



UVEAL TUMORS

Late isolated brain metastasis following enucleation for choroidal melanoma

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Ophthalmic Surg Lasers Imaging. 2005 Mar-Apr;36(2):151-4.

Brain metastases from choroidal melanoma are rare and usually have a grave prognosis. A case of successfully treated late isolated brain metastasis from choroidal melanoma is described. A 35-year-old man presented with epileptic seizures of recent origin, 9 years following enucleation for choroidal melanoma. Imaging studies revealed a lesion of the right frontal lobe that was surgically removed. Results of pathologic examination were compatible with metastatic choroidal melanoma. The patient is asymptomatic 5 years postoperatively. Late isolated brain metastases from uveal melanoma may be treatable by local resection. Close, lifelong follow-up is required to diagnose and aggressively treat metastatic disease.

Ultrasound biomicroscopy features of iris and ciliary body melanomas before and after brachytherapy

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BACKGROUND AND OBJECTIVE: To determine ultrasound biomicroscopy (UBM) features of iris and ciliary body melanomas before and after brachytherapy.

PATIENTS AND METHODS: Four uveal anterior melanoma cases undergoing brachytherapy were retrospectively studied. All cases were examined by UBM prior to treatment and repeatedly after treatment.

RESULTS: Before brachytherapy, UBM examination showed a solid mass in the iris, ciliary body, or both in all four cases and allowed its characterization, sizing, and positioning. Two cases had a pigmented scleral lesion corresponding to the tumor location, but UBM did not detect any scleral infiltration at those sites. After brachytherapy, all lesions showed progressive decrease in size and progressive attenuation of their limits. Internal reflectivity was variable. Complications related to brachytherapy were demonstrated, including cataract, peripheral anterior synechiae at tumor location, and secondary scleral thinning.

CONCLUSION: UBM played an important role as a complementary diagnostic method for anterior uveal melanomas, particularly ciliary body melanomas, but also allowed therapeutic planning (brachytherapy or surgery) and follow-up after treatment.

Free keratin and dermoid cyst of the iris

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No abstract available.

Uveal melanocytomas: genetic comparison with uveal and dermal melanomas

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OBJECTIVE: Melanocytomas of the eye are typically benign tumors that may be associated with nevi and melanomas. In this study, we assessed the genetic data of melanocytomas and compared them with nevi and melanomas of both the eyes and the skin.

DESIGN: We microdissected 8 melanocytomas, 13 uveal melanomas, and 10 cutaneous melanomas and analyzed loss of heterozygosity markers on chromosome bands 1p36, 6q22-23.3, 9p21, and 10q23, which represent genetic loci associated with advanced dermal melanocytic lesions.

RESULTS: There was no loss of heterozygosity in any of the melanocytomas. However, many loss of heterozygosity events were found in uveal and cutaneous melanomas, most frequently involving chromosome 1 damage followed by chromosome 9 and 10 alterations.

CONCLUSION: Based on the absence of loss of heterozygosity in melanocytomas, specifically the locus that is lost most often in dysplastic nevi of the skin, we conclude that melanocytomas represent an entity that is different from melanomas or may be similar to that of dermal benign nevi.

CLINICAL RELEVANCE: Our results confirm that melanocytomas represent nonaggressive lesions that do not demand radical surgery.

"Finger-tip" cryoprobe assisted enucleation

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Am J Ophthalmol. 2005 Mar;139(3):559-61.

PURPOSE: To report the use of a new cryotherapy probe to induce proptosis during enucleation surgery.

DESIGN: Interventional case report. **METHODS:** Two patients with uveal melanoma and secondary glaucoma were treated by enucleation. A large surface area (70 mm²), spatulated, end-freezing cryotherapy probe ("Finger-tip" probes, MIRA, Inc.) was used to induce proptosis during optic nerve transection.

