

# Choroidal melanoma with massive extraocular extensions through sclera

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**Abstract:** We report a case of choroidal melanoma with massive extraocular extensions through the sclera. A 64-year-old woman reported blurred vision in her right eye. At the first visit, visual acuity was 10/20 OD. An ophthalmological examination revealed a raised choroidal mass and exudative retinal detachment in the lower retina. A diagnosis of peripheral choroidal melanoma was confirmed by additional test results. Because the tumor size was large and no systemic metastasis was found, we recommended enucleation. However, the patient refused and requested only to be followed without treatment. Seven months later, the tumor showed extraocular extensions through the sclera into subconjunctival space and she finally agreed to undergo enucleation. Histopathologic findings showed that the tumor was a mixed cell malignant melanoma of the choroid. The eye was filled with tumor cells, and the tumor had massive extraocular extensions into the orbit through the sclera and scleral emissarium vessels. The intraocular tumor was markedly necrotic, which indicated rapid growth. Choroidal melanomas can increase quickly in size resulting in extraocular extensions through the sclera.

**Keywords:** choroidal melanoma, extraocular extension, pathology

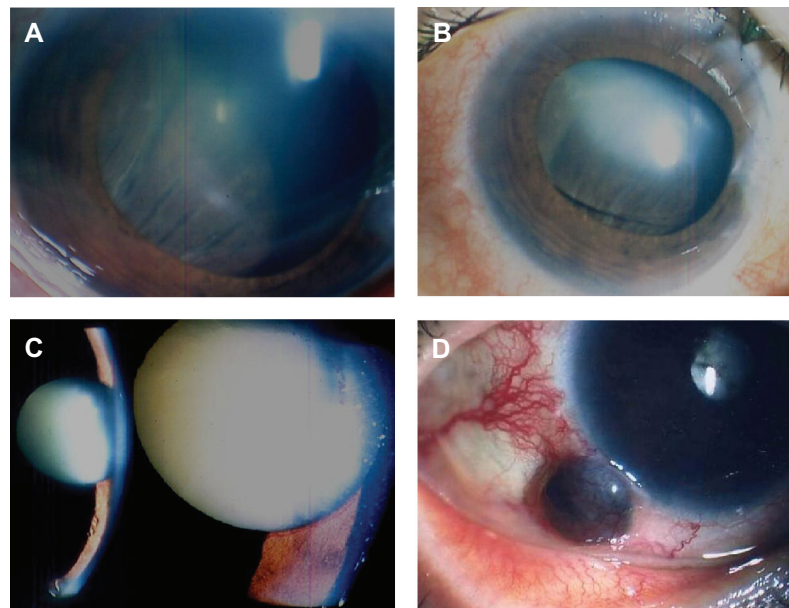
## Introduction

Recently, use of conservative therapies for choroidal melanomas, eg, irradiation and local resection, has been increasing, while enucleation is recommended only for advanced cases to improve the prognosis. Most cases of choroidal melanomas show growth into the subretinal space, and extraocular extensions are not common. Here, we report a case of choroidal melanoma with massive extraocular extensions through the sclera.

## Case report

A 64-year-old woman reported blurring of vision in the right eye that began a few months prior to visiting us. At the initial examination, best corrected visual acuity was 10/20 OD and 20/20 OS, while intraocular pressure was 19 mmHg and 21 mmHg, respectively. The left eye was normal. Ophthalmoscopy revealed a raised choroidal mass behind the lens (Figure 1A) and exudative retinal detachment in the lower retina of the right eye. Ultrasonography showed that the choroidal mass (14 × 14 mm, height 12 mm) was a solid mushroom-shaped lesion. A diagnosis of peripheral choroidal melanoma was confirmed by further examinations using computed tomography, magnetic resonance imaging, and brain perfusion scintigraphy. Systemic metastasis was not found in a thorough examination.

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**Figure 1** External photographs showing anterior segment of the eye. **(A)** Initial visit. A raised choroidal mass can be seen behind the lens. **(B)** Six weeks after first visit. The choroidal mass behind the lens has expanded to push the lens upward. **(C)** Three months after the first visit. The eye had a mature cataract, the anterior chamber has disappeared, and iris rubeosis is severe. **(D)** Seven months after the first visit. The tumor showed extraocular extensions through the sclera into the subconjunctival space.

