Intrascleral neurilemmoma

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SUMMARY The case of an anteriorly situated, intrascleral, neurilemmoma in an 11-year-old girl is reported. Only two similar cases have been described in the literature, and the diagnosis was made only after detailed histological examination, including electron microscopy and specialised staining techniques, to differentiate a neurilemmoma (schwannoma) from a neurofibroma.

Neurilemmomas are benign, encapsulated tumours composed of proliferating Schwann cells, usually unassociated with neurofibromatosis. They arise from ciliary nerves in the orbit and account for about 1% of primary orbital tumours.¹

Shields *et al.*² in 1981 reviewed the literature for solitary nerve sheath tumours of the globe and found 17 (including the one they described) involving the uveal tract and three involving the sclera. The average age of those patients with uveal tumours was 37, with a female:male ratio of 3.6:1. Only two of the 17 were associated with neurofibromatosis. These authors pointed out, however, that many of the cases reported had not been investigated by modern histological techniques and that it was impossible to state with certainty whether the tumours were neurilemmomas or neurofibromas. When Rosso *et al.*³ considered these cases, they believed only six to be proved neurilemmomas.

Neurilemmomas derived from ciliary nerves in the sclera occur even more rarely than in the uveal tract. One tumour has been described situated posteriorly in a 76-year-old female, in whom it simulated an orbital tumour,⁴ and two cases of limbal neurilemmomas occurring in a 14-year-old female and a 12-year-old male have been reported in the older German literature.^{5,6}

Case report

An 11-year-old Cypriot girl presented to the Casualty Department in July 1981 with a one-month history of

a small lump in the upper part of her left eye. On examination the visual acuity was 6/5 unaided, and two small, apparently separate masses were noted in the superotemporal aspect of the globe just posterior to the limbus. No other abnormality was detected, and a full blood count and differential white cell count performed at that time were normal. A decision was made to withhold surgery until the masses caused symptoms.

Over the next three years the masses enlarged, and by the time she returned in July 1984 requesting surgical treatment they were clearly parts of the same

Fig. 1 Eyelid raised to demonstrate tumour.

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Fig. 2 Demonstration of dumb-bell contour of tumour.

lesion. The visual acuity was 6/6 unaided and she had developed a mechanical ptosis.

By June 1985 the eye had become uncomfortable, the ptosis had increased to 3 mm, with the contours of the lump visible through the upper lid, and the mass had noticeably enlarged (Figs. 1, 2). The superior part of the cornea had become distorted, and fundal examination revealed a deep indent corresponding to the external position of the posterior part of the mass. General physical examination did not suggest any underlying systemic disease, and serological tests for

parasitic infections were negative. An ultrasound examination showed the mass to be solid.

In October 1985 the tumour was excised under general anaesthetic. Highly vascular conjunctiva was readily separated from the underlying, well encapsulated mass, which in turn was separated from the globe by blunt dissection to reveal a patch of partial-thickness sclera with a large area of bare choroid centrally. Minimal manipulation led to rupture of the globe, and it was necessary to close the defect with donor sclera.

The postoperative course was uneventful, and when she was last seen in the summer of 1987 the patient was asymptomatic.

PATHOLOGY

The dumb-bell shaped tumour measured 19×7×7 mm. There was a hyalinised connective tissue capsule round the vacularised tumour, which consisted of intertwining fascicles of spindle cells showing little pleomorphism. In some areas there was palisading of nuclei (Fig. 3) and in others myxoid degeneration. Collagen deposition was marked (Fig. 4). The differential diagnosis included fibroma, neurofibroma, and neurilemmoma. No axons were demonstrable within the substance of the tumour on immunohistochemical staining with antineurofilament protein antibody, but the S-100 stain was strongly positive in the spindle cell areas. Electron microscopy showed Luse bodies (Fig. 5) between cells with flattened and invaginated nuclei; microfibrils were present within the cytoplasm with occasional lysosomes and mitochondria.

Fig. 3 Palisaded nuclei (arrowed) in spindle area of tumour. (H and E, ×180.)

Fig. 4 Extensive reticulin framework present within the tumour. (Silver preparation, $\times 180$.)

Fig. 5 Electron micrograph showing Luse bodies (arrowed) composed of long-spaced collagen. ($\times 15\,000$.)

The tumour was therefore classified as a neurilemmoma arising from a nerve sheath.

Discussion

This tumour showed the extensive collagenisation recorded within orbital tumours but in addition had the classical Antoni A areas of palisaded spindled cells, mixed with the more myxoid areas of degeneration, known as Antoni B. The latter areas, with their cystic potential, are said to account for the intermittent waxing and waning in size noticed clinically by some patients. Both neurilemmomas and neurofibromas are derived from a common stem cell, the Schwann cell, and both may be expected to show S-100 positivity. The more universal staining pattern, however, in this tumour was that associated with a neurilemmoma. Fibroblastic tumours do not stain with S-100 and on electron microscopy do not have the prominent basal laminae or interdigitating processes seen in this case. The Luse bodies⁷ seen in Fig. 5 consist of aggregates of long-spaced or broad-banded collagen with an axial periodicity of 130 nm. These bodies were originally thought to be pathognomic of neurilemmomas but are now known to occur in other tumours, including basal and squamous carcinomas and in naevoid tumours, also of neural crest origin.⁸ Elsewhere in the eye similar collagen is seen in structures such as Descemet's membrane, of neural crest derivation.

From the surgical point of view this case illustrated the potential risk associated with the removal of an apparently superficial tumour from the globe. If the sclera is thin, there is a risk of rupture, and the possibility that a scleral patch graft may be required should be borne in mind.

We are grateful to Professor Roy Weller, of Southampton, for performing the neurofilament staining.

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Accepted for publication 28 July 1988.