RESULTS: This new probe offers homogeneous freezing over a relatively large surface area. This new cryoprobe was used to create a large cryo-adhesion on the cornea, for an excellent purchase of the eye during enucleation surgery. Using this adhesion, the eye was lifted, enabling transection of the optic nerve.

CONCLUSIONS: This report includes photographs of the cryoprobe, the corneal cryo-adhesion, and describes its use for traction during enucleation surgery.

Iris melanocytoma: clinical features and natural course in 47 cases

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Am J Ophthalmol. 2005 Mar;139(3):468-75.

PURPOSE: To describe the clinical features, natural course, management and histopathologic features of iris melanocytoma. **DESIGN:** Single-center retrospective case series.

METHODS: Forty-seven consecutive patients (47 eyes) with iris melanocytoma. Data regarding patient and tumor features were analyzed for their impact on the main outcome measures using univariate and multivariate regression models. Kaplan-Meier estimates were used to analyze the main outcomes as a function of time. Increased intraocular pressure (IOP), tumor seeding, and tumor growth were the outcome measures.

RESULTS: Associated findings at initial presentation included iris stromal seeds in 20 patients (43%), and anterior chamber angle seeds in 12 (26%). Intrinsic vascularization and sector cataract were not seen in any eyes. The management at presentation included observation in 39 patients (83%), tumor removal by sector iridectomy/iridocyclectomy in 7 (15%), and enucleation for blind painful eye with secondary increased IOP in 1 (2%). The diagnosis was confirmed by histopathologic examination in 11 patients (23%). The mean follow-up was 58 months. Using Kaplan-Meier estimates, clinical evidence of growth was observed in 23% at 5 years, 48% at 10 years, and 74% at 15 years. New tumor seeds developed in 34% at 5 years, 63% at 10 years, and 75% at 15 years. Increased IOP was observed in 11% at 5 years, 11% at 10 years, and 55% at 15 years.

CONCLUSIONS: Iris melanocytoma represented only 3% of all iris nevi. Related iris stromal and anterior chamber angle seeds were common, and secondary glaucoma occurred in 11% at 5 years. Growth was observed in 23% at 5 years but no malignant transformation was found.

Multiple bilateral choroidal metastatic tumors from a small-cell neuroendocrine carcinoma of unknown primary site

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PURPOSE: To report one case of multiple and bilateral choroidal tumors from a poorly differentiated small cell neuroendocrine carcinoma of unknown primary.

METHODS: The case of a 30-years-old white female who developed multiple and bilateral choroidal tumors from a poorly differentiated small cell neuroendocrine carcinoma of unknown primary is presented.

RESULTS: The patient had a disseminated disease and died 6 months after. The oncologic work-up, including physical examination, laboratory and radiographic study, fails to identify the primary site.

CONCLUSIONS: Intraocular involvement from a poorly differentiated small cell neuroendocrine carcinoma of unknown primary has not yet reported. We describe this case together with a review of the literature.

Iris and anterior chamber angle neovascularization after iodine 125 brachytherapy for uveal melanoma

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PURPOSE: Iris neovascularization (INV) and anterior chamber angle neovascularization after radiotherapy for uveal melanoma may lead to neovascular glaucoma and enucleation. However, neovascularization of the anterior ocular segment may respond favorably to treatment with panretinal photocoagulation. The purpose of this study was to evaluate the frequency, interval to development, and predisposing factors of anterior ocular segment neovascularization following iodine 125 (I125) brachytherapy for uveal melanoma.

DESIGN: Retrospective, interventional, consecutive case series.

PARTICIPANTS: Sixty-five patients (65 eyes), consecutively treated with I125 brachytherapy for uveal melanoma from 1995 through 2000 and followed up after radiation therapy for 24 months or more.

METHODS: Clinical findings and ultrasonography characteristics as well as treatment parameters were analyzed.

OUTCOME MEASURES: The frequency of INV was determined and the interval to development of INV as well as the predisposing factors were analyzed statistically.

