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Ciliary body malignant melanoma: A dilemma on staging

Melanoma maligno do corpo ciliar: um dilema no faseamento

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Abstract

Choroidal melanomas are the most common primary intraocular malignant tumor in adults. They tend to be more malignant; because of their location hidden behind the iris they can not be detected until they become larger. Therapeutic strategy is related by size, extension, number and location of tumor and growth patterns. High frequency ultrasound biomicroscopy (UBM) gives high resolution, cross-sectional images of the anterior segment lesions. Postequatorial lesions and intracranial extension of the melanomas are scanned by magnetic resonance imaging (MRI). We report a case of bilobed tumor with confusing appereance in preoperative imaging studies and macroscopy following enucleation. MRI is the perfect imaging method to reveal extension and size of the tumor in the posterior chamber. Combined use of UBM and MRI provides appropriate staging of ocular melanomas.

Keywords: *Melanoma/diagnosis; Ultrasound; Cliary body/pathology; Magnetic resonance imaging; Microscopy acoustic; Neoplasm staging.*

Resumo

Melanomas coroidais são os tumores malignos intra-oculares primários mais comuns em adultos. Eles tendem a ser mais malignos; devido à sua localização ser escondida por detrás da íris eles não podem ser detectados até se tornarem maiores. A estratégia terapêutica está relacionada com tamanho, extensão, número e localização dos padrões tumorais e de crescimento. O biomicroscopio ultra-sónico de alta frequência (BMU) fornece imagens transversais de alta resolução das lesões do segmento anterior. Lesões pós-equatoriais e de extensão intracraniana dos melanomas são digitalizadas em ressonância magnética (RM). Relatamos um caso de tumor com dois lóbulos, com aparência confusa em exames de imagem pré-operatórios e macroscopia após enucleação. A RM é o método de imagem perfeito para revelar a extensão e o tamanho do tumor na câmara posterior. O uso combinado de BMU e MRI fornece o faseamento apropriado dos melanomas oculares.

Descritores: Melanoma/diagnóstico; Corpo ciliar/patologia; Imagem por ressonância magnética; Microscopia acústica; Estadiamento de neoplasias.

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Introduction

Iris and ciliary body melanomas constitute about 15% of ocular melanoma cases. Ciliary body melanomas tend to be more malignant and because of their location hidden behind the iris they can not be detected until they become larger. Because of the agressive behaviour of the tumor, the therapeutic strategy is related by the size, extension, number and the location of the tumor and the growth patterns.[1] Therefore preoperative evaluation is very crucial. A case of bilobed ciliary body melanoma tumor causing confusion about the number and extension on preoperative imaging studies and enucleation material is reported.

Case report

Ninety-one years old age male patient applied to our clinic because of decreased vision of the right eye for two months. In the clinical history, he had many previously excised skin lesions at different times and he had been diagnosed as malignant melanoma, basal cell carcinoma and squamous cell carcinoma pathologically. In some of these excisions, the surgical margins were reported to be tumor positive, however he had not received any additional therapy.

Ophtalmologic examination revelaed that the visual acuity was at hand movements level in the right eye; 10/10 in the left eye. On biomicroscopic evaluation of the right eye a 3x3.2 mm in size, well defined, vascularized nevus was seen at the 6 o'clock radial location, protruded from the sclera and subconjunctivally, 1-2 mm distance to the limbus. Anterior segment was normal, pupil was regular and lens was centralized. Left eye was pseudophakic. On funduscopic examination of the right eye, vitreous was degenerated and liquefied, the tobacco dust sign was positive and retina was detached near totally; the left eye was normal. Ocular conventional sonography detected near total retinal detachment. Ultrasound biomicroscopy (UBM) revelaed a smooth, well defined, round solid mass arising from the ciliary body (Figure 1). Magnetic resonance imaging (MRI) of the both orbitas and brain was performed (Figure 2). At 5-7 o'clock location, crescentic lesion measuring 5x14x7 (apxtxcc) mm in size over the retina inferior to the lens was detected. The nodule was hyperintense on T1 weighted images; hypointense on T2 weighted images, with contrast enhancement. There was also retinal detachment. The lesion was thought to be a malignant melanoma of the ciliary body. There was not extraocular extension.

