Schwannoma of the ciliary body treated by block excision

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Abstract

A 26-year-old man developed a non-pigmented ciliary body tumour of his right eye. A 7 mm block excision and tectonic corneoscleral graft were performed. The excised tissue was studied using histopathological, immunohistochemical, and electron microscopic techniques. The tumour revealed characteristic features of a Schwann cell neoplasm including Antoni A and B patterns, acid mucopolysaccharides, S-100, and vimentin positivity, and – by electron microscopy – Luse bodies. It was classified as a schwannoma. Although rare, schwannoma should be included in the clinical differential diagnosis of non-pigmented ciliary body tumours. Local excision should be considered to avoid over-treatment by enucleation.

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Ciliary body schwannomas (neurilemmomas) are very rare benign peripheral nerve neoplasms.^{1–9} In two large histopathological studies that reviewed a total of 955 globes enucleated for uveal melanoma or pseudomelanoma, only one uveal schwannoma (0.1%) was found.^{4,10} Ciliary body schwannomas are frequently misdiagnosed as malignant melanomas resulting in unnecessary enucleation.^{1,2,4,6,8} Therefore, we report the clinical, histological, immunohistochemical, and ultrastructural findings of a ciliary body schwannoma that was treated by block excision.

Clinical history

A 26-year-old white Turkish man presented with a prominent ciliary body tumour of his right eye. His medical history included inactive pulmonary tuberculosis but was of no help otherwise. There were no clinical signs of neurofibromatosis. For 3 years, the patient had noticed slight slowly progressive blurred vision of his right eye. Two years earlier, an 'iris cyst' of the right eye had been diagnosed and 'laser treatment' had been performed in Turkey. Corrected visual acuity was right eye 20/25, left eye 20/32, and intra-ocular pressure was right eye 18 mm Hg and left eye 16 mm Hg. On examination, the left eye was completely normal except for anisometropic amblyopia. Slit-lamp examination of the right eye revealed a prominent, non-pigmented ciliary body tumour extending over 3 clock hours from 8 o'clock to 11 o'clock with erosion of the basal iris pigment epithelium. The tumour appeared solid with a smooth vascularised surface (Fig 1). Gonioscopy revealed a closed anterior chamber angle in the area of the tumour without tumour invasion into the anterior chamber or dispersion of tumour cells. The lens showed focal cortical

opacification in the area of the tumour. There were no vitreous infiltrates, and the retina was flat without signs of shifting subretinal fluid. The optic nerve head appeared normal. A and B scan ultrasonography revealed a solid, homogeneous ciliary body tumour. Tumour prominence was 5 mm. Aqueous flare measured with the laser flare cell meter was normal (6.7 photon counts/ms). Computed tomography or magnetic resonance imaging scan were not performed. The clinical differential diagnoses included adult medulloepithelioma and adenoma of the non-pigmented ciliary body epithelium. However, as a non-pigmented malignant melanoma could not be excluded, a 7 mm block excision of the ciliary body tumour including adjacent cornea, sclera, iris, and ciliary body was performed and the defect was closed with a corneoscleral graft. The postoperative course was complicated by vitreous haemorrhage which was treated by vitrectomy. Following vitrectomy, a traction retinal detachment developed which necessitated scleral buckling, membrane peeling, and sulphur hexafluoride gas tamponade. After this procedure, the retina was attached and corrected visual acuity was 20/100.

Material and methods

The excised tissue consisted of cornea, sclera, ciliary body, and iris. A whitish solid ciliary body tumour with a vascularised surface was present (Fig 2). Tumour diameter was 5 mm, and tumour prominence was 5 mm. The tissue was bisected radially and processed for light microscopy, immunohistochemistry, and transmission electron microscopy.

Figure 1 Slit-lamp photograph showing the ciliary body tumour in the patient's right eye. Notice the non-pigmented smooth tumour surface.

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Figure 2 Gross photograph of excised tissue consisting of cornea, sclera, ciliary body, and solid non-pigmented prominent tumour (asterisk) measuring 5×5 mm (×9).

Figure 3 Photomicrograph showing ciliary body tumour (asterisk) and occluded chamber angle (arrowheads) (periodic acid Schiff, ×33).

Figure 4 Photomicrograph of densely packed spindle shaped tumour cells showing fascicular arrangement (Antoni A pattern) (periodic acid Schiff, ×146).

