SOLITARY ORBITAL MALIGNANT NEURILEMMOMA*†

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A SERIES of 500 cases of proptosis included two due to orbital malignant neurilemmoma, which according to Duke-Elder (1952) and Hogan and Zimmerman (1962) is a very rare condition.

Case Reports

Case 1, a 50-year-old woman (Fig. 1), had a left forward painless proptosis of 7-months' duration.

Examination.—The right eye was normal and the visual acuity 6/9.

The left eye showed proptosis 23 mm. (right eye 16 mm.), oedema of the lids, and limitation of the ocular movements in all directions. The visual acuity was counting fingers at 10 cm. The left fundus showed optic atrophy. A mass was palpable in the outer lower quadrant of the orbit. Skull *x* rays showed the left orbital cavity to be wider than the right.

FIG. 1.—Case 1. Left proptosis due to orbital malignant neurilemmoma in a woman aged 50 years.

FIG. 2.—Case 1. Orbital malignant neurilemmoma, showing non-capsulated lobulated tumour removed at operation.

FIG. 3.—Case 1. Section of tumour, showing palisading of nuclei. $\times 540$.

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Operation.—The tumour was removed through a lower fornix conjunctival incision; it was grey in colour, soft in consistency, non-capsulated and lobulated (Fig. 2), and measured $5 \times 3 \times 1$ cm.

Histopathological Examination.—The tissue was highly cellular with spindle-shaped cells and hyperchromatic elongated nuclei arranged mostly in palisade formation (Fig. 3), but occasionally in whorls. The stroma consisted of loose reticular tissue. Mitotic figures and tumour giant cells were present in many fields. The appearance was that of a malignant neurilemmoma.

Result.—The patient refused left orbital exenteration and treatment was continued by irradiation of the orbit, but she died 6 months later of liver metastases.

Case 2, a 55-year-old man (Fig. 4) complained of right proptosis of 4 months' duration.

Examination.—The left eye was normal, with a normal fundus and visual acuity 6/12.

The right eye showed proptosis 24 mm. (left eye 15 mm.), with ptosis and limitation of ocular movements in all directions. The visual acuity was 6/60. The right fundus showed papilloedema. A mass was palpable in the upper inner quadrant of the orbit. Skull x rays showed no abnormality.

Operation.—The tumour was removed through an upper-inner orbital incision; it was grey in colour, soft in consistency, non-capsulated and lobulated, and measured $4 \times 3 \times 2$ cm.

FIG. 4.—Case 2. Right proptosis due to orbital malignant neurilemmoma in a man aged 55 years.

FIG. 5.—Case 2. Section of neurilemmoma. $\times 95$.

FIG. 6.—Case 2. Section of tumour, showing palisading of nuclei. $\times 540$.

Histopathological Examination.—The appearance was that of a malignant neurilemmoma (Figs 5 and 6).

Result.—The patient refused right orbital exenteration, and treatment was continued by irradiation of the right orbit, but the tumour recurred after 9 months. This time the right orbit was exenterated but the patient died 4 months later of lung metastases.

Discussion

Malignant orbital neurilemmomata have been reported by Cerveira (1953), Ginde (1953), Schiff-Wertheimer and Loisillier (1954), and Reese (1963a), and approximately one-half of such cases arise in patients with von Recklinghausen's disease (Reese, 1963b). Hosoi (1931) estimated that malignant neurilemmomata developed in 13 per cent. of cases of von Recklinghausen's disease, but Stout (1949) considered this figure too high. Tumours occurring in cases of generalized neurofibromatosis are better considered as neurofibrosarcomata.

Summary

Two rare cases of solitary orbital malignant neurilemmomata causing proptosis (the first in Egypt) are described. There was no associated neurofibromatosis.

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