The patient was consulted by medical oncology and radiation oncology departments, no metastasis was detected. Enucleation

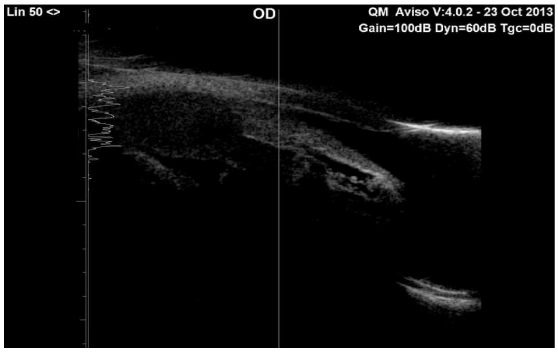

Figure 1: Ultrasound biomicroscopy shows hipoechogenic nodular, solitary lesion measuring 5mm at the ciliary body.

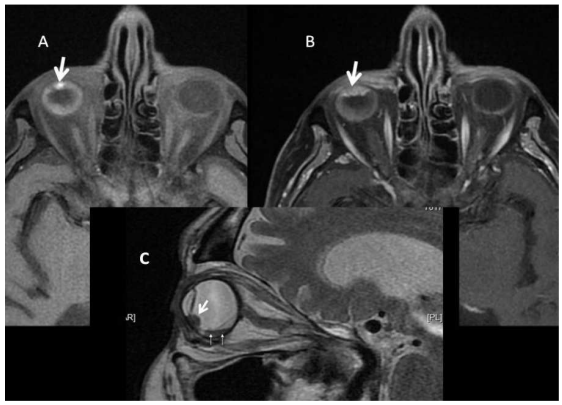

Figure 2: A) Precontrast fat saturated T1 W axial MRI shows a hyperintense micronodule at 6 o'clock. **B)** Postcontrast fat saturated axial scans reveal a lentiform lesion with homogenous and avid enhancement. **C)** T2 weighted sagital scan. The tumour at the ciliary body (arrow) extending posteriorly along the choroidea. There is not extraocular extension. Retinal detachment is also seen (thin arrows).

associated with porous sphere implantation was performed to enable mobile prosthetic application. Following enucleation, unexpectedly two distinct pigmented micronodular lesion was detected (Figure 3 A). The first lesion was at the 6 o'clock, a second lesion was observed behind the former. The second lesion thought to be missed on MRI but macroscopic examination revealed a black-brown colored 1.5x1x0.5 cm tumor starting from ciliary body and continuing all the way along choroidea at the posterior by forming bilobar nodular mass (Figure 3B). Each nodule was approximately 0.5cm in diameter and connected to each other by a thin rim of tumor tissue. Appropriate tissue samples from the tumor were prepared by routine tissue processor and 4µ thickness sections were stained by Hematoxylin-Eosine for routine light microscopic evaluation. The tumoral tissue consisted of short bundles of spindle cells with oval hyperchromatic nuclei and prominent eosinophilic nucleoli. Some of the tumor cells contained intracytoplasmic rough granular dark brown pigment. Spindle cell bundles were haphazardly intersecting each other and mitotic figures were evident. Tumoral lesion did not invade the sclera or optic nerve but performed complete retina elevation. On immunohistochemical evaluation, tumoral lesion showed positive cytoplasmic expression with HMB-45 and S-100. KI-

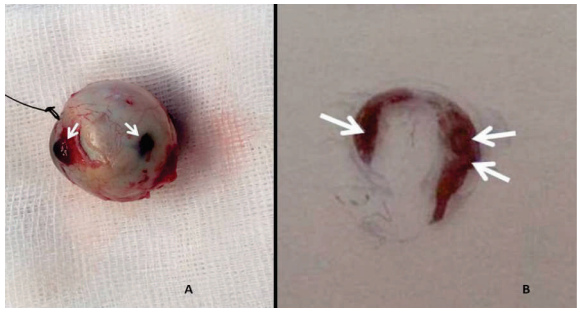

Figure 3: A) Following enucleation, two discrete hyperpigmented masses, one in preequatorial region anterior to the rectus muscle insertion and the other in the postequatorial location. **B)** The globe was dissected into two hemispheres between the cornea anteriorly and the optic nerve exit posteriorly. The tumor was starting from the ciliary body with nodular bulging appereance but simultaneously was extending posteriorly throughout the choroidea and forming a second nodular growing.

