A CASE OF MELANOCYTOMALYTIC GLAUCOMA

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Case report

A 35-year-old white male was referred with an iris lesion in the left eye, asymptomatic and present since the age of 10 years old. The visual acuity was 6/4 in both eyes and the intraocular pressure (IOP) was normal. The left anterior segment harboured a deeply pigmented iris tumour infero-nasally with pigment accumulation in the angle on gonioscopy, especially inferiorly (Figure 1A). Ultrasound B scan showed medium internal echogenicity and an elevation of 1.8 mm. The appearances were consistent with a melanocytoma.

Four months later, the patient noticed left eye photophobia. There were now pigment granules in the anterior chamber with pigment seeding across the iris, causing heterochromia and increased trabecular meshwork pigmentation throughout the iridocorneal angle (Figure 1B). The IOP in the left eye was 26 mm Hg and the optic disc appeared healthy. Unilateral melanocytomalytic pigmentary glaucoma was treated with topical dorzolamide 2% and timolol 0.5%. The intraocular pressure failed to respond, despite maximal medical therapy (dorzolamide 2%, timolol 0.5%, apraclonidine 0.5% topically and oral acetazolamide), with IOP of 47 mm Hg. Left laser coagulation of the ciliary body was attempted to lower the IOP, but this too was insufficient. Local resection of the melanocytoma was performed to make a tissue diagnosis and to reduce pigment shedding in the anterior chamber. Histological analysis confirmed the diagnosis of iris melanocytoma without mitotic activity. There was no necrosis (Figure 1C). Following surgery, the IOP returned to normal (14 mm Hg) with only topical anti-hypertensive therapy (Figure 1D) (dorzolamide 2%, timolol 0.5%, apraclonidine 1%).

Discussion

Melanocytoma is a magnocellular nevus, which in the iris is most often located in the inferior and lateral quadrants[1]. Clinically, melanocytomas are deeply pigmented, with an irregular

"cobblestone" surface. These tumors can grow slowly, but malignant transformation is exceptionally rare.[3,4] Secondary glaucoma occurs in 11% of patients[2] as a consequence of pigment dispersion, which is thought to be related to tumour necrosis,[3] but has to be differentiated from iris ring melanoma, to avoid unnecessary enucleation.[5] On histological analysis, the melanocytoma cells have also been shown to invade the intertrabecular spaces.[6] In cases where medical treatment alone fails to normalize the intraocular pressure, excision of the iris melanocytoma should be considered in an attempt to prevent further dispersion of pigment and cells from the lesion.[2] The present case shows that dispersion of tumour cells is not always caused by tumour necrosis but could also be attributed to poor cohesion of melanocytoma cells. The mechanism of secondary glaucoma in our patient is thought to be related either to pigment dispersion or infiltration of trabecular meshwork by melanocytoma cells. However, this cannot be ascertained in the absence of histological analysis of the angle, which would only become possible if the patient requires enucleation in the future.

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Figure 1 legend

1A: well-demarcated heavily pigmented iris lesion diagnosed clinically as melanocytoma

1B: pigment seeding causing iris heterochromia, 4 months after Fig 1A

1C: histological section confirming the diagnosis of iris melanocytoma with no mitotic activity and no necrosis

1D: The anterior segment following local resection of the melanocytoma