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MIXED CELL MALIGNANT MELANOMA OF THE CILIARY BODY

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INDEXING WORDS

malignant melanoma; ciliary body; diagnostic problems; management

SYNOPSIS

Malignant melanomas of the ciliary body are uncommon tumors and the ratio of occurrence in the ciliary body to that in the choroid is 1:10[27]. These tumors may have a nodular or diffuse growth pattern or combination of the two[31].

The authors recently encountered a malignant melanoma of the ciliary body. After evaluation of the results of clinical examination, computed tomography(CT) and magnetic resonance imaging(MRI) a tentative diagnosis of malignant melanoma of the ciliary body in the left eye was made and an enucleation was done. Histopathological examination revealed the case as a mixed cell malignant melanoma of the ciliary body. The patient was followed up for approximately one year with no signs of metastasis or recurrence.

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Infrequently problems are related to the clinical diagnosis of small malignant melanomas of the ciliary body. Therefore, we recommend that orbital CT and MRI should be done in conjunction with other usual examinations in both the suspected and unsuspected melanomas of the ciliary body.

INTRODUCTION

Both benign and malignant melanomas are encountered less frequently in the ciliary body than in the choroid. Reese[27] reviewed 271 cases of malignant melanoma of the uveal tract, only 15 of the neoplasms were found in the ciliary body. It has been noted that uveal malignant melanomas are rare in blacks but common in whites[23,35] and, a black and white ratio was found to be 1:98[19]. The Asians have an incidence in between blacks and whites[23].

It is generally accepted that the malignant melanoma of the ciliary body possesses a great affinity for metastasis and the tumor can be life-threatening; therefore, early diagnosis and treament is very important. The treatment of malignant choroidal melanomas with photocoagulation has been reported by Vogel[32] while the role of enucleation in the management of uveal melanomas has been elaborately described by others[33,34]. Low mortality rate among untreated uveal melanomas has also been recorded[1,6,13,14,16]. Some investigators stated the various prognostic factors in the management of uveal melanomas[10,11,12,20,21,22,29]. We would like to report a case of malignant melanoma of the ciliary body describing its clinical and prognostic features.

CASE REPORT

[redacted] aged 79 first consulted us at the department of Ophthalmology, Kobe University School of Medicine on October 17, 1994 for the complaints of foreign body sensation in her left eye since September 30, 1994. The patient denied any history of infection, trauma or foreign body in the left eye. She did not give any history of drug abuse or allergies but she was hypertensive.

On general examination her oral temperature was found to be 37° C; pulse 70/min. and respiration 18/min. Her blood pressure was within normal limits at the time of examination. Ophthalmic examination showed visual acuity 0·5 in the right eye and 0·2 in the left eye. The lens opacities accounted for the reduction of visual acuity in both eyes with and without correction. Pupils were circular and reactive to light and accommodation. The intraocular tension was 8 mmHg in the right eye and 12 mmHg in the left eye and did not rise with mydriatics. Extraocular muscles were full and orthophoric. Eyelids and eyelashes were found to be normal. No abnormality was seen in the conjunctiva, cornea and sclera of either eye. Slit lamp examination revealed iris pigments on the inner surface of the cornea and on the anterior surface of the lens of the left eye. The optic discs and fundi were normal.

Results of the following laboratory tests were within normal limits: a complete thyroid profile, plain roentgenograms of the skull and orbita but liver function tests showed liver dysfunction and blood cell count showed a decreased number of white cells. B-mode ultrasonography was done and no abnormality was seen in either eye. Orbital tomograms and MRI(Fig.1A) revealed a mass around the 12 O'Clock position in the left eye. There was no evidence of extraocular extension. A tentative diagnosis of malignant melanoma of the ciliary body was entertained and enucleation advised. The patient underwent complete metastatic evaluation including Gallium test. Findings from these were normal. Thus she was admitted to Kobe University Hospital in October 24, 1994. Under retrobulber anesthesia, an enucleation was performed and the eye underwent a histopathological examination.

