Distant Subconjunctival Recurrences following Proton Therapy for a Choroidal Melanoma with Extrascleral Extension

DOI 10.1055/a-0796-6299
Klin Monatsbl Augenheilkd 2019; 236: 422–424

Publisher and Copyright:
© 2019 by
Georg Thieme Verlag KG
Rüdigerstraße 14
70469 Stuttgart
ISSN 0023-2165

Thieme

Distant Subconjunctival Recurrences following Proton Therapy for a Choroidal Melanoma with Extrascleral Extension

Entfernte subkonjunktivale Rezidive nach Protonentherapie für ein choroidales Melanom mit extraskleraler Extension

Background

Orbital recurrences after proton therapy for uveal melanoma are rare. They either occur in cases with an extrascleral extension at presentation [1,2] or following transvitreal surgery or a transscleral biopsy [3,4]. The former tends to be located outside the scleral wall facing the tumor scar and the latter at the site of scleral entry. We report our first case since 1984 of a double distant subconjunctival recurrence from a choroidal melanoma with an extrascleral extension at presentation, treated with proton therapy and without a biopsy procedure.

History and Signs

An 80-year-old gentleman was referred because of an inferotemporal choroidal melanoma (LE) in June 2016 (▶ **Fig. 1 a**). On B-scan ultrasonography, the dome-shaped tumor's height was 5.4 mm. During tantalum clip surgery, a minor extrascleral extension (D: 2.5 × 2.5 mm; H = 1 mm) was noted and managed as usual with thermocauterization of the involved vortex vein, starting distant from the extension and continuing towards the sclera. Proton therapy was applied (60 CGE in 4 fractions), adapting the collimator to include the extrascleral extension in the target volume (▶ **Fig. 1 b–d**). Eighteen months later, two new pigmented subconjunctival lesions at 9 and 12 o'clock had appeared, at a distance from the infero-temporal primary tumor scar. On UBM 35 MHz ultrasonography, no corresponding intraocular tumor mass could be revealed (▶ **Fig. 2**).

Therapy and Outcome

A general check-up excluded systemic metastases. Both the nasal lesion, adherent to the sclera, and the superior nodule, mobile, were surgically excised. Histopathology confirmed the diagnosis of a double orbital recurrence from a uveal epithelioid melanoma with the loss of BAP1 nuclear expression. An orbital MRI excluded other, more posterior recurrences. However, 6 months later, hepatic metastases were discovered on abdominal ultrasonography.

