ORBITAL NEURILEMMOMA WITH CAFÉ-AU-LAIT PIGMENTATION OF THE SKIN*†

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IN the two cases here described, a solitary orbital neurilemmoma was associated with pigmentation of the skin suggesting an association with von Recklinghausen's disease (Strachov and Shepkalova, 1941; Reese, 1963).

Case Reports

There was no important family history and no history of trauma or x-ray therapy, and a complete general medical examination was negative.

Case 1, a 35-year-old woman (Fig. 1), complained of right proptosis of one year's duration. Her skin showed areas of *café-au-lait* pigmentation (Fig. 2).

FIG. 1.—Case 1. Right proptosis of one year's duration due to orbital encapsulated neurilemmoma in a woman aged 35 years.

FIG. 2.—Case 1. *Café-au-lait* areas of pigmentation on the skin of the back.

Examination.—The left eye was normal with visual acuity 6/12. The right eye showed proptosis 20 mm. (left 15 mm.), and deviated outwards with limitation of ocular movements inwards. The fundus showed optic atrophy and the visual acuity was hand movements. A firm mass was felt in the inner side of the orbit extending backwards.

Surgery.—Through a medial fornix conjunctival incision the tumour was removed by blunt finger dissection. It was encapsulated and firm, measuring 3 × 3 cm., with a smooth surface (Fig. 3, opposite) and pink in colour.

Histopathological Examination.—There were spindle Schwann cells with elongated nuclei showing the characteristic palisade arrangement. Between the cells were long slender straight or serpentine reticulum fibres giving the appearance of spindle cellular fibrillar tissue of Antoni type A (Fig. 4, opposite). There were no nerve fibrils. The appearances were those of a neurilemmoma.

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FIG. 3.—Case 1. Encapsulated tumour after removal.

FIG. 4.—Case 1. Section of neurilemmoma. $\times 400$.

Case 2, a 60-year-old woman (Fig. 5), complained of left gradual proptosis of 2 years' duration.

Examination.—The right eye showed a mature senile cataract; the visual acuity was hand-movements with good projection of light.

The left eye showed upward proptosis 20 mm. (right 15 mm.), an immature senile cataract, and visual acuity 1/60. The fundus showed optic atrophy. There was a palpable mass between the globe and the lower orbital margin.

FIG. 5.—Case 2. Left proptosis of 2 years' duration due to orbital encapsulated neurilemmoma in a woman aged 60 years.

Surgery.—Through a lower fornix conjunctival incision the tumour was removed by blunt finger dissection. It was encapsulated, firm, and pink, measuring 2×3 cm. (Fig. 6, overleaf).

Histopathological Examination.—The appearances were those of a neurilemmoma (Fig. 7, overleaf).

A 5-year follow-up of the two cases showed neither proptosis nor recurrence of the neoplasm.

Discussion

It is the general opinion that a neurilemmoma is an isolated entity, while a neurofibroma may be a local manifestation of von Recklinghausen's neurofibromatosis (Duke-Elder, 1952). Herbut (1959) said that it mattered little from which type of cell the tumour originated or what name it was given as they have similar pathological and clinical properties; when the tumour is multiple the condition is referred to as neurofibromatosis. A neurilemmoma may show nerve fibres in its periphery and a neurofibroma usually contains spindle cells frequently arranged with palisading of the nuclei.

FIG. 6.—Case 2. Encapsulated tumour after removal.

FIG. 7.—Case 2. Section of neurilemmoma. $\times 90$.

Summary

(1) Although neurilemmoma is considered to be an isolated entity not associated with neurofibromatosis, these two solitary orbital encapsulated neurilemmomata were associated with patches of *café-au-lait* pigmentation.

(2) These are the first two orbital neurilemmomata to be described from Egypt.

REFERENCES

- DUKE-ELDER, S. (1952). "Text-book of Ophthalmology", vol. 5, p. 5579. Kimpton, London.
 HERBUT, P. A. (1959). "Pathology", 2nd ed., p. 1391. Kimpton, London.
 REESE, A. B. (1963). "Tumors of the Eye", 2nd ed., p. 202. Harper and Row, New York, Evanston, and London.
 STRACHOV, V. P., and SHEPKALOVA, V. M. (1941). *Vestn. oftal.*, **18**, 12.