Pathological findings

Gross Pathology

The tumor measured approximately 4x10x4mm and was located in the ciliary body and protruded into the vitreous(Fig.1B). The entire mass was examined and it was found to be a well-circumscribed pigmented tumor. There was neither apparent infiltration to the sclera nor melanosis in any other part of the eye.

Microscopic findings

Multiple sections were taken throughout the extent of the tumor and the surrounding tissue and hematoxylin-eosin and reticulin silver impregnation stain slides were prepared. The retina was found to be detached. The tumor was composed of pleomorphic cells with abundant eosinophilic cytoplasm and it was moderately pigmented. Nuclei were large, round to oval in shape with prominent nucleoli(Fig.1C), showing frequent mitotic figures. Like epithelial neoplasms, pleomorphic tumor cells arranged cohesively with scarce intercellular reticulin. In some areas, tumor cells were spindle shaped and proliferated in a fascicular pattern(Fig.1D). Many melanophages aggregated surrounding the tumor and infiltrated to the adjacent sclera and iris. These melanophages were also observed in the region of the anterior chamber. No invasion was seen to the optic nerve. On the basis of these pathological findings we diagnosed the case as a malignant melanoma of the ciliary body.

DISCUSSION

Malignant melanomas of the ciliary body may be asymptomatic or a variety of clinical features can be produced by them depending on their growth patterns and effects on the adjacent ocular structures[31]. These tumors usually assume a nodular or diffuse growth pattern. The diffuse type (ring melanoma) is more aggressive and generally carries a worse prognosis. Melanomas in the ciliary body may cause cataract, subluxation of the lens and vitreous hemorrhage. These tumors can grow forward into the anterior chamber and produce secondary glaucoma or hyphema[31]. Usually these features are not observed in the choroidal melanoma unless the melanoma is large or localized in the peripheral region.

There is little agreement concerning the management of ciliary body or choroidal melanoma. Cryosurgery[17], photocoagulation[24,32], chemotherapy[27], cobalt plaque implantation[2,5,18] and eye wall resection[26] have been advocated as alternatives to enucleation. In a recent study[36] proton-beam irradiation of the tumor has been described, but the efficacy of such radiation therapy is still unclear. Vogel[32] reported a 10 years follow-up study on 54 patients of malignant

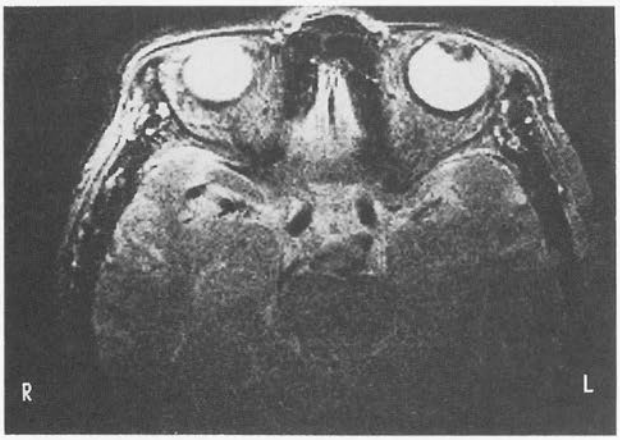

Fig. 1A.

Orbital MRI showing a mass around the 12 O'Clock position in the left eye.

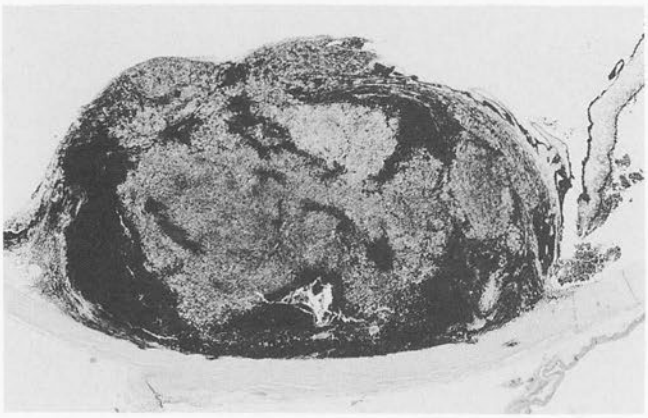

Fig. 1B.

