

## CASE NOTES

### PRIMARY MALIGNANT MELANOMA OF THE CORNEA\*

BY

H. B. STALLARD

*London*

DUKE-ELDER (1938) states that isolated pigment flecks in the corneal epithelium are very rare, and quotes Steiner (1905, 1923), who noted two instances in natives of Java, one in a normal eye and the other in a trachomatous eye. Krämer (1906) also observed such flecks in a Mexican.

Very rarely, associated with diffuse melanosis of the conjunctiva, there is an extension of pigment over the cornea, diffusely or in tongue-shaped processes, the pigment being limited to the basal epithelial cells.

Congenital pigmentation extending from the conjunctiva into the cornea is less rare in animals than in man and has been noted in the horse, dog, pig, sheep, guinea-pig, and frog.

Some pathologists have the opinion that the basal cells of the corneal epithelium are potential melanoblasts. Indeed, melanin is found in Stähli's and Hudson's lines, and it is an occasional sequel to irradiation of the eye. It is, however, still disputed whether malignant melanoma could arise from the Schwannian cells of the corneal nerves or from the cells of the epithelium; the present trend is probably towards the latter view.

Because of the rarity of a benign melanoma or melanosis of the corneal epithelium which ultimately becomes malignant it seems of interest to record the following case:

#### Case Report

A man aged 62 attended Moorfields Eye Hospital, stating that the vision of the right eye had been defective since the age of 18. Many clusters of fine brown pigment dots were seen in the corneal epithelium (Fig. 1, opposite).

He failed to return for follow-up until 10 years later, when a large nodular pinkish mass covered the cornea and part of the sclera of the right eye (Fig. 2, opposite). The mass moved freely with the ocular movements. The eyelids were lifted by it but not infiltrated. There was no lymphadenopathy and no radiographic evidence of metastases in the skull, chest, and spine.

*Operation.*—The right orbit was exenterated and the cavity lined by a split skin graft retained in place by an alginate mould. The graft took well (Fig. 3, opposite), and a camouflage prosthesis was fitted (Fig. 4, opposite).

---

\* Received for publication February 13, 1961.

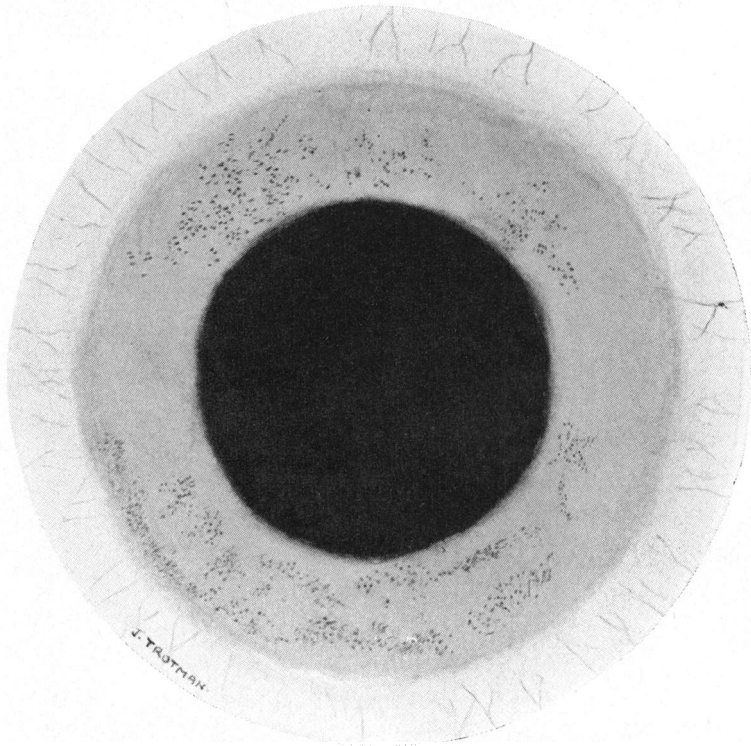


FIG. 1.—Corneal epithelium, showing pigment dots.



FIG. 2.—Mass covering cornea and part of sclera in right eye.