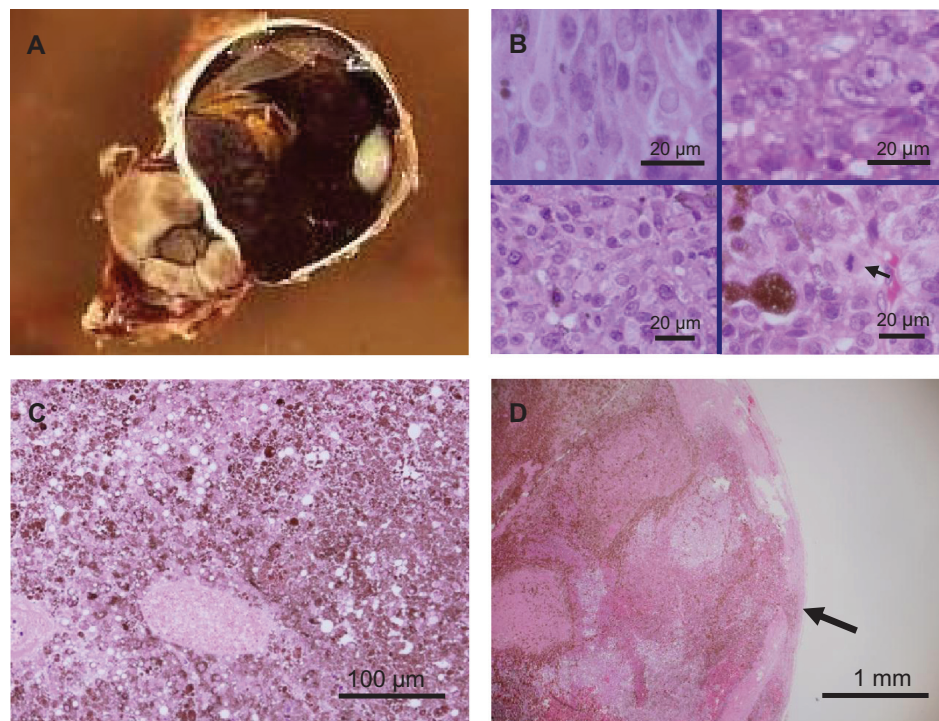
The Collaborative Ocular Melanoma Study reported that there was no difference between iodine-125 brachytherapy and enucleation in regard to 5-year survival of patients with a medium-sized choroidal melanoma, while external-beam radiation to the orbit prior to enucleation also did not improve survival as compared with enucleation alone.<sup>1</sup> Our patient consulted with a radiotherapy specialist, but neither heavy particle radiotherapy nor transpapillary thermotherapy were indicated because of the size and location of the mass. In addition, it was noted that radiation therapy might induce severe eye pain due to secondary glaucoma. Thus, enucleation was considered to be the best treatment for improving the mortality risk, which we proposed enucleation several times. However, she refused and asked to be only followed without treatment.

Six weeks after our initial examination, the choroidal mass behind the lens had expanded to push the lens upward (Figure 1B), and corrected visual acuity was reduced in regard to light perception. The anterior chamber had become quite shallow and intraocular pressure was increased to 40 mmHg OD, although the patient did not complain of eye pain. Three months later, the right eye had a mature cataract, the anterior chamber had disappeared, and iris rubeosis was severe (Figure 1C).

Seven months after the first visit, the tumor showed extraocular extensions through the sclera into the subconjunctival space (Figure 1D). The eye had no light perception

and intraocular pressure was 42 mmHg OD. Finally, after receiving informed consent from the patient, we performed enucleation of the right eye, as it was considered difficult to preserve, during which time systemic metastasis was revealed by 2-[<sup>18</sup>F]-fluoro-2-deoxy-D-glucose positron emission tomography. We did not attempt an invasive operation, such as exenteration of the orbit, because the patient was at a high risk of mortality.

Macroscopic observations showed that the enucleated eye had a massive choroidal tumor along with exudative retinal detachment and multiple extraocular extensions into the orbit through the sclera (Figure 2A). The choroidal tumor was seen as a brown-black intraocular mass with brown extraocular nodules of lobulated structures. The extraocular portion was not pigmented, while the intraocular portion was pigmented. We diagnosed the tumor as a mixed cell melanoma, because the tumor cells consisted of spindle type A, spindle type B, and epithelioid cells (Figure 2B).<sup>2</sup> The intraocular portion showed marked necrosis that indicated rapid growth of the tumor and karyokinesis (Figure 2B and C). The tumor had directly invaded the sclera (Figure 2D) and extended through the scleral emissarium vessels, but had not invaded the optic disc. In the anterior segment, the mass extended into the ciliary body and anterior chamber angle, which resulted in iris neovascularization and peripheral anterior synechia. Following enucleation, the patient underwent chemotherapy,



**Figure 2** Histopathology findings of enucleated eye. **(A)** Macroscopic appearance. The eye had a massive choroidal tumor with exudative retinal detachment and multiple extraocular extensions into the orbit through the sclera. **(B)** Higher magnification of representative histological section showing spindle type A, spindle type B, and epithelioid cells, as well as cells with karyokinesis. Upper left: spindle type A cells. Upper right: epithelioid cells. Lower left: spindle type B cells. Lower right: cells presenting karyokinesis (arrow). **(C)** Necrosis of tumor cells. **(D)** Direct extraocular extension through the sclera (arrow).

but it was not effective. She was transferred to a terminal care hospital and died about 6 months after the procedure.

## Discussion

It has been reported that the prognosis of patients with a choroidal melanoma is poor in cases with extraocular extensions,<sup>3-5</sup> with a 3-year survival rate of 37% according to Affeldt et al, and 5-year survival rates of 27% reported by Shammas et al and 52% by Pach et al.<sup>3-5</sup> Those reports also noted some factors, eg, tumor size, cell type, extraocular extension, and karyokinesis, that were important for prognosis.<sup>3-5</sup>

Our case presented with four factors related to poor prognosis, ie, size  $\geq 1.0$  cm<sup>3</sup>, presence of epithelioid cells, extraocular extensions, and karyokinesis. Immediate enucleation might have improved the prognosis because systemic metastasis was not found at the initial visit. However, the patient refused and wished to be followed only without treatment. Therefore, we observed the natural course of tumor expansion for a period of 7 months.

Recently, it has become quite rare to observe the natural expansion of a choroidal melanoma because of the availability of techniques for early detection and development of effective therapies. In conclusion, we observed rapid growth

of a choroidal melanoma along with extraocular extensions through the sclera into the subconjunctival space and orbit in the present patient.

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## Disclosure

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