RESULTS: In 15 of 65 eyes (23%), INV was detected after I125 brachytherapy at a mean +/- standard deviation of 26.66 +/- 11.63 months (median, 24 months; range, 9-48 months). Risk factors displaying the stronger correlation with INV were greater maximal tumor height ($P < 0.01$), greater tumor vascularity ($P < 0.01$), and disinsertion of horizontal rectus muscles ($P = 0.01$).

CONCLUSIONS: After I125 brachytherapy for choroidal melanoma, INV developed in 23% of eyes and was correlated with larger tumor size, greater tumor vascularity, and disinsertion of a horizontal rectus muscle.

Prognostic value of serum vegf in melanoma patients: a pilot study

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Anticancer Res. 2004 Nov-Dec;24(6):4255-8.

BACKGROUND: Vascular endothelial growth factor (VEGF) is involved in angiogenesis. We investigated the association of VEGF serum levels (pre-treatment and follow-up) with outcome in patients with melanoma.

PATIENTS AND METHODS: Serum levels of VEGF in melanoma patients at diagnosis and during follow-up were analysed with enzyme-linked immunoassays. Patients were followed up with physical examination and ultrasound scans of the liver every three months and thorax X-ray annually. The VEGF serum level was evaluated six-monthly.

RESULTS: From February 1996 to February 2000, 33 patients were enrolled. Ninety-two serum blood samples were collected. Patients had a median age of 60 years (range 32-82). Twenty patients were males, 13 females. One patient presented with stage IA disease, 2 with stage IB, 11 with stage IIA, 4 with stage IIB, 8 with stage III and 5 with stage IV. Two patients were affected by uveal melanoma. The melanomas were predominantly located at the extremities or trunk (26/33). The median serum level of VEGF at diagnosis was 249 ng/ml (minimum: 9 ng/ml, maximum: 1215 ng/ml). The median survival of all 33 patients was 45.1 months. The median time-to-progression was 36.7 months. Patients with lower or higher serum VEGF values showed no statistically significant differences in survival. In contrast, high serum VEGF values were associated with shorter disease-free survival as compared with lower values (median DFS: 25 vs 60 months, $p = 0.048$ at log-rank test).

CONCLUSION: Our results suggest that serum VEGF could be of prognostic value in melanoma.

Management of prominent iris vascular tufts causing recurrent spontaneous hyphema

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Cornea. 2005 Mar;24(2):224-6.

PURPOSE: To report the management of recurrent, spontaneous hyphema associated with florid iris vascular tufts in a patient presenting for cataract surgery.

METHODS: Interventional case report and review of the literature; presentation of clinical findings, iris angiography, and the argon laser regimen used to minimize potential corneal complications with increased total treatment energy.

RESULTS: An 80-year-old man with a 20-year history of bilateral, recurrent, spontaneous hyphema associated with extensive iris vascular tufts presented with visually significant cataracts. Serial argon laser photocoagulation treatment of the prominent, circumferential iris vascular tufts of the left eye arrested further episodes of spontaneous hyphema and facilitated uneventful cataract surgery. Argon laser parameters were titrated to therapeutic effect during the initial treatment sessions, and sectoral photocoagulation of the circumferential vascular tufts was performed during a 5-month period to accommodate increased laser power and energy. The total energy required to complete treatment of the extensive lesions was substantially more than that in similar previous reports; however, no adverse corneal complications were associated with the laser therapy.

CONCLUSIONS: This case appears to represent the first description of chronic, bilateral, recurrent spontaneous hyphema associated with iris vascular tufts. Argon laser treatment of symptomatic iris vascular tufts promotes resolution of recurrent, spontaneous hyphema and may serve to mitigate the risk of hemorrhage from these lesions during subsequent intraocular surgery. Conservative management of increased total treatment energy may minimize the potential risk of corneal decompensation with argon laser therapy.

