Spontaneous Regression of Iris Melanocytoma: A Case Report

DOI 10.1055/s-0043-123642
Klin Monatsbl Augenheilkd 2018; 235: 473–475

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Georg Thieme Verlag KG
Rüdigerstraße 14
70469 Stuttgart
ISSN 0023-2165

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Spontanregression von einem Irismelanozytom: ein Fallbericht

Introduction

A melanocytoma is a rare, darkly pigmented nevus subtype, also called "magnocellular" nevus, as it consists of large and uniform polygonal cells with small nuclei and an intensely pigmented cytoplasm.

It was first described by Zimmerman in 1962 at the level of the optic disc [1], and was subsequently identified within the iridociliary complex. The choroid, sclera, or conjunctiva are exceptional locations [2].

Iridociliary melanocytomas are benign tumors, whose evolution can be marked by slow and limited growth (23% after 5 years) [3], which can be difficult to differentiate clinically from malignant transformation (lifetime risk: 2–5%) [3–5]. Spontaneous necrosis with secondary pigment dispersion glaucoma is a frequent complication (11–30%) [2,3], but to our knowledge, a nearly complete and spontaneous regression has never been reported.

Case Report

In April 2005, an 18-year-old female was referred to the Ocular Oncology Unit because of a progressive iris tumor (LE), noticed for the first time 3 years earlier.

Visual acuity was 1.0 in both eyes, with an intraocular pressure (IOP) of 18 mmHg OU. A darkly pigmented tumor was located within the nasal iris and associated with a discrete pigment dispersion over the whole iris and lens surface and within the iridocorneal angle (▶ **Fig. 1a, b**). On ultrasoundbiomicroscopy (UBM), tumor thickness was 1.4 mm (▶ **Fig. 1c**). A presumed diagnosis of iris melanocytoma was made, and a periodic observation proposed.

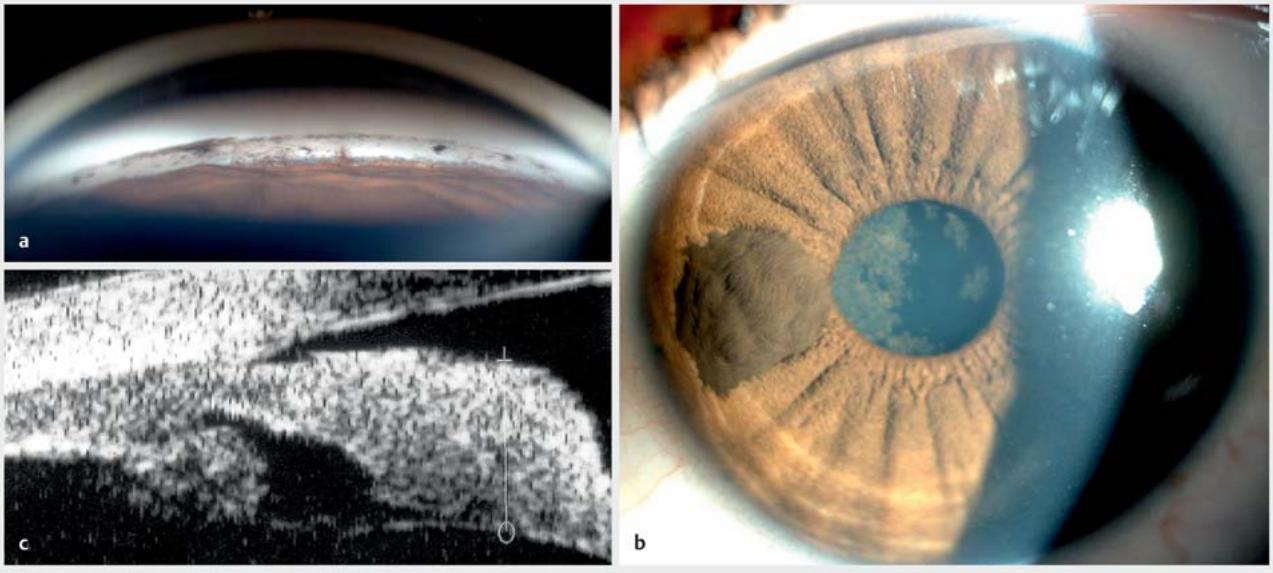
After 6 months, the tumor had not evolved. However, the IOP had increased from 18 to 28 mmHg, compatible with melanocytomalytic glaucoma, which was successfully treated with anti-glaucomatous drops (Brimonidine and Timolol).

