

## Case Report

# Massive Extraocular Extension of Choroidal Melanoma

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## Keywords

- Choroidal melanoma
- Extraocular extension
- Subtotal exenteration
- Dermis-fat graft

## Abstract

We report two cases of choroidal melanoma with massive extraocular extension treated in East Tallinn Central Hospital Eye Clinic. Extraocular extension of intraocular melanoma is rare and its management is controversial. The medical records of two patients with histologically proven choroidal melanomas with large orbital extension were analyzed retrospectively. None of the patients showed any systemic metastasis at the time of diagnosis. An eyelid preserving subtotal orbital exenteration combined with socket reconstruction using autologous dermis-fat graft was performed on both patients with very good cosmetic results. The patients survived 3.5 and 9 years respectively after the surgery with no orbital recurrence. They both died of hepatic metastatic disease.

## INTRODUCTION

Uveal melanoma is the most common primary intraocular malignant tumor in adults. The incidence ranges from 5.3 to 10.9 cases per million population, with about 90% arising from the choroid, 5% involve the ciliary body and 5% the iris [1]. The majority of choroidal melanomas show growth into the subretinal space and extraocular extensions are rare [2-4]. Uveal melanoma disseminates hematogenously with the most common site of metastases (93%) being the liver [5]. Liver metastases develop within 15 years after the initial diagnosis and treatment in approximately 50% of patients with choroidal melanoma and the metastatic disease remains the leading cause of death [1]. A life-long follow-up of patients is important as metastases of choroidal melanoma can occur decades after treatment [6]. After detection of metastases 80% of patients die within 1 year and 92% within 2 years [7].

The prognosis of patients with choroidal melanoma is mostly poor in case of extraocular extensions regardless the treatment [2-4]. The 5-year survival rate of 27% was reported by Shammas et al. [4], and 52% by Pach et al. [2]. The aforementioned reports also state the prognostic markers having negative effect on patient survival, e.g. intraocular tumor size, epithelioid cell type, advanced age and extraocular extension.

The management of uveal melanomas with extraocular extension has undergone considerable changes over the last decades with more conservative methods being used depending upon the type of extraocular extension and how and when the extrascleral extension was detected [8,9]. There is still a lack of consensus concerning the optimal management of orbital invasion in uveal melanoma.

## CASE PRESENTATION

We report two cases of choroidal melanoma with histologically

proven massive orbital extension. The patients were treated and seen by the third author AK from 1998-2010.

## Case report No. 1

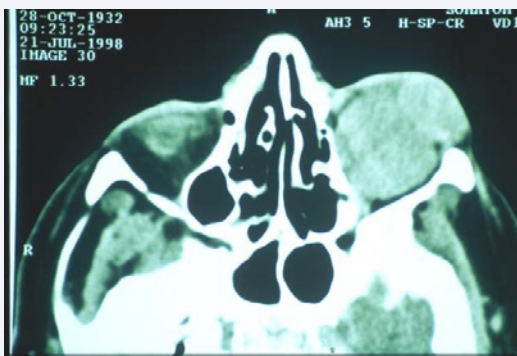
A 65-year-old woman reported gradual painless vision loss from her left eye during for a couple of years prior to the presentation. She denied any previous ocular history. At presentation she showed moderate proptosis of her left eye. The best corrected visual acuity was 1.0 OD and no light perception OS. Her left eye showed slight irritation, the cornea was deformed and under the slit lamp a dark grayish mass was seen right behind it. Her right eye was normal (Figure 1). CT scan revealed a large intraocular mass filling the left eye with an even larger extraocular extension into the left orbit (Figure 2). No systemic metastases were found at the time of diagnosis. A subtotal exenteration of the left orbit with preserving the eyelids was performed. A tumor of dark grayish-greenish color was filling the eye with an extrascleral lobulated extension into the orbit (Figure 3). Histologically the tumor was mixed cell type malignant melanoma of the choroid. It also showed an invasion of the optic nerve (Figures 4-6). 2 weeks after the exenteration an autologous dermis-fat graft taken from the patient's left lower abdominal region was transplanted into the left orbit. One month after the surgery the patient received individually made cover shell prosthesis (Figure 7). The patient had regular follow-up visits after the surgery and showed no orbital regrowth. The patient survived 9 years after the first visit and died due to metastatic disease of the liver.

