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Research Article

Uveal Malignant Melanoma: A Case Report

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Abstract

Uveal malignant melanoma arises from melanocytes in the uveal tract of the eye which comprises of iris, ciliary body and choroid (common). It is the most common intraocular malignancy seen in Caucasian adults and is more in males than females. Early diagnosis and local treatment is crucial as survival correlates with primary tumor stage. Approximately 50% of the patients develop metastasis even after 20 years of diagnosis and may die in 6 to 12 months after the confirmation of metastatic disease, although the recurrence is infrequent. We report a case of malignant melanoma of uvea in 37 years old female.

Keywords: Uveal Malignant Melanoma; Choroid Melanoma; Enucleation; Melanocytes

Introduction

Malignant melanoma is a rare malignancy arising from the melanocytes located at various anatomical locations including skin (91%), mucous membrane (1%), ocular region (5%), and rarely from some unknown primary sites (2%) [1]. Uveal melanoma arises from melanocytes in the uveal tract of the eye which consists of iris, ciliary body and choroid. Choroid is the commonly involved part (85-90%) followed by ciliary body (5-8%) and iris (3-5%) [2,3]. It is the most common intraocular malignancy seen in Caucasian adults and is more in males than females. The overall mean age adjusted incidence of uveal melanoma is 5.1 cases in US, 1.3-8.6 cases in Europe, 0.2-0.3 cases in Africa and Asia per million per year [4,5]. Uveal melanoma is more commonly seen in older age groups with median age of diagnosis as 62 years and range of 65-79 years [2,6] The predisposing factors include fair skin, light eye color, blonde hair and inability to tan which maybe related to less melanin present in these individuals, resulting in less protection from ultraviolet light and sunlight exposure [7]. Uveal melanoma is treated either by enucleation or radiotherapy. Early diagnosis and local treatment is crucial as survival correlates with the primary tumor stage. Approximately 50% of the patients develop metastasis even after 20 years of diagnosis and may die in 6 to 12 months after the confirmation of metastatic disease, although the recurrence is infrequent [8]. We report a case of malignant melanoma of uvea in 37 years old female.

Case Presentation

A 37 years old female reported to our department with the chief complaints of gradual loss of vision on the right side of eye

and headache on the same side from last 2 months. Loss of vision was gradual in onset. On ocular examination loss of vision was seen in the right eye with dark grey mass in the posterior segment andretinal detachment. Left eye was normal with normal vision.B scan Ultrasonography of Right eye showed a well defined hypo-echoic focal lesion of size 12 * 16 mm in posterior segment with feeding vessels seen at lateral orbital wall and retinal detachment at temporal half as shown in figure 1. MRI of brain was done to further evaluate the extent and exact location of the lesion. MRI showed a well defined lobulated intraocular T1 hyperintense and T2 hypointense lesion along the lateral aspect of right eye. The lesion orginated from the choroid with size of 1.12*1.28*1.80 cm (AP*TRA*CC), is bilobed with broad base (1.6 cm) extending into the iris with mild enhancement on post contrast. Retinal detachment was present with no extra scleral extension. Rest of right orbit and left orbit appear unremarkable and is shown in figure 2. The clinical and imaging features were suggestive of choroidal or uveal melanoma. PET CT Scan was performed and it showed no regional or distant metastasis. The patient was treated with enucleation of right eye. The Histopathological report was confirmative of uveal malignant melanoma with TNM stage PT3bNxMx as shown in figure 3. The microscopic photographs of various resolutions is shown in figure 4.

Figure 1: B scan Ultrasonography of Right eye showing a well defined hypo-echoic focal lesion of size 12 * 16 mm in posterior segment with feeding vessels seen at lateral orbital wall and retinal detachment at temporal half.

Figure 2: MRI showing a well defined lobulated intraocular T1 hyperintense and T2 hypointense lesion along the lateral aspect of right eye. The lesion originated from the choroid and is extending into the iris with mild enhancement on post contrast. Retinal detachment is present with no extra scleral extension. Rest of right orbit and left orbit appear unremarkable.

Figure 3: Final histopathological report.

Figure 4: (a) Photograph of histopathological section (H&E, 40X) Sclera on top, part of cilliary body at left lower and tumor island in right lower corner. (b): (H&E, 10x: Tumor cells with spindled morphology arranged in fascicles with abundant melanin pigment deposition. (c): (H&E, 40X): Tumor cells with epithelioid morphology with prominent nucleolus arranged in sheet, mitosis in the center and intracytoplasmic melanin pigment.