After 19 months, the tumor had regressed in thickness, which was associated with an overall increase in the anterior chamber pigment dispersion (▶ **Fig. 2a, b**) and witnessing of a spontaneous tumor necrosis, which was confirmed by a reduced thickness on UBM of 1.1 mm (▶ **Fig. 2c**). IOP was 15 mmHg with the same drops.

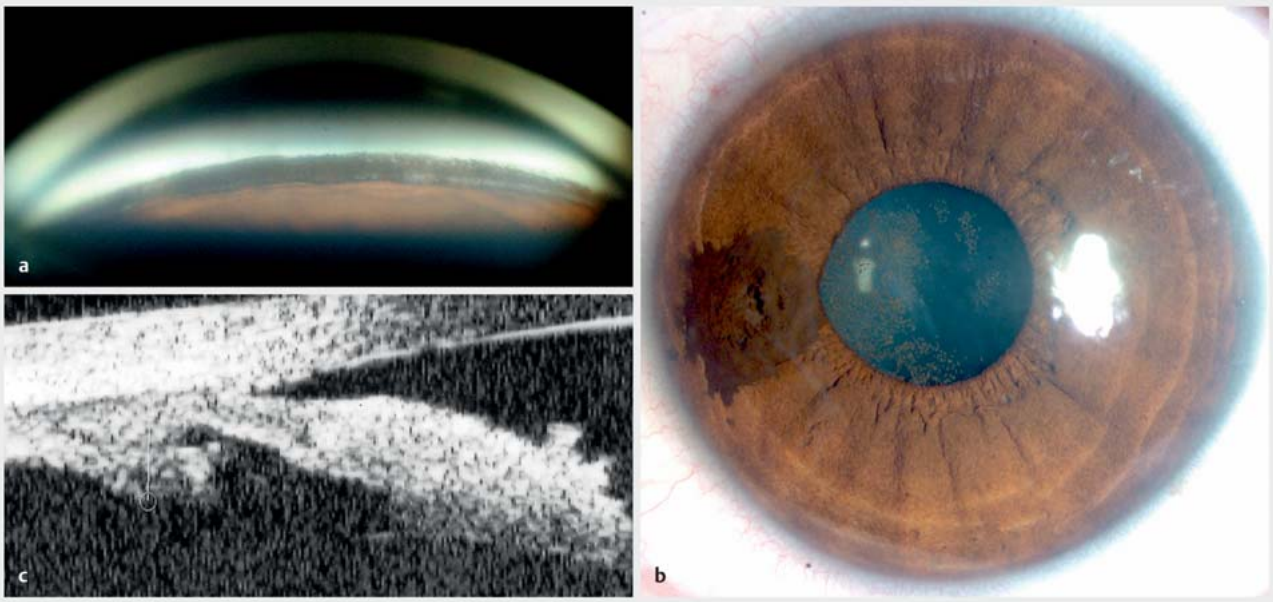
Over 8 years of follow-up, a progressive tumor atrophy was observed (▶ **Fig. 3a, b**), with a continuing decrease in thickness on UBM to 0.3 mm in 2013 (▶ **Fig. 3c**). IOP normalized (19 mmHg) without any drops since 2009. There were no signs of glaucomatous optic neuropathy.