Malignant melanoma of the choroid associated with misdiagnosed ocular melanocytosis

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Acta Ophthalmol Scand. 2005 Feb;83(1):109-10.

No abstract available.

Retinal vascular occlusion with overlying vitreous hemorrhage masquerading as a tumor

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No abstract available.

Choroidal neovascular membrane associated with choroidal osteoma treated with trans-pupillary thermo therapy

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Indian J Ophthalmol. 2004 Dec;52(4):329-30.

Choroidal neovascular membrane, a known complication of choroidal osteoma causing visual loss when located subfoveally, can be successfully treated with transpupillary thermo therapy.

Treatment of metastatic tumors of the choroid with proton beam irradiation

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Ophthalmology. 2005 Feb;112(2):337-43.

OBJECTIVE: To describe the clinical outcomes of patients treated by proton beam irradiation for choroidal metastatic tumors. **DESIGN:** Noncomparative case series.

PARTICIPANTS: A retrospective chart review was performed on a series of 63 patients (76 eyes) with choroidal metastases treated with proton beam therapy between December 1989 and September 2000.

METHODS: Patients were treated with 2 fractions of 14 cobalt gray equivalents (CGEs) (CGE = proton Gy x relative biological effectiveness 1.1), each using a nonoperative "light-field" technique. Ophthalmologic follow-up was available for 46 patients (55 eyes), with a mean follow up time of 10 months. The medical record or the Social Security Death Index was used to obtain survival status, which was available in 94% of cases.

MAIN OUTCOME MEASURES: Tumor regression, recurrence, treatment-associated complications, and visual acuity were evaluated by ophthalmologic examination and ultrasonography. Eye retention and length of survival also were assessed.

RESULTS: At the time of ocular diagnosis, 49 patients reported a history of a primary cancer. Median survival time after ocular diagnosis was 16 months through May 2003. Most choroidal metastases were dome shaped (62%) and located at the posterior pole (95%). Mean tumor height was approximately 3.5 mm, and serous retinal detachment was seen in 63% of cases. Eighty-four percent of treated tumors regressed completely within 5 months of treatment, and none of these recurred. Retinal detachment resolved in 82% of patients within 3.8 months after treatment, and visual acuity was preserved or improved in 47% of the patients. Complications occurred in 56% of cases and included madarosis, keratitis, dry eye syndrome, cataract, neovascular glaucoma, chorioretinal atrophy, radiation papillopathy, and radiation maculopathy. None of the treated eyes required enucleation.

CONCLUSIONS: Proton beam irradiation is a useful therapeutic approach for choroidal metastases; it allows retention of the globe, achieves a high probability of local tumor control, and helps to avoid pain

and visual loss. Although complications occur in most cases, many of these are minor and are not associated with a change in function. This modality is accurate and efficient, because it only entails 2 treatment fractions and does not require surgery for tumor localization.

Descending atrophic tracts associated with choroidal hemangioma

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Retina. 2005 Feb-Mar;25(2):216-8.

No abstract available

Screening for uveal melanoma metastasis: Literature review

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Local tumour control in uveal melanoma has improved in the last decades. However, 5-year mortality due to metastases from large uveal melanomas remains high. Recently both isolated liver perfusion therapy and chemotherapy have reached encouraging results in improving metastasis survival. As such screening at an early stage, especially for liver metastases, becomes imperative.

Fluorescence angiography in the diagnosis of pigment-free melanomas of the choroid

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Vestn Oftalmol. 2004 Nov-Dec;120(6):8-11.