Discussion

Uveal malignant melanoma is a highly malignant tumor of epithelial origin. It usually presents with headache, blurred vision, pain, photopsia, floaters, loss of vision, visible tumor or is asymptomatic [9]. Choroidal melanoma presents as a dome shaped or mushroom shaped mass because of the rupture of bruch's membrane. The lesion can be pigmented (55%) or non-pigmented (15%) or has a mixed color (30%) [10]. Retinal detachment is seen in almost 70% of cases. The AJCC classifies choroidal melanoma based on the tumor thickness, the tumor base dimension, ciliary body involvement and associated extraocular extension [11]. The diagnosis of uveal melanoma is primarily based on clinical examination by biomicroscopy and indirect ophthalmoscopy. Other tests include ultrasonography (USG), indocyanine green

angiography (ICGA), fundus fluorescein angiography (FFA), fundus autofluorescence (FAF), color fundus photography, optical coherence tomography (OCT), and ultrasoundbiomicroscopy (UBM) can be done in order to confirm diagnosis [12]. In contrast to the basic principles of oncology, histological or cytologic evaluation is not routinely used in the diagnosis of intraocular neoplastic lesions. The most common differential diagnoses include choroidal nevus, congenital hypertrophy of the retinal pigment epithelium, peripheral exudative hemorrhagic chorioretinopathy, circumscribed choroidal hemangioma, hemorrhagic detachment of the retina or pigment epithelium, and age-related macular degeneration [13]. Histo-pathologically choroidal melanoma is classified into four types, based on the presence of spindle or epitheloid cells (modified callender's classification) [14].

- Pure Spindle cell type - 45%.
- Pure epitheloid cell Melanomas - 5% .
- Mixed cell melanoma - 45% (both spindle cell and epitheloid cell types).
- Necrotic melanoma - 5% (presence of unrecognizable type of cell).

The present case is type 3. Immunohistochemically, malignant melanomas are reactive for HMB-45, S-100 proteinand Mart-1 (Melan-A) [15].

Treatment options for uveal melanoma include conservative Eye surgery like photocoagulation and transpupillary thermotherapy for small lesions of less than 4 mm thickness, radiotherapy for small to medium lesions of <18mm in diameter and < 12 mm in thickness, enucleation for large posterior uveal melanomas of > 18mm in diameter and >12 mm in thickness and orbital exenteration for extraocular extension. There are two main types of radiation therapy which include plaque brachytherapy (iodine-125, ruthenium-106, or palladium-103, or cobalt-60) and teletherapy (proton beam, helium ion, or stereotactic radiosurgery using cyber knife, gamma knife, or linear accelerator) [16]. Randomized, multicenter clinical trials conducted by the Collaborative Ocular Melanoma Study (COMS) group showed no difference in long-term survival rates of patients treated with plaque radiotherapy or enucleation in medium-sized tumors (basal diameter of 16 mm and apical height 2.5-10 mm) [17].

Local tumor control of uveal melanoma is excellent, but this malignancy is associated with relatively high mortality secondary to distant metastasis. Various clinical, histopathological, cytogenetic and gene expression features help in estimating the prognosis of uveal melanoma. The clinical features associated with poor prognosis in patients with uveal melanoma include older age at presentation, male gender, ciliary body location, diffuse tumor configuration, larger tumor basal diameter and thickness, association with ocular/oculodermal melanocytosis, extraocular tumor extension, and advanced tumor staging [18]. Histopathological features suggestive of poor prognosis include epithelioid cell variant, high mitotic activity, higher microvascular density, extravascular matrix patterns, tumor-infiltrating macrophages, tumor-infiltrating lymphocytes, higher expression of human leukocyte antigen Class I and II and higher expression of insulin-like growth factor-1 receptor [18]. Monosomy 3, 1p loss, 6q and 8q loss, and those classified as Class II by gene expression are predictive of poor prognosis of uveal melanoma [18].

Uveal malignant melanoma has a high tendency for distant metastasis which is the reason for its higher mortality [19]. The common sites of metastasis include liver (89%), lung (29%), and bone (17%) [20]. Approximately, 50% of patients with uveal melanoma succumb to metastasis within 10 years of diagnosis, irrespective of the type of treatment. Median survival after metastasis is 6 to 12 months and is better in patients receiving any form of treatment than those receiving no treatment [20,21].