Discussion

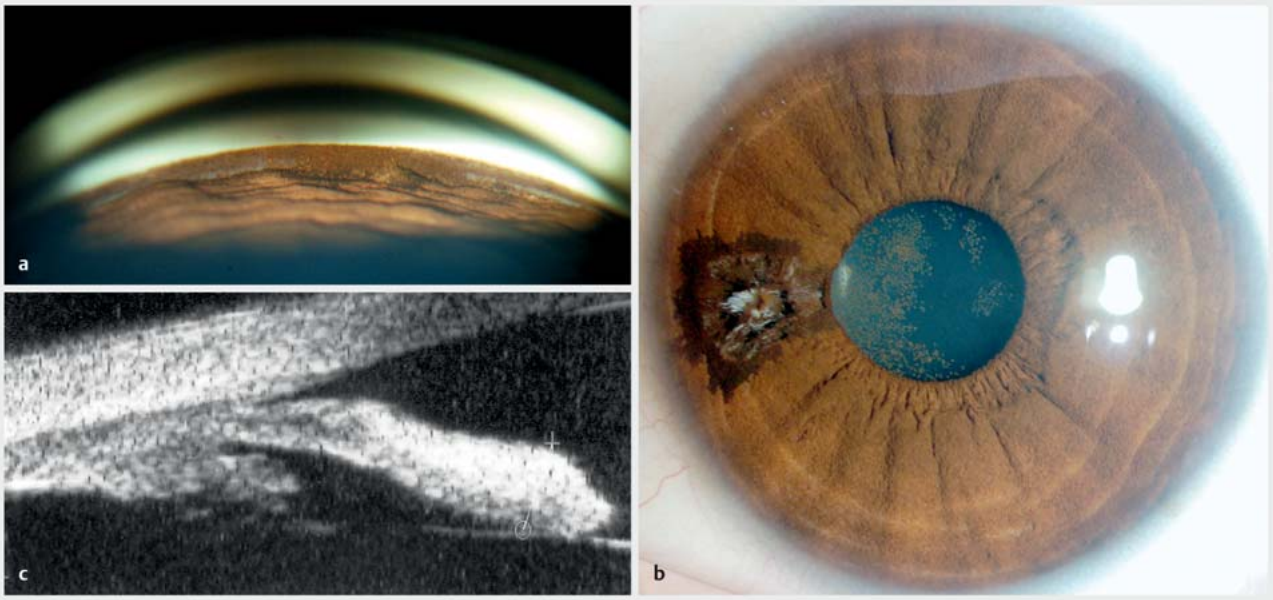
We report a case of presumed iris melanocytoma that presented a spontaneous, nearly complete regression over 8 years, associated with a transient intraocular hypertension.



▶ **Fig. 1** Initial presentation: Presumed iris melanocytoma (LE) associated with discrete pigment dispersion within the anterior chamber and iridocorneal angle. **a** Gonioscopy of the inferior quadrant. **b** Slit-lamp photography. **c** UBM: the tumor thickness is 1.4 mm.



▶ **Fig. 2** After 19 months of follow-up: The iris melanocytoma had regressed, with an overall increase in the anterior chamber and angle pigment dispersion, witnessing of a spontaneous tumor necrosis. **a** Gonioscopy of the inferior quadrant. **b** Slit-lamp photography. **c** UBM: the tumor thickness is 1.1 mm.



▶ **Fig. 3** After 8 years of follow-up: The tumor has become atrophic, with a white necrotic center, associated with a slight decrease in angle pigment dispersion. **a** Gonioscopy of the inferior quadrant. **b** Slit-lamp photography. **c** UBM: flat tumor scar, with a residual thickness of 0.3 mm.

We speculate that the mechanism of this regression represents an extreme form of spontaneous melanocytoma necrosis, of which the mechanism is still not fully understood. According to Teichmann and Karcioglu [6], melanocytomas have a very high metabolic demand because of their high melanosome content and melanin production. Minor circulatory alterations, e.g., by slow growth shutting down thin-walled vascular channels, can disrupt cell metabolism and damage cell walls and

junctions. A lack of a tumor capsule and continuous exposure to aqueous flow lead to a shedding of the necrotic tumor cells and pigment within the anterior chamber. Obstruction of the trabecular meshwork, either by cell debris, pigment, or melanin-loaded macrophages, explains the secondary ocular hypertension [6, 7].

Melanocytoma necrosis can present clinically as a "krater" at the summit of the tumor, or ultrasonographically as necrotic "cysts" within the tumor mass. However, a nearly complete regression of a previously dome-shaped tumor has never been reported and should be included in the possible natural evolution patterns of iris melanocytomas that can be observed during their periodic observation.

Conflict of Interest

The authors declare no conflict of interest.

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DOI https://doi.org/10.1055/s-0043-123642
Published online 28.3.2018 | Klin Monatsbl Augenheilkd 2018; 235: 473–475 © Georg Thieme Verlag KG Stuttgart · New York | ISSN 0023-2165