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CASE REPORT

Orbital melanoma

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Abstract: Primary or secondary orbital melanomas are extremely rare tumors; they represent less than 1 % of primary orbital neoplasms. Over 90 % of primary orbital melanomas arise from melanocytes (congenital ocular melanosis, oculodermal melanosis). In the Department of Ophthalmology, Comenius University, Bratislava, in period 2001–2007, a total number of 79 patients with uveal melanoma (dg. C69) were treated with radical surgery, combined techniques or isolated stereotactic radiosurgery, 45 (57 %) patients with ciliary body and choroidal melanomas were treated with primary enucleation, 34 (43 %) patients were managed with combined techniques (parsplana vitrectomy with endoresection, brachytherapy plus stereotactic radiosurgery) or with "conservative treatment", namely with stereotactic radiosurgery alone. In group of melanoma patients, primary orbital melanoma was present only in 1 case (1 %) *(Fig. 4, Ref. 5)*. Full Text in free PDF *www.bmj.sk.*
Key words: orbital melanoma, rare tumors, neoplasms, stereotactic surgery.

Primary or secondary orbital melanomas are extremely rare tumors; they represent less than 1% of primary orbital neoplasms. They belong to a heterogeneous group of pigmented tumors. The prognosis is very poor and the tumor character can present diagnostic challenges in the beginning. Since they may cause a protrusion of the eye, the management of treatment leads almost in all cases to exenteration.

Material

In the Dpartment of Ophthalmology, Comenius University., Bratislava, in period 2001–2007, 79 patients with uveal melanoma (dg. C69) in total were treated by radical surgery or combined techniques or isolated stereotactic radiosurgery. From this group, 44 (56 %) patients with ciliary body and choroidal posterior uveal melanomas were treated with enucleation. Other group of 34 (43 %) patients with uveal melanoma (posterior, ciliary body) were managed with combined techniques (pars plana vitrectomy with endoresection, brachytherapy plus stereotactic radiosurgery) or with "conservative treatment", namely with stereotactic radiosurgery alone. In group of melanoma patients, primary orbital melanoma was present only in 1 case (1 %). Relapse of melanoma in the orbit after enucleation in a three-year interval was in 1 case (1 %). We did not mention a relapse in the orbit in patients with uveal melanoma treated with stereotactic radiosurgery.

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Case 1

A 78-year-old man was sent by an ophthalmologist to our department for right-eye cataract operation due to low visual acuity. The patient had a painless proptosis of 2 mm with no diplopia. He reported no problems in the past and had never visited an ophthalmologist before. He "could see with the other eye". The visual acuity of the right eye was on a level of ability to count fingers, the left one yielded 6/9 of Snellen chart. Intraocular pressure was normal, the ultrasound A and B scan examinations showed a large tumor mass in the posterior pole and also in retrobulbar space.

The patient was without any other symptoms; extensive systemic workup including chest radiography, abdominal imaging, and liver tests were negative. The clinical examination showed the tumor to be in T4 stage of uveal melanoma. A radical surgical attitude was indicated in form of enucleation plus exenteration (Figs 1–3).

The first step of operation was focused on enucleation of the right eye with peroperative biopsy of the tumor mass. Since the biopsy showed a malignant melanoma, the operation was finished with a total exenteration of the orbit (with eyelids).

Eighteen months after exenteration, the patient's status is good and with no presence of distant metastases. He undergoes systemic workups in 6 month intervals, including chest X-ray, abdominal imaging, computed tomography of brain and orbit. There are no signs of paranasal, ethmoidal extension of the tumor. PET (positron emission tomography) to check the presence of distant metastases was not performed because the patient did not approve. A relapse of solid multiple melanoma nodules is present in the wall of the orbit (nasal, temporal margin) while the latter nodules are excised in three-month intervals and histopathologically verified as malignant melanoma (spindle cells of