67 proliferation index was stated as 5%. According to light microscopic and immunohistochemical evaluation, the case was diagnosed as malignant melanoma- spindle B type (Figure 4).

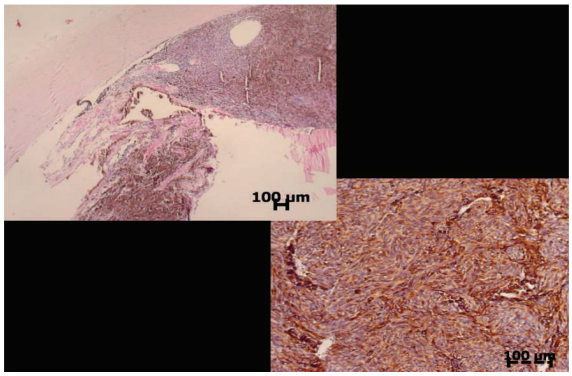

Figure 4: 5X, H.E: Tumoral lesion composed of pigment containing spindle cells starting from ciliary body and continuing all the way along choroidea. 20X, HMB-45 Tumoral cells showed diffuse strong positive cytoplasmic staining with HMB-45 immunohistochemistry.

Discussion

Preoperative imaging findings and enucleation material were in contradiction about the tumor's number and extension. One tumor at the ciliary body extending to posterior chamber along the choroidea was found in preoperative UBM and MRI imaging studies. Nevertheless enucleation specimen revealed two distinct lesions suggesting that MRI missed the second tumor. The patient was risky to develop multiple melanomas because of his previous skin melanomas history, therefore we first suggested multiple ocular melanomas or de novo lesion appeared in scanning-surgery interval. But histopathologic study concluded that there was one bilobed lesion. When the MRI scans were retrospectively investigated, the size and posterior extension of the lesion was compatible with pathologic size but superficial spread was not clearly visible.

Choroidal melanoma is classically a unilateral and unifocal tumor. Multifocal tumors are rare but they may occur in cases associated with immune deficiency, systemic malignancy, cutaneous melanoma, neurofibromatosis, familial atypical mole and melanoma syndrome, or Li–Fraumeni syndrome that might contribute to multifocality of cancer in general.[2] Our case was risky to have multiple choroidal melanomas because of his malignant melanoma history.

Choroidal melanomas appear as a mushroom- shaped, cresentic or flat tumor arising from choroid layer.[2,3] Without local invasion or extraocular extension, the size of the tumor is critical for therapeutic management.[4] Conventional ultrasonography, may visualize posterior chamber but the resolution is highly limited. UBM is a very valuable technique for high resolution in vivo evaluation of anterior segment tumours. UBM reaches a resolution up to 50 mm, with a 4 to 5 mm of tissue penetration which is similar to low power light microscopy; therefore the correlation with histological characteristics is good. Even small melanocytosis lesions are detectable by UBM.[5] Tumor borders, surface, internal echotexture and local extension can be delineated, therefore UBM is very helpfull in treatment planning.[1] The high resolution, surface imaging ability of UBM is limited for the lesions located on the anterior segment of the eye where the probe applications can be performed. Accordingly superficial lesions located behind the equator of the eye can not be analyzed by UBM. Postequatorial lesions and intracranial extension of the melanomas are scanned by MRI. Exudative retinal detachment can be associated with uveal melanomas. Differentiation of choroidal melanoma from retinal detachment can be difficult in some cases.

Magnetic resonance imaging reveal also extraocular extension of the tumor. Melanotic melanoma appears typically hyperintense on T1 weighted images, hypointense on T2-weighted images with contrast enhancement following gadolinium based contrast injection.(4) Retinal detachment have a pathognomonic V shape appereance with hyperintense on T1 and hypointense on T2-weighted images due to intracellular methemoglobin without contrast enhancement.

Melanotic melanoma is usually associated with retinal detachment and noncontrast images can not distinguish the tumor from the detachment.(4) Because of high cellularity of the melanomas, they can be differentiated from retinal detachment on diffusion weighted images (DWI).(6) We had DWI sequence for brain, with thick slices (1 cm), because of thick slice and small size of the tumors, none of the tumors were visible on DWI.

In conclusion, MRI is the perfect imaging method to reveal extension and size of the tumor in the posterior chamber. Combined use of UBM and MRI provides appropriate staging of ocular melanomas.

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