The ophthalmoscopic and angiographic patterns of 52 patients with pigment-free melanomas of the choroid, aged 27 to 73 were investigated. The mean age of examinees was 52. The prominence of tumors ranged from 1.3 to 6.5 mm. All patients underwent standard ophthalmoscopic general-clinical examinations. The tumor sizes were verified echographically. Fluorescence angiography was made according to the commonly used technique of serial pictures with computer processing. Diagnostically valuable angiographic signs were defined: contrasting of tumor vessels, spotty nature of fluorescence, beginning of fluorescence (arterial phase), pick of fluorescence (late venous phase), intensification velocity of fluorescence, late fluorescence, angiopathy of retinal vessels, large choroidal vessels and tumor-associated epitheliopathy of retina (small pigment foci, druses and hypofluorescence). Considering the research results, fluorescence angiography can be regarded as a principle diagnostic tool in pigment-free melanomas of the choroid.

Bilateral choroidal metastasis from carcinoma ex pleomorphic adenoma of the parotid gland.

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Clin Experiment Ophthalmol. 2005 Feb;33(1):70-2.

The histological, clinical and angiographic findings are reported of a 34-year-old man with bilateral visual loss who had left parotidectomy with subsequent radiotherapy due to carcinoma ex pleomorphic adenoma of the parotid gland 1 year before. Funduscopy disclosed choroidal masses with surrounding serous retinal detachment in both posterior poles. At the time of ocular diagnosis, lung, pleura and pharynx metastases had recently been revealed. Because of the extent of disease and its poor prognosis, no treatment was offered. Although parotid gland carcinoma usually spreads via lymphatics, choroidal involvement may rarely occur due to haematogenous dissemination.

Midperipheral mottling pigmentation with familial choroidal osteoma

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Retina. 2005 Jan;25(1):63-8.

PURPOSE: To describe a rare presentation of familial choroidal osteoma in two siblings.

METHODS: The clinical findings in two siblings over 4 years' follow-up.

RESULTS: Two brothers (15 and 12 years old) had bilateral choroidal osteomas. Both had bilateral peripapillary yellowish-white lesions and midperipheral mottling pigment appearance, which are not seen in sporadic cases. Extensive midperipheral area with mottling pigment appearance was noted by fluorescein angiography (FA) as scattered multiple hyperfluorescent dots. The yellowish-white lesions showed diffuse hyperfluorescence with FA and hypofluorescence with indocyanine green angiography (ICG). ICG also revealed irregular hyperfluorescent areas within the tumor, indicating abnormal choroidal vessels on the tumor. In the left eye of the younger brother, the subretinal fibrosis due to choroidal neovascularization superior to the macula extended down toward the foveal region over 2 years, resulting in visual deterioration.

CONCLUSION: The midperipheral mottling pigment appearance of familial choroidal osteoma cases is unique and different from most sporadic cases, suggesting that familial choroidal osteoma might have separate etiologic or modified factors.

Adenoma arising from nonpigmented ciliary epithelium concomitant with neovascularization of the optic disk and cystoid macular edema

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PURPOSE: To report a case of adenoma of the nonpigmented ciliary epithelium concomitant with marked neovascularization of the optic disk and cystoid macular edema (CME).

DESIGN: Observational case report. **METHODS:** A 34-year-old woman presented with a nonpigmented, vascularized tumor behind the iris and neovascularization of the optic disk (NVD). Visual acuity decreased gradually as a result of the development of CME.

RESULTS: Iridocyclectomy to excise the tumor was performed 8 months after initial examination. Histopathologic study revealed an adenoma arising from the NPCE. The levels of vascular endothelial growth factor were significantly elevated in both aqueous and vitreous humor obtained at surgery. The NVD and CME regressed after surgery, and the postoperative visual acuity improved to 20/40.

CONCLUSIONS: NVD and CME may develop secondary to adenoma of the nonpigmented ciliary epithelium. Increased levels of vascular endothelial growth factor in intraocular fluids may play a role in NVD and CME development.

Systemic non-hodgkin b-cell lymphoma encountered as a vanishing choroidal mass

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Arch Ophthalmol. 2005 Jan;123(1):105-9.

No abstract available.