Conclusion

Uveal melanoma is an aggressive tumor with favourable prognosis in anterior than posterior lesions. Patients should be followed for long-term due to presence of distant metastasis even after twenty years of treatment. Early detection, proper treatment and long term surveillance are key factors for long term local and distant control.

Acknowledgement

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Conflict of Interest

None declared.

Bibliography

1. Chang AE., *et al*. "The National Cancer Data Base report on cutaneous and noncutaneous melanoma: a summary of 84,836 cases from the past decade". The American College of Surgeons Commission on Cancer and the American Cancer Society". *Cancer* 83.8 (1998): 1664-1678.

2. McLaughlin CC., *et al*. "Incidence of noncutaneous melanomas in the U.S". *Cancer* 103.5 (2005): 1000-1007.

3. Damato B. "Progress in the management of patients with uveal melanoma: the 2012 Ashton Lecture". *Eye (Lond)* 26.9 (2012): 1157-1172.

4. Virgili G., *et al*. "Incidence of uveal melanoma in Europe". *Ophthalmology* 114 (2007): 2309-2315.

5. Kivelä T. "The epidemiological challenge of the most frequent eye cancer: retinoblastoma, an issue of birth and death". *British Journal of Ophthalmology* 93.9 (2009): 1129-1131.

6. Andreoli MT., *et al*. "Epidemiological trends in uveal melanoma". *British Journal of Ophthalmology* 99.11 (2015): 1550-1553.

7. Weis E., *et al*. "The association between host susceptibility factors and uveal melanoma: a meta-analysis". *Archives of Ophthalmology* 124.1 (2006): 54-60.

8. Krantz BA., *et al*. "Uveal melanoma: epidemiology, etiology, and treatment of primary disease". *Clinical Ophthalmology* 11 (2017): 279-289.

9. Damato EM and Damato BE. "Detection and time to treatment of uveal melanoma in the United Kingdom: an evaluation of 2,384 patients". *Ophthalmology* 119.8 (2012): 1582-1589.

10. Shields CL., *et al*. "Metastasis of uveal melanoma millimeter-by-millimeter in 8033 consecutive eyes". *Archives of Ophthalmology* 127.8 (2009): 989-998.

11. Edge SB., *et al*. "Malignant melanoma of the uvea". In: AJCC Cancer Staging Manual, 7th edn. Springer: New York, NY, USA (2010): 547-559.

12. Tarlan B and Kıratlı H. "Uveal Melanoma: Current Trends in Diagnosis and Management". *Turkish Journal of Ophthalmology* 46.3 (2016): 123-137.

13. Shields JA., *et al*. "Pseudomelanomas of the posterior uveal tract: the 2006 Taylor R. Smith Lecture". *Retina* 25.6 (2005): 767-771.

14. IW McLean., *et al*. "Reappraisal of Callender's spindle A type of malignant melanoma of choroid and ciliary body". *American Journal of Ophthalmology* 86 (1978): 557-564.

15. MJ Hendrix., *et al*. "Biologic determinants of uveal melanoma metastatic phenotype: role of intermediate filaments as predictive markers". *Lab Investigation* 78 (1998): 153-163.

16. S Kaliki and CL Shields. "Uveal melanoma: relatively rare but deadly cancer". *Eye* 31 (2017): 241-257.

17. Collaborative Ocular Melanoma Study Group. "The COMS randomized trial of iodine 125 brachytherapy for choroidalmelanoma: V. Twelve-year mortality rates and prognosticfactors: COMS report No. 28". *Archives of Ophthalmology* 124.12 (2006): 1684-1693.

18. Kaliki S., *et al*. "Uveal melanoma: estimating prognosis". *Indian Journal of Ophthalmology* 63.2 (2015): 93-102.

19. Singh AD., *et al*. "Uveal melanoma: Trends in incidence, treatment, and survival". *Ophthalmology* 118 (2011): 1881-1885.

20. Diener-West M., *et al*. "Development of metastatic disease after enrollment in the COMS trials for treatment of choroidalmelanoma: Collaborative Ocular Melanoma Study Group Report No 26". *Archives of Ophthalmology* 123 (2005): 1639-1643.

21. Gragoudas ES., *et al*. "Survival of patients with metastases from uvealmelanoma". *Ophthalmology* 98 (1991): 383-389.