## Case report No. 2

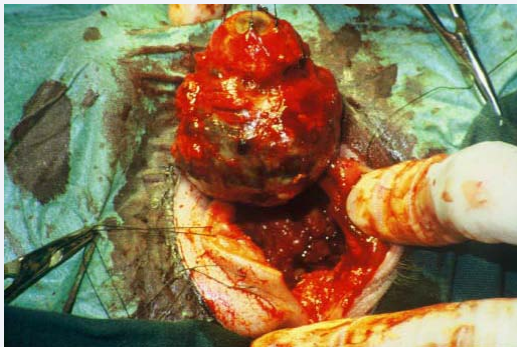
A 72-year-old woman presented to a local ophthalmologist with a chief complaint of painful left eye with no light perception. She had gradually lost vision about 1.5 years before presentation



**Figure 1** Patient No.1. External photograph of a 65-year-old woman with large intraocular melanoma.



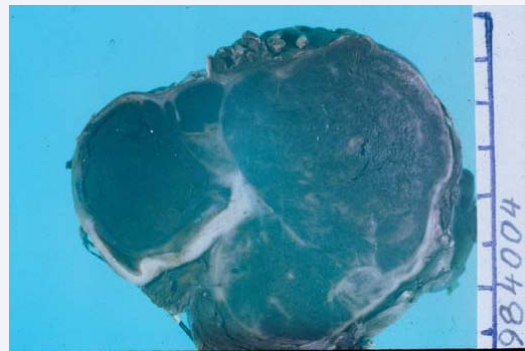
**Figure 2** Patient No.1. Axial CT image demonstrating intraocular melanoma with massive extension into the left orbit deforming the globe.



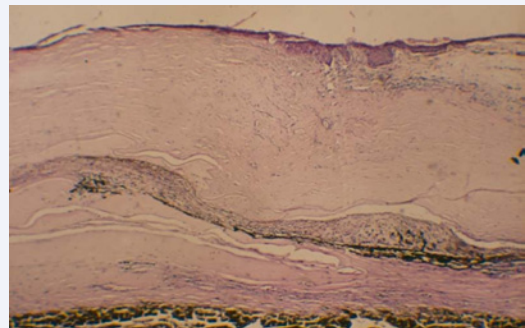
**Figure 3** Patient No.1. Exenteration of the left eye with subtotal orbital tissue removal.

but as she lived in a rural area and the eye wasn't painful initially she didn't seek for any medical attention until her left eye became increasingly proptotic and painful. The right eye was normal. She also denied any prior ocular history. As the left eye showed severe corneal exposure at the initial visit, temporary tarsorrhaphy had been performed by her local ophthalmologist already (Figure 8). CT scan showed an intraocular mass filling the left eye with massive extraocular extension into the left orbit (Figure 9). At the time of diagnosis no metastases were detected. A subtotal left orbital exenteration with preserving the eyelids was performed.

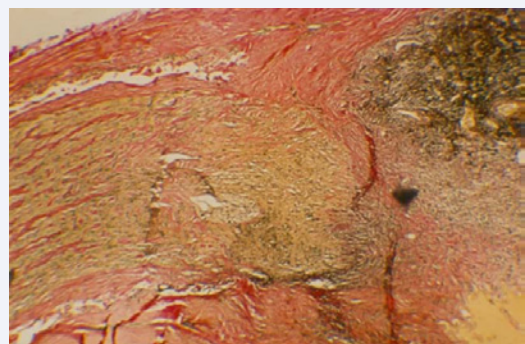
Histopathological examination confirmed the diagnosis of mixed cell type choroidal melanoma with massive extraocular extension through sclera into the left orbit. An autologous dermis-fat graft was transplanted into the left orbit 2 weeks after exenteration (Figures 10-12). The patient was under close follow-up and survived 3.5 years after the exenteration without any orbital recurrence. Eventually she started complaining of epigastric pain and ultrasonography and CT scan revealed hepatic metastases. The patient died 4 months after the diagnosis of liver metastases was made.



**Figure 4** Patient No.1. Histopathologic specimen showing the macroscopic appearance of large intraocular and massive orbital portion of the tumor.



**Figure 5** Patient No. 1. Histopathology findings, higher magnification showing cornea and iris pressed together.



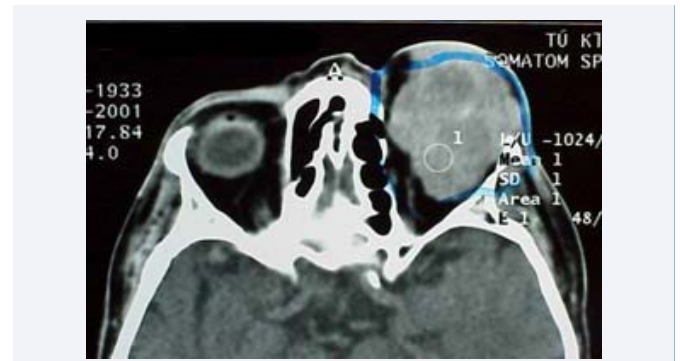
**Figure 6** Patient No.1. Histopathology findings. Higher magnification showing intraocular melanoma cells' invasion into the optic nerve.

## DISCUSSION

The two cases presented have much in common. Both patients presented with advanced choroidal melanomas with massive extraocular extension into the orbit. The extraocular extension of choroidal melanoma is rare and the management of such cases has still remained a subject of controversy. The presented cases were both managed in a similar manner. The patients underwent subtotal orbital exenteration preserving the eyelids followed by socket reconstruction using an autologous dermis-fat graft transplant. Individually made cover shell prosthesis was fitted thereafter.