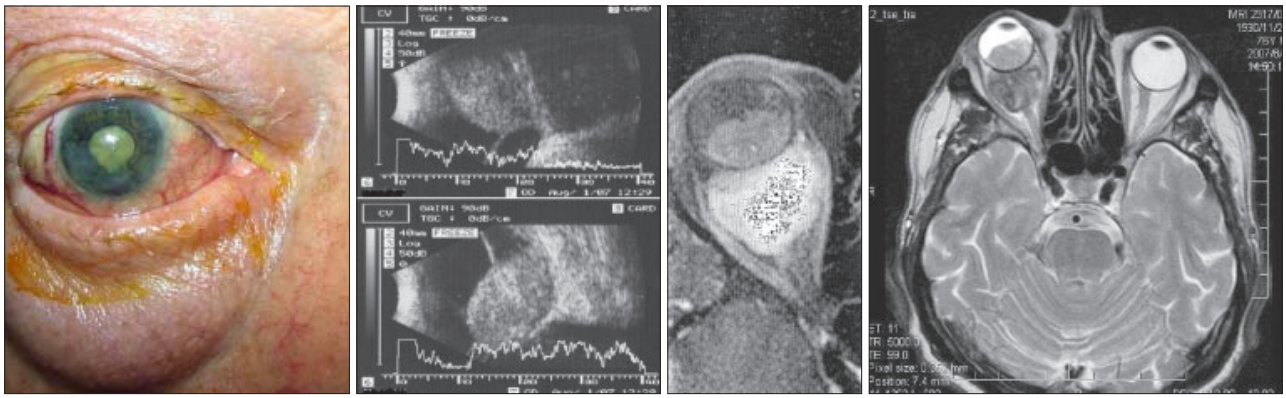

Fig. 1. a) Clinical findings before therapy, mature cataract, hyphaema, neovascularisation of the iris, b) Ultrasound A, B modes, c) Computed tomography and magnetic resonance revealed a large well-circumscribed enhancing orbital tumor mass with intrabulbar and retrobulbar location, d) Magnetic resonance findings.

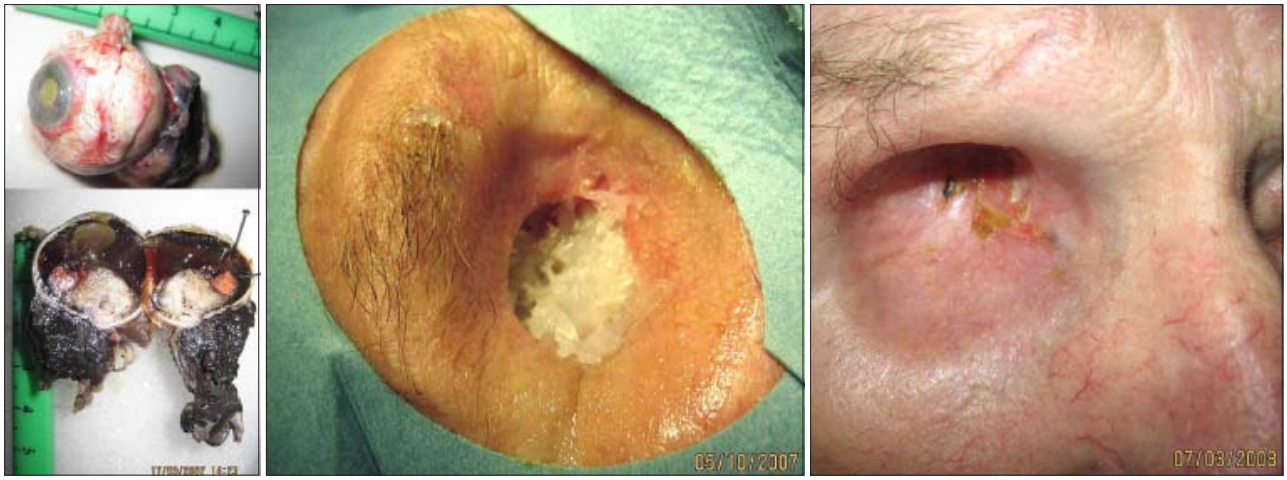

Fig. 2. a) Right orbital specimen with an enucleated eye. The tumor is inside the eye globe. The volume of tumor mass is over 2.5 cm^3, the volume of the solid retrobulbar and intraconal tumor mass is over 2.6 cm^3. Histologically, both parts of the tumor contained spindle cells of mixed type while the cells stained positively for HMB45 and S100. ICD 8720/3, b) Patient one month after the exenteration of orbit, c) Half year after the exenteration of orbit; solid subcutaneous nodules are present on the margin and the nasal part of the orbit.