Discussion

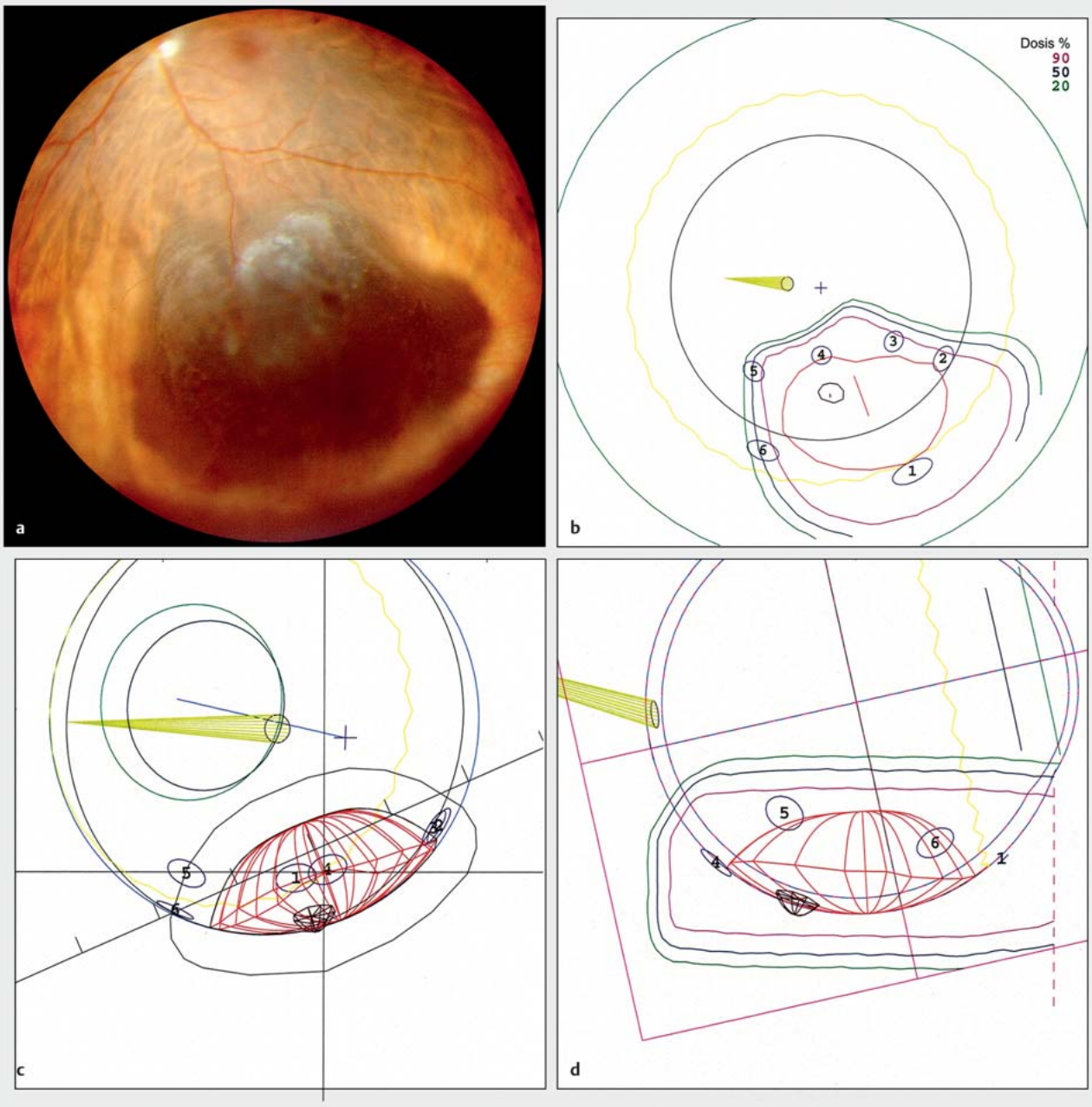
Since 1984, out of 245 uveal melanoma patients with an extrascleral extension, on a total of 5846 cases treated with proton therapy, we describe our first case of distant subconjunctival recurrences. They were diagnosed 18 months following our usual management protocol consisting of a precautious thermocauterization of the involved vortex vein, followed by proton therapy with an adapted target volume.

In the literature, only one other case of a distant orbital recurrence was described by Daiker et al. [5]. Their episcleral recurrence was on the opposite side of the globe, without any connection with the primary tumor, which had not presented an initial extrascleral extension. With the pathogenesis remaining unclear, the authors hypothesized that the recurrence had resulted from systemic blood dissemination, or lymphatic or surgical dissemination secondary to tantalum clip suturing. In our case, the episcleral recurrences were presumably caused by some malignant cells from the extrascleral extension that disseminated peroperatively and were not included in the adapted target volume.

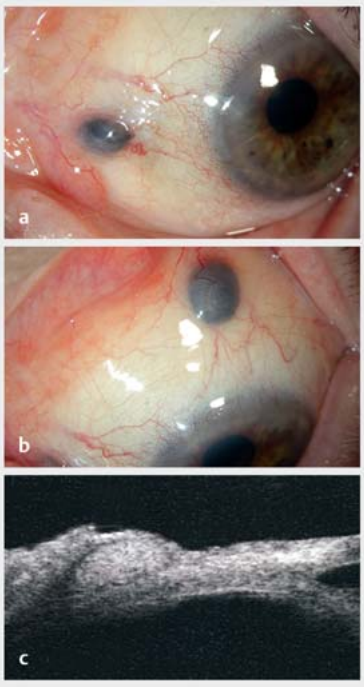
Nowadays, uveal melanomas with a circumscribed extrascleral extension can be treated conservatively, with either proton or plaque radiation therapy, depending on the size and type of the extension. In 1992, Bercher et al. of the Lausanne/PSI group reported on 41 cases with an extrascleral extension that were treated with proton therapy without any orbital recurrences being documented [6]. Bellmann et al. observed similar results at the Curie institute, where a safety margin of 2.5 mm, instead of 2 mm, was applied. They reportedly treated smaller, exteriorized tumors located anteriorly to the equator and with a height inferior to 6 mm, with 125-iodine brachytherapy [7]. Augsburger et al. and Gündüz et al. also published their results on ^{125}-I plaque therapy, with no orbital recurrences [8,9].