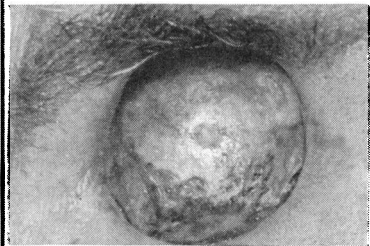


FIG. 3.—Right orbit after exenteration and skin graft.



FIG. 4.—Prosthesis in right orbit.

*Pathological Report*

**MACROSCOPIC.**—A bilobed, firm, pinkish-white neoplasm covered the whole cornea and bulbar conjunctiva and its cut surface was white, firm, and homogeneous (Fig. 5).

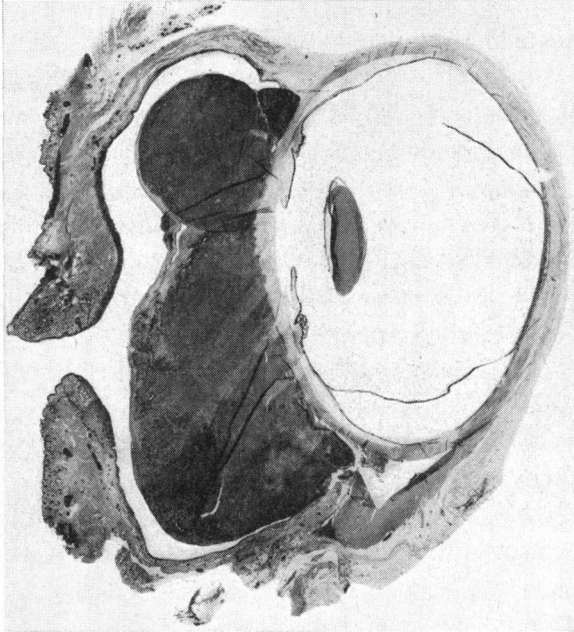


FIG. 5.—Neoplasm covering cornea and bulbar conjunctiva.

There were two areas of finely granular flat diffuse pigmentation, one in the lateral fornix and the other in the lower fornix and over the palpebral conjunctiva of the lower lid.

**MICROSCOPIC.**—An almost unpigmented melanoma composed of interlacing bundles of spindle cells (Fig. 6, opposite) with areas of cytoplasmic vacuolations had destroyed most of the corneal epithelium, Bowman's membrane, and the bulbar conjunctiva, and had infiltrated the substantia propria of the cornea. The sections showed intra-epithelial malignant changes and cellular proliferation in various stages of evolution. Other features were corneal folding, vascular sclerosis in the posterior choroidal vessels, and retinal oedema. There was no evidence of malignant extension into the eye or orbit.

The patient died of intercurrent disease three years after exenteration.

### Discussion

In the literature since 1892 I have been unable to find more than five cases in which it was certain that malignant changes occurred in a benign melanoma arising in the corneal epithelium. In a number of others it seemed probable that the growth had its origin at the limbus and extended thence across the cornea. The latter I have disregarded for the purpose of this paper.

In 1886 Blanquinque had under his care a woman aged 68, who for 20 years had a circular reddish-brown fleck on the cornea in its upper temporal quadrant with 1 mm. of clear cornea between the neoplasm and the limbus. For 11 years this neoplasm had not grown and during the following 9 years