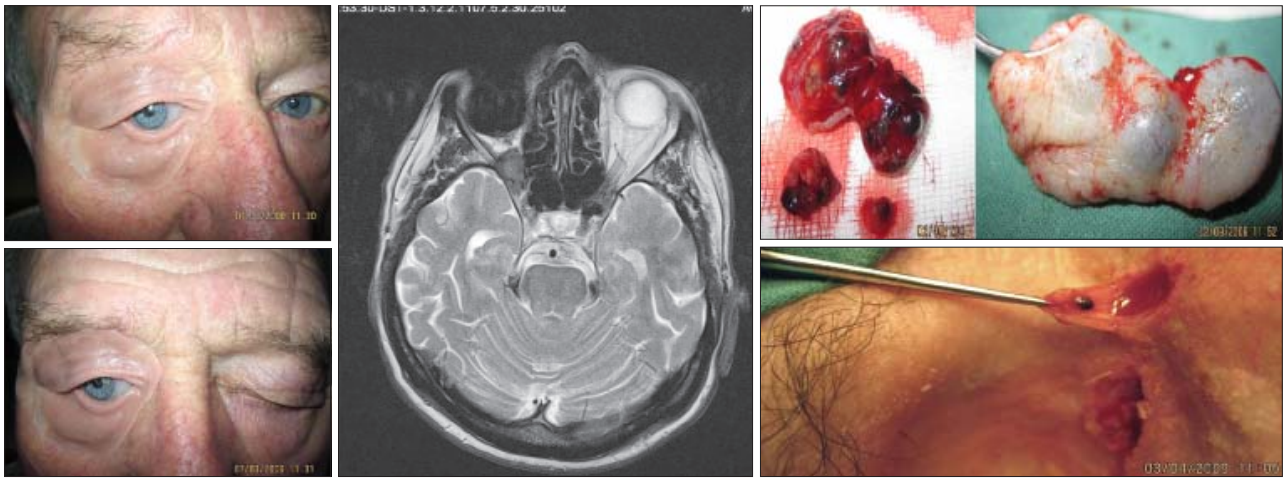

Fig. 3. a) The patient has an individual epithesis (prosthesis) of the right orbital side, b) Magnetic resonance imaging half year after exenteration, without progression to ethmoids, c) One year after radical operation; an extension of relapsed melanoma nodule was present on the temporal margin of orbit and in the nasal part. Excision of multiple melanoma nodules twelve months after exenteration, d) Excision of relapsing subcutaneous orbital melanoma – melanoma nodules – eighteen months after the exenteration of orbit.

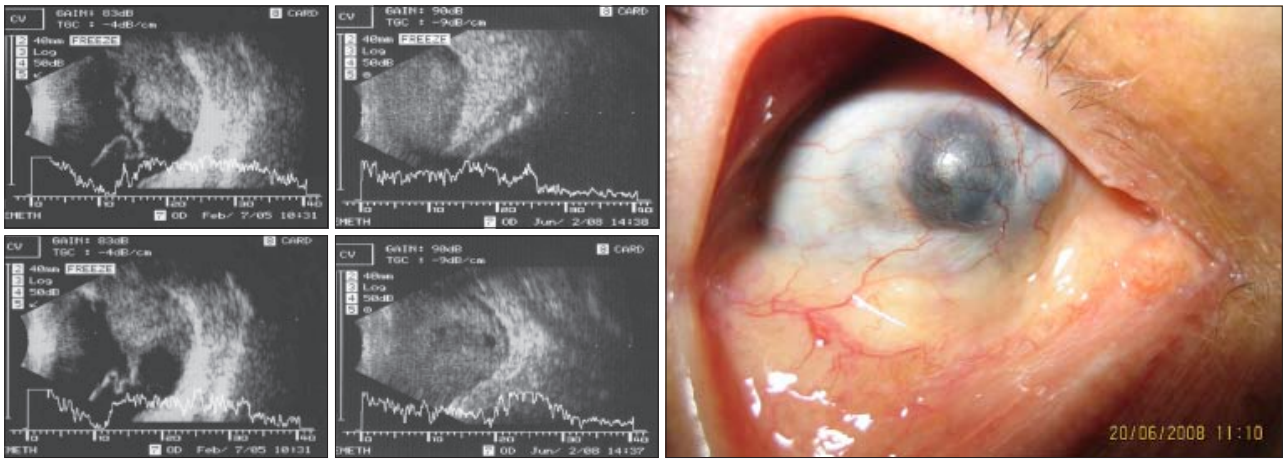

Fig. 4. a) Ultrasound A, B modes in 2005 before enucleation, b) Ultrasound of the orbit in A, B modes in 2008, c) Clinical findings of the patient with tumor mass in the orbit three years after enucleation.

B or mixed type). There are no signs of inflammation, extraorbital extension as well as no presence of distant metastases.

Case 2

A 79-year-old male patient underwent enucleation in 2005 due to malignant uveal melanoma in stage T2 of type A of spindle cells, with no presence of extrascleral expansion (Fig 4a). In 2008, there was a solid tumor mass present in the orbit of volume over 2.5 cm^3 (Figs 4b, 4c). In this period, distant hepatic metastases appeared and the patient died in 2009.

Comment

Primary orbital melanoma can has an extreme tendency to appear *de novo*, as well as primary optic nerve melanoma, which can occur due to malignant transformation of an optic disc melanocytoma. Although rare, it should be considered in cases of rapidly progressing proptosis in young Caucasian patients, even where no predisposing ocular disease is present (1). Although uveal melanoma is actually the most common intraocular tumor of adults, the incidence of orbital melanoma is not known (2). The incidence of orbital melanoma reported in literature is about 1% of all uveal melanomas, as is the case in our report. The incidence of orbital melanoma after brachytherapy or radiosurgery of intraocular melanoma is extremely rare (3).

The mortality rate in patients with orbital melanoma exceeds that of 1/3 of patients in the first year after the radical surgery. The approach toward the treatment is very individual in every patient; radical surgical treatment is known to be combined with local radiotherapy or in some patients with chemotherapy (4). The treatment is individual depending on the lesion but also on the internal status of patients, and of course on the presence of distant metastases at the moment of diagnosis. The therapy guidance is based on observing the patient, local resection or radical surgical techniques combined with radiotherapy, immunotherapy or chemotherapy (5). The approach depends also on prognostic factors (cell type, mitotic count). First-step examination by ultrasound (eye and orbit) is important for every patient with mature cataract sent for cataract operation, as was the case in our patient. It is also important not to overlook serious intraocular or possibly expanding retrobulbar lesions.

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