Section of the eye showing tumor of the ciliary body(HE stain. x100).

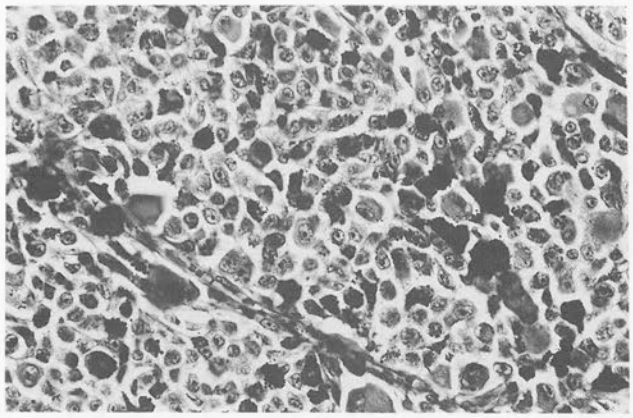

Fig. 1C.

The tumor composed of epitheloid cells with round to oval nuclei(HE stain. x700).

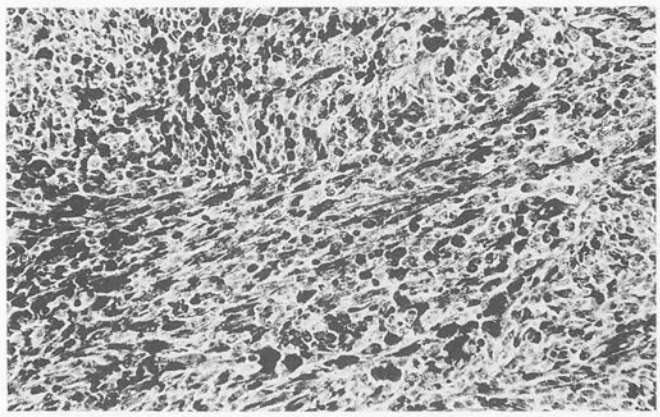

Fig. 1D.

The photomicrograph showing spindle shaped cells with fascicular proliferations(HE stain. x350).

melanoma of the choroid treated by photocoagulation and he was in favor of photocoagulation for small melanomas. Enucleation is the most widely used surgical technique for treating uveal melanomas. But there are different opinions about the effectiveness of enucleation as a modality of treatment. Zimmerman and coworkers[33,34] described a higher mortality rate following enucleation for uveal melanomas. This higher mortality may be attributed to the dissemination of tumor cells at the time of surgery. So, they suggested to develop a new technique for enucleation to prevent the dissemination of tumor cells.

Malignant melanomas of the ciliary body have been notoriously unpredictable in respect of metastasis. It is generally believed that melanoma of the choroid disseminates earlier than do similar tumors located more forward in the uveal tract. In a review of 432 ciliary body and choroidal melanomas, some investigators described the evidence of orbital extension in 45 cases[30]. The incidence of orbital extension was influenced primarily by the cell type and the size of the neoplasm. In 23% of these cases, they found local orbital recurrence with evidence of extrascleral extension. They also described that exenteration had better prognosis than patients treated by chemotherapy, radiotherapy or local excision.

In general, ciliary body and choroidal melanomas carry the same prognosis[15] and the prognosis of these tumors correlate closely with their pigmentation[4,25], cell type[3,8], size of the tumors[7,9] and extension of the tumors[15]. It has been recorded that the largest tumor diameter is the most important clinical and pathological prognostic factor[29]. In another study, McLean and co-workers[20] found that seven factors are closely related to the prognosis of malignant melanomas of the choroid and ciliary body. These are pigmentation, size, cell type, scleral extension, mitotic figure, location of anterior margin of the tumor and optic nerve involvememt. In our case, the tumor was small in size and composed of mixed cells, and the pigmentation was of a moderate degree. There was no scleral or extraocular extension. So we conclude that our patient carries a moderate prognosis.

In some cases, diagnostic problems are related to the small melanomas of the ciliary body; so we advocate that orbital CT and MRI must be done in addition to other routine examinations in suspected and unsuspected melanomas of the ciliary body.

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