Results

Histologically, the chamber angle was closed (Fig 3). The ciliary body tumour was partially surrounded by a pseudocapsule and composed of spindle cells with rather small oval nuclei, eosinophilic cytoplasm, and small cytoplasmic extensions. Some areas of the tumour appeared solid with a densely packed fascicular arrangement of tumour cells (Antoni A pattern) (Figs 4, 5). In other areas, a loose, myxoid arrangement of tumour cells and extracellular mucoid substances were present (Antoni B pattern). The tumour cells appeared benign as the cells were uniform and mitoses or nuclear anomalies were absent. Using colloidal iron stains, acid mucopolysaccharides were demonstrated in the tumour. Immunoperoxidase staining showed tumour cell expression of S-100 protein and vimentin, whereas staining for desmin was negative. Transmission electron microscopy demonstrated spindle cells with electron lucent cytoplasm containing cytoplasmic filaments and sparse organelles (scattered mitochondria and lysosomes, short segments of rough endoplasmic reticulum) and long, delicate cytoplasmic processes (Fig 6). The tumour cells and their processes were covered by basement membrane material. Aggregates of banded basement membrane material (Luse bodies) were also present (Fig 7).

Given the histopathological, immunohistochemical, and electron microscopic findings, the diagnosis of a benign schwannoma (neurilemmoma) was made.

Discussion

Schwannomas (neurilemmomas) are benign nerve sheath tumours of Schwann cell origin. In ophthalmic patients, they arise most frequently in the orbit where they account for approximately 0.5–1% of orbital tumours.^{11–13} Other periocular or ocular locations include the eyelids,¹⁴ the area of the nasolacrimal duct,¹⁵ conjunctiva,¹⁶ caruncle,¹⁷ limbus, sclera,^{18,19} choroid,^{20–23} and ciliary body.^{1–9} The development of ocular and periocular schwannomas may be associated with neurofibromatosis.^{20,24,25}

To our knowledge, a total of nine cases of ciliary body schwannoma have been reported so far.^{1–9} Six of those were clinicopathological case reports,^{1,2,5,6,8,9} two were brief descriptions and illustrations in ocular pathology textbooks,^{3,7} and one was briefly mentioned in a study of lesions mistaken for uveal melanomas.⁴ Electron microscopic and/or immunohistochemical verification of the diagnosis was done only in four of the nine reported cases.^{6–9} As light microscopic differentiation of schwannoma from other benign spindle cell tumours and especially from neurofibroma is extremely difficult, and as the terms neurofibroma and neurilemmoma were not sharply separated before electron microscopy was available, the diagnosis of schwannoma in the early reports is probably somewhat uncertain. In none of the nine reported cases was a schwannoma suspected clinically, and seven of the nine eyes were enucleated because a malignant melanoma was suspected.^{1–4,6–8}

On clinical grounds, it is impossible to differ-

Figure 5 Higher magnification of tumour cells showing Antoni A pattern (periodic acid Schiff, $\times 294$).

Figure 6 Electron micrograph showing closely packed and intertwined cell processes of Schwann cells. Schwann cell bodies and processes are diffusely covered by basal lamina (arrowheads). Asterisk indicates Schwann cell nucleus ($\times 4785$).

Figure 7 Electron micrograph of characteristic aggregate of banded basement membrane material (Luse body) (arrow) ($\times 6195$).

entiate ciliary body schwannomas from other benign tumours such as leiomyoma, mesectodermal leiomyoma, adult medulloepithelioma, and adenoma of the non-pigmented ciliary epithelium. The most important differential diagnosis of ciliary body schwannoma is non-pigmented malignant melanoma, and it may be very difficult or even impossible to exclude a melanoma with certainty on clinical grounds. Clinical signs indicating malignant melanoma include rapid tumour growth, shifting subretinal fluid, invasion of the anterior chamber, and dispersion of tumour cells on the iris surface or throughout the chamber angle structures. Ancillary diagnostic studies such as fluorescein angiography, ultrasonography, computed tomography, and magnetic resonance imaging may be useful in the differential diagnosis. Non-invasive quantification of aqueous flare with the laser flare cell meter may also be helpful in the differential diagnosis of malignant and benign uveal lesions. It has been shown that aqueous flare is significantly increased in eyes with choroidal and ciliary body malignant melanomas.²⁶ Interestingly, aqueous flare was in the normal range in our case. Fine needle biopsy and cytological examination may be another means of differentiating ciliary body tumours of unknown origin.^{27–28}

Histologically, the tumour showed characteristics of a benign spindle cell tumour. For exact diagnosis, immunohistochemical studies and electron microscopy are required in addition to light microscopy. Electron microscopy displayed the characteristic findings of schwannomas including identification of Schwann cells with long cell processes and basal lamina long spacing basement membrane material referred to as Luse bodies.^{29–31} In our case, block excision of the ciliary body tumour with adjacent cornea, full thickness sclera, ciliary body, and iris was performed.³² Thereby, the tumour was removed in toto and enucleation was avoided.

Benign schwannoma should be included in the differential diagnosis of non-pigmented ciliary body lesions, and enucleation should be avoided in these cases. In prominent non-pigmented ciliary body tumours that are localised, extend over less than 6 clock hours, and do not show diffuse infiltration of the iris surface or chamber angle structures, block excision may be an appropriate therapeutic measure to remove the tumour in toto, allow histological diagnosis, and avoid overtreatment by enucleation or radiation.

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