On the other hand, tumors with large nodular or diffuse extrascleral extensions are managed with enucleation. Adjuvant orbital radiation therapy was advocated by Hykin et al. after observing an orbital recurrence in 22% of patients without and 6% of those with orbital irradiation following enucleation [10].

In conclusion, we describe an exceptional case of distant subconjunctival recurrences from a choroidal melanoma with an extrascleral extension at presentation and management with thermocauterization followed by adapted conservative proton therapy. With the risk of a distant orbital recurrence less than 0.5%, the presence of an extrascleral extension does not require systematic adjuvant orbital irradiation. However, awareness of the possibility in cases at risk should be present during follow-up, allowing for rapid diagnosis and appropriate management.



▶ **Fig. 1** Proton therapy target volume planning for a choroidal melanoma with extrascleral extension. **a** Panoramic fundus picture of a dome-shaped infero-temporal choroidal melanoma with associated retinal detachment. **b** Schematic fundus view of the tumor base marked by six tantalum clips and distribution of the radiation isodoses around the tumor. **c** Proton beam view. The tumor target volume (in red) is surrounded by the adapted collimator with a safety margin of 2 mm around the tumor and the extrascleral extension. **d** Axial tumor view, its extrascleral extension and the radiation isodoses.



▶ **Fig. 2** Nasal and superior distant subconjunctival tumor recurrences. **a,b** Slit lamp color pictures. **c** UBM 35 MHz ultrasonography. No corresponding intraocular tumor mass can be visualized.

Conflict of Interest

The authors declare that they have no conflict of interest.

Authors

Victoria Mouvet[1], Alexandre Moulin[1], Alessia Pica[2], Ann Schalenbourg[1]

1 Jules-Gonin Eye Hospital, Department of Ophthalmology, University of Lausanne, FAA, Switzerland

2 Center for Proton Therapy, Paul Scherrer Institute, ETH Domain, Villigen, Switzerland

Correspondence

Ann Schalenbourg
Hopital ophtalmique Jules-Gonin
Adult ocular oncology unit
Av. de France 15
1004 Lausanne
Switzerland
Phone: + 41 (0) 2 16 26 81 11
Fax: + 41 (0) 2 16 26 88 89
ann.schalenbourg@fa2.ch

References

[1] Blanco G. Diagnosis and treatment of orbital invasion in uveal melanoma. Can J Ophthalmol 2004; 39: 388–396

[2] Coupland SE, Campbell I, Damato B. Routes of extraocular extension of uveal melanoma: risk factors and influence on survival probability. Ophthalmology 2008; 115: 1778–1785

[3] Caminal JM, Sanz S, Carreras M et al. Epibulbar seeding at the site of a transvitreal fine-needle aspiration biopsy. Arch Ophthalmol 2006; 124: 587–589

[4] Schefler AC, Gologorsky D, Marr BP et al. Extraocular extension of uveal melanoma after fine-needle aspiration, vitrectomy, and open biopsy. JAMA Ophthalmol 2013; 131: 1220–1224

[5] Daicker B, Zografos L, Müller O. [Homolateral episcleral metastasis or surgical seeding of a proton-irradiated ciliary body melanoma?]. Klin Monatsbl Augenheilkd 1988; 192: 579–581

[6] Bercher L, Zografos L, Egger E et al. [Treatment of exterior extension of choroid melanomas by accelerated proton beams]. Klin Monatsbl Augenheilkd 1992; 200: 440–443

[7] Bellmann C, Lumbroso-Le Rouic L, Levy C et al. Uveal melanoma: management and outcome of patients with extraocular spread. Br J Ophthalmol 2010; 94: 569–574

[8] Augsburger JJ, Schneider S, Narayana A et al. Plaque radiotherapy for choroidal and ciliochoroidal melanomas with limited nodular extrascleral extension. Can J Ophthalmol 2004; 39: 380–387

[9] Gündüz K, Shields CL, Shields JA et al. Plaque radiotherapy for management of ciliary body and choroidal melanoma with extraocular extension. Am J Ophthalmol 2000; 130: 97–102

[10] Hykin PG, McCartney AC, Plowman PN et al. Postenucleation orbital radiotherapy for the treatment of malignant melanoma of the choroid with extrascleral extension. Br J Ophthalmol 1990; 74: 36–39

Bibliography

DOI https://doi.org/10.1055/a-0796-6299
Klin Monatsbl Augenheilkd 2019; 236: 422–424

ISSN 0023-2165