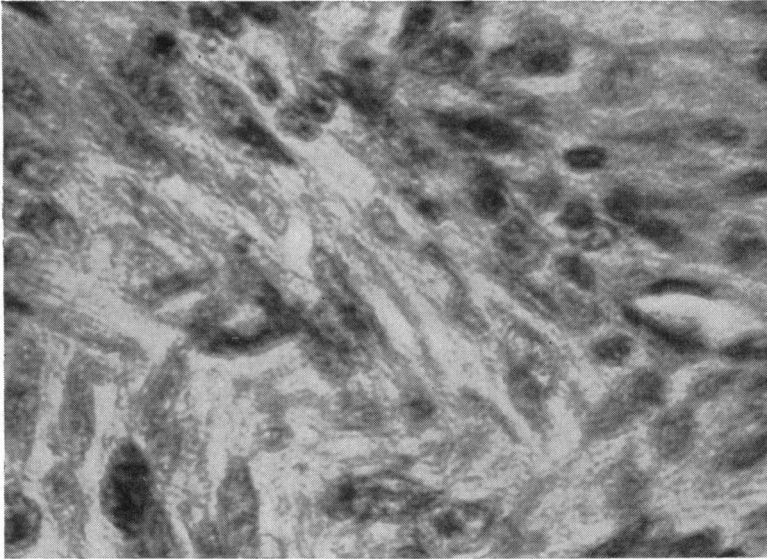


FIG. 6.—Section showing melanoma composed of interlacing bundles of spindle cells.  $\times 700$ .

it had slowly increased in size to become 6 mm. in diameter and 3 mm. thick. On April 13, 1886, a partial keratectomy was done, and the neoplasm was found to be malignant. The patient was still alive with good vision 4 years later (Blanquinque, 1892).

Dean (1913) treated by partial keratectomy a woman aged 63, who had a rust-coloured neoplasm in the upper half of the cornea. Its diameter was 5 mm. and its thickness 2 mm. It extended a little below the 3 to 9 o'clock meridian and there was 1 mm. of clear cornea between its upper edge and the limbus. Histological examination showed that the neoplasm was composed of round, oval, oat-shaped and spindle cells with many mitotic figures. The corneal epithelium had been eroded in the centre. Bowman's membrane was intact except at the centre of the base where there was slight infiltration of the substantia propria.

Reese (1951) commented on the rarity of malignant melanomata arising from the corneal epithelium and wrote of a man, aged 89, who had a malignant melanoma of the cornea unconnected with the limbus.

Davies and Bailey (1954) described the case of a white man, aged 53, who had a pigmented neoplasm arising from the cornea,  $8 \times 5$  mm. in area and 2 mm. thick. The summit of the neoplasm was ulcerated and the eye was very painful. The neoplasm, which had extended between the epithelium and an intact Bowman's membrane, was composed of sheets and whorls of spindle cells. The pigment, vascularity, and reticulin content were moderate.

François, Gildemyn, and Rabaey (1956) reported a patient aged 55, who had carcinomatous melanosis in the temporal half of the cornea with a band

of clear cornea between the periphery of the neoplasm and the limbus. As the authors considered that the neoplasm was slowly penetrating, a partial keratectomy was done with a good result.

### Summary

A description is given of the clinical and pathological features of a benign melanoma (melanosis) of the corneal epithelium in a man aged 62 which, 10 years later, became malignant when the patient was aged 72.

The sparse literature about malignant melanomata arising in the corneal epithelium is briefly reviewed.

I thank my friend Mr. J. H. Doggart for his kindness in wishing me to publish Fig. 1, a painting which he had made of the corneal pigment in this patient some years before we exchanged clinics at Moorfields Eye Hospital. My thanks are also due to Prof. Norman Ashton for the pathological report, and to Dr. Peter Hansell of the Medical Illustration Department of the Institute of Ophthalmology for help with the illustrations.

### REFERENCES

- BLANQUINQUE, M. (1892). *Recueil Ophtal.*, **14**, 214.  
 DAVIES, W. S., and BAILEY, W. H. (1954). *A.M.A. Arch. Ophthalm.*, **52**, 923.  
 DEAN, L. W. (1913). *Ann. Ophthalm. (St. Louis)*, **22**, 764.  
 DUKE-ELDER, S. (1938). "Text-book of Ophthalmology", vol. 2, p. 1396. Kimpton, London.  
 FRANÇOIS, J., GILDEMYN, H., and RABAËY, M. (1956). *Ann. Oculist. (Paris)*, **189**, 496.  
 KRÄMER (1906). *Zbl. prakt. Augenheilk.*, **30**, 39.  
 REESE, A. B. (1951). "Tumors of the Eye". Hoeber, New York.  
 STEINER, L. (1905). *Zbl. prakt. Augenheilk.*, **29**, 293.  
 ——— (1923). *Ann. Oculist. (Paris)*, **160**, 137.