Dear Editor,

Choroidal melanoma is the most common primary intraocular malignancy and has a high potential for site-specific metastasis. It is known to metastasize to the liver and lungs through hematogenous spread. However, brain metastasis is rare without a prior liver metastasis. In general, the presence of metastasis determines a patient’s prognosis and survival. Sachin AB et al. reported a case of choroidal melanoma with an isolated brain metastasis that developed following enucleation[1]. Similarly, in this report, we present a rare case of choroidal melanoma with extensive leptomeningeal metastasis that occurred a year following intensity modulated radiation therapy (IMRT) and enucleation with hydroxyapatite implantation.

A 49-year-old female visited the retina specialty clinic in September 2015 for continuous visual deterioration and metamorphopsia over the previous two months. She had received intravitreal anti-VEGF(Vascular endothelial growth factor) for exudative retinal detachment(RD) at another hospital. At initial examination, corrected visual acuity was 10/200 in the left eye with exudative RD. Orbital sonography and magnetic resonance imaging(MRI) revealed an inferior choroidal pigmented mass of 14.73 mm diameter and 7.14 mm height. No other systemic metastases were found.

Based on the Collaborative Ocular Melanoma Study(COMS), the tumor was a medium-sized choroidal melanoma, and there was no difference in the 5-year survival following treatment with either iodine-125 brachytherapy or enucleation[2]. The patient refused enucleation and was referred to the radiology department for IMRT. In October 2015, she received a total dose of 60 Gy over 10 days, and her vision subsequently deteriorated to no light perception(LP-).

After radiation therapy, exudative RD was continuously observed, and the vision continued to be LP-. It was previously reported that vitrectomy can be safely performed in eyes with previously irradiated uveal melanoma in order to prevent a patient from having to undergo enucleation. Several studies have shown that vitrectomy does not increase the risk for intraocular, orbital, or systemic dissemination of the tumor[3]. Therefore, the patient in the present case underwent a vitrectomy with oil injection on June 2016 to prevent enucleation although the visual prognosis was not favorable.

Despite close follow up for 6 months, she developed a recurred melanoma with abrupt rupture through the sclera and extraocular extension into the subconjunctival space(Fig.1 A,B). In December 2017, an emergent
enucleation was performed. After tumor free margins were confirmed by multiple frozen sections of the conjunctiva, tenon’s layer and superior rectus muscles, an 18 mm hydroxyapatite implant was placed. Histopathology showed an epithelioid cell type choroidal melanoma with a tumor-free margin around the optic nerve, despite tumor extension through the optic nerve(Fig.1 C,D). At the time of enucleation, no systemic or local metastasis was observed. In January 2019, the patient was admitted to the emergency department with vomiting and headache. MRI showed extensive leptomeningeal seeding in the brain without metastasis to the liver or lungs(Fig.1 E,F). The patient agreed to undergo palliative treatment without chemotherapy at a terminal care hospital, and she died comfortably 3 months later.

A choroidal melanoma with extraocular extension has a 3-year survival rate of 37% and a 5-year survival rate of 27%[4]. Tumor size, location, cell type, extraocular extension, and age at initial presentation determine the prognosis. The poor prognostic indicators in the present case were tumor size > 1.0 cm³, presence of epithelioid cells, and extraocular extension.

The high dose radiation of IMRT could have caused scleral extension of the tumor by weakening the scleral tissue[5]. We assume that combined vitrectomy and IMRT could have triggered extraocular extension. Additionally, we consider microscopic extension of tumor into the orbit that might have been missed during enucleation despite ensuring tumor free margins on frozen sections. The hydroxyapatite implant might have impaired early detection of the recurrence and prevented quick tumor growth. A subtotal exenteration without implant might have improved the prognosis either by facilitating complete microscopic excision or by not inhibiting early detection of the recurrence. Finally, adjuvant orbital radiation following enucleation should have been considered for undetected microscopic metastasis.

To conclude, we present a rare case of choroidal melanoma with rapid and extensive leptomeningeal metastasis following IMRT and enucleation. Thus, in patients with high metastatic potential, we recommend early and aggressive surgical intervention combined with adjunct therapies and regular follow up with brain imaging to detect early spread.

Taehwan Kim, Boram Kim, Jae Sang Ko, Jin Sook Yoon,
Department of Ophthalmology, Institute of Vision Research, Severance Hospital, Yonsei University College of Medicine, Seoul, Korea E-mail (Jin Sook Yoon): YOONJS@yuhs.ac

Conflict of Interest
No potential conflicts of interest relevant to this article are reported.
References


Fig. 1. (A,B) Slit-lamp photograph of recurred choroidal melanoma with abrupt rupture through the sclera and extraocular extension into the superior subconjunctival space. (C) Gross photograph of specimen after enucleation. (D) Histologic findings of epitheloid cell type choroidal melanoma (hematoxylin and eosin stain, x200). (E,F) Magnetic resonance imaging findings of extensive leptomeningeal seeding in the brain.