the free termination of the lamina vitrea and through the intermediary tissue of Kuhnt. This, as Reese (1925) pointed out, is the line of least resistance in this region. It seems likely that this malignant change occurred in a benign juxta-papillary melanoma. In one area near the disk malignant cells can be seen extending along a blood vessel and they form a small nidus of growth in the pia near the lamina cribrosa being thus an extra-ocular extension of the tumour. No direct connection could be seen between this malignant tumour and the mass of pigmented tissue on the nerve head; the cells of the latter appear to be melanoblasts and they do not have the appearance of pigment invasion from a choroidal neoplasm. There is a marked difference between the dense pigment on the nerve head and the comparatively poorly pigmented cellular mass of the choroidal tumour.

Discussion

The striking ophthalmoscopic appearance of this case naturally aroused

FIG. 6.—Portion of melanoma of disk showing densely pigmented melanoblasts. Haematoxylin and eosin. $\times 300$.

interest at once, and observation showed that the striated mass of pigment overlying the nerve head had remained unchanged over a period of 4 months while the solid detachment of the retina adjacent to the disk increased in extent. This suggests that the former was a benign melanoma while a malignant change was responsible for the latter. The histological appearances supported this interpretation.

The striated appearance of the mass of pigment closely resembles that illustrated in a number of the previously reported cases, notably those of Liebreich, Hirschberg, and Pálch Szántó. It suggests at the least a close spatial relationship with the nerve fibres. It is noteworthy from the clinical point of view that perimetry did not give any evidence of a nerve-fibre bundle defect, nor was there any peripheral depression. These negative findings argued against the presence of growing tumour tissue in the nerve head itself. The case is thus in line with that reported by Pálch Szántó and contrasts with that of Oguchi in which a growing tumour was present on the disk.

The origin of melanomata of the optic disk has given rise to some discussion. Following the researches of Berger (1882), who maintained that the innermost layer of the lamina cribrosa is derived from the choroid, it has been recognized that its peripheral part contains chromatophores from the proliferation of which pigmentation of the disk might arise. Such pigmentation would be expected to be mainly deep to the nerve fibres and indeed, if small in amount, might not be distinctly visible clinically.

Such a choroidal (mesodermal) origin has been accepted by Ogawa (1905),

Oguchi (1933), Thomson and Ballantyne (1903), and Scheerer (1922). The pigmentation would develop between the 5th and 7th month of gestation.

On the other hand, a retinal (ectodermal) origin was postulated by Pick (1900), who pointed out that prolongations of the internal pigmented layer might be cut off before the 7th week by developing nerve bundles and might persist and proliferate instead of regressing.

A glial origin was suggested by Krückmann (1906; quoted by Ginsberg, 1908; Scheerer, 1922; and Michail, 1934), and Vetter (1950) emphasized the pigmentation in the sheaths of the nerve in his case.

Summary

A case of benign melanoma of the optic disk with associated juxta-papillary malignant melanoma of the choroid is described and the histological appearances are illustrated. There is a brief historical survey and discussion.

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