The eyelid sparing technique of orbital exenteration is indicated mainly for advanced primary orbital malignancies and for certain secondary orbital malignancies such as orbital extension of uveal melanoma. Advantages of the eyelid-sparing method are more rapid healing and earlier fitting of prosthesis [8]. The advantage of dermis-fat graft lies in its ability to replace the orbital volume and maintain the fornix and conjunctiva [9]. Autogenous deepithelialized skin with subcutaneous fat is implanted into the socket and dermis is sutured to the periosteum [10].

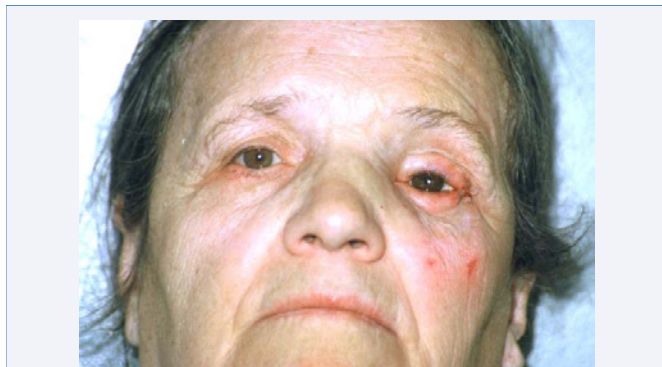
Although both patients had no clinical evidence of systemic metastasis at initial presentation, they had several other factors related to poor prognosis, i.e. the presence of epithelioid cells,



**Figure 9** Patient No.2. Axial CT showing intraocular melanoma with large extension in to the left orbit.



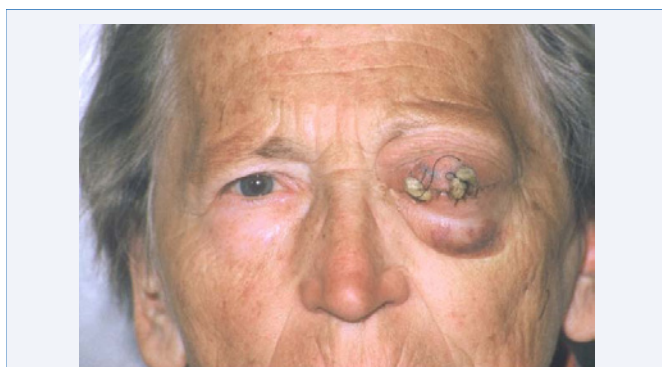
**Figure 10** Patient No.2. Histopathologic specimen after sub total orbital exenteration.



**Figure 7** Patient No.1 3 years after surgery with good cosmetic result wearing cover shell prosthesis on the left.



**Figure 11** Patient No.2. after autologous dermis-fat graft transplantation.

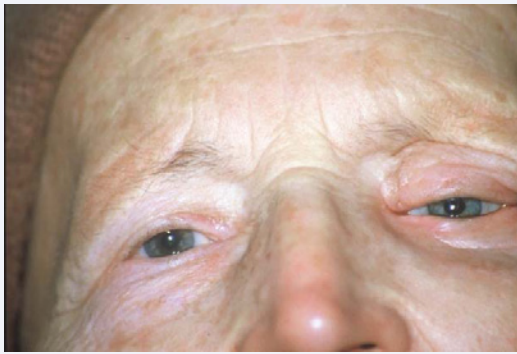


**Figure 8** Patient No.2. External photograph of a 72-year-old woman with choroidal melanoma with extraocular extension in to the left orbit, tarsorrhaphy hasbeen performed due to corneal exposure.

tumor size  $\geq 1.0 \text{ cm}^3$  and extraocular extension. The available data is not adequate to determine whether exenteration is beneficial, neutral or detrimental to the mortality rate in patients with extrascleral extension of choroidal melanoma [11,12].

In our cases the subtotal orbital exenteration preserving the eyelids and autologous dermis-fat graft implantation with subsequent individual cover shell prosthesis resulted in complete local control and very good cosmetic results.

As life-time follow-up for patients treated for choroidal melanoma is needed, both patients were screened for hepatic



**Figure 12** Patient No.2. 2 years after treatment wearing cover shell prosthesis on the left.

metastases by ultrasonographic liver imaging and biochemical liver function tests every 6 months. They survived 9 and 3.5 years respectively without any orbital recurrence and died of metastatic disease. We presume a radical exenteration would not have improved the prognosis or survival of the patients. These cases also illustrate our still insufficient knowledge concerning the prognostic factors of choroidal melanoma. Although the patients showed remarkable resemblance concerning both the disease and treatment, and none of them showed any systemic metastases at the time of initial visit, they both died of hepatic metastases with survival difference of 5.5 years.

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