Uveal and cutaneous melanoma: shared expression characteristics of melanoma associated antigens

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Invest Ophthalmol Vis Sci. 2005 Jan;46(1):24-30.

PURPOSE: Downregulation of melanoma-associated antigens (MAAs), against which natural cytolytic T lymphocytes (CTLs) exist in humans, is one of the mechanisms that aids in evasion of immune surveillance. In view of putative re-expression strategies for MAAs during immunotherapy, this study was conducted to investigate MAA silencing in malignant melanoma.

METHODS: The expression of the MAA Melan-A/MART-1 was analyzed in 10 uveal and 10 cutaneous patient-derived melanoma cell lines by Western blot analysis and RT-PCR. Expression characteristics of four other MAAs-Tyr, Tyrp1, Dct, and gp100/Pmel17-were analyzed by RT-PCR. DNA methylation patterns at the Melan-A/MART-1 promoter region were investigated by methylation-sensitive restriction enzyme digestion and subsequent Southern blot analysis. Exogenous promoter activity was assessed in all 20 melanoma cell lines to correlate the DNA methylation patterns with Melan-A/MART-1 expression.

RESULTS: MAA expression was observed in 15 of the 20 melanoma cell lines. Furthermore, there is a direct correlation between DNA methylation patterns at the Melan-A/MART-1 promoter region, exogenous Melan-A/MART-1 promoter activity, and Melan-A/MART-1 protein expression. These data reveal the division of patient-derived melanoma cell lines into two distinct subsets, which are identical for both uveal and cutaneous tumor types.

CONCLUSIONS: The authors propose a categorization of melanoma cell lines into two different panels based on shared MAA-expression characteristics: panel I, MAA-expressing cell lines, and panel II, MAA-deficient cell lines. This categorization can be used to obtain knowledge about the regulation of MAA-expression and for further research concerning MAA-based immunotherapy.

Ultrasound biomicroscopy in the management of melanocytoma of the ciliary body with extrascleral extension

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Br J Ophthalmol. 2005 Jan;89(1):14-6.

AIM: To demonstrate the ultrasound biomicroscopic features of a ciliary body melanocytoma with extrascleral extension, and a conservative approach in its management.

METHOD: Observational case reports. Two cases of ciliary body melanocytoma were suspected at presentation, confirmed histologically by biopsy, and subsequently monitored for change by serial ultrasound biomicroscopic imaging. The main outcome measures were anatomical and functional preservation of the eye, with avoidance of formal surgical excision. **RESULTS:** Ultrasound biomicroscopy allows clear visualisation of the tumours, and the ultrasound characteristic is of low homogeneous internal reflectivity. 5 year follow up with observation only demonstrates success with this conservative management approach. Histopathological evaluation confirmed melanocytoma.

CONCLUSIONS: Melanocytoma is a rare tumour. However if considered in the differential diagnosis at presentation and confirmed histologically, further management with use of the ultrasound biomicroscope as an accurate mode of imaging is an acceptable technique for preservation of the eye and avoids surgical excision.

Primary extracutaneous malignant melanoma: a comprehensive review with emphasis on treatment

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Onkologie. 2004 Oct;27(5):492-9.

Extracutaneous malignant melanomas (EMM) require special consideration in the field of oncology due to their rareness and--depending on the localization—the frequency of late diagnosis with consecutive poor prognosis. Only 4-5% of all primary melanomas do not arise from the skin. Most frequently they originate from the mucous membranes lining the respiratory, digestive, and genitourinary tracts or in the eyes as well as in the cerebral meninges. Extracutaneous melanomas are considered to be biologically more aggressive than cutaneous melanomas. The Clark level and Breslow index used for evaluation of cutaneous melanomas are not applicable to EMM and, at present, there are no consistent, internationally accepted therapy standards for this form of the disease. For this reason, this review focuses primarily on the literature pertaining to therapeutic strategies as well as epidemiologic, biological, and diagnostic aspects of this disease.