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A case of ciliary body mesectodermal leiomyoma with rapid growth and loss of vision necessitating enucleation

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ABSTRACT

Introduction: Mesectodermal leiomyoma of the ciliary body is a benign rare tumor that rarely presents acutely with a complicated clinical course.

Presentation of case: We are reporting a 39-year-old healthy female who presented with subacute blurred vision in her right eye secondary to a ciliary body mass. Initial fine needle aspiration biopsy ruled out a melanoma but the patient meanwhile experienced rapid complicated growth of the mass with vision loss, for which her right globe was eventually enucleated. The tumor was diagnosed histopathologically to be mesectodermal leiomyoma.

Discussion: The clinical course of our case was unique because of the subacute onset of her symptoms, the rapid growth of her benign tumor, and the complicated tumor behaviour leading to enucleation. An acute presentation of mesectodermal leiomyoma has been reported only once among similar cases in the recent literature. The tumor in our case showed the characteristic histopathological and immunohistochemical findings described before.

Conclusion: Ophthalmologists should be aware of the unusual acute/subacute presentation of a rather benign lesion. Delayed diagnosis may result in ophthalmic complications and loss of the globe.

1. Introduction

Mesectodermal leiomyoma is the proper term for a benign smooth muscle tumor arising in the ciliary body (CB). This term was first coined in the year 1977 by Jacobiec et al. owing to the embryologic origin of the CB muscles from neural crest cells (mesectoderm) [1]. Since then, this origin has been confirmed in several reported cases through expression of smooth muscle and neural immunohistochemical (IHC) markers by the tumor cells, as well as electron microscopic demonstration of characteristic thin filaments and focal densities within the cytoplasm of the cells [2]. Benign leiomyoma is expected to exhibit slow growth, often with complications but acute presentation is unusual and has been first reported recently by Kim et al. [3] This case report has been prepared in compliance with the updated SCARE 2018 criteria [4]. A general written informed consent was taken from the patient, which includes permission for anonymous use of information and photos for the purpose of publication.

2. Case presentation

A 39-year old healthy female presented with subacute blurred vision in her right eye over the past month. Her family history, genetic background, and past medical history were irrelevant, and she was not on any medications. The right eye examination revealed visual acuity of counting fingers and intraocular pressure of 18 mmHg. External examination of that eye showed dilated scleral sentinel vessels temporally with adjacent conjunctiva, clear cornea, and normal lens (Fig. 1a). A large pigmented mass originating from the ciliary body was seen following dilatation of her right pupil (Fig. 1b). The mass extended posteriorly near the equator with a basal diameter of 18 mm and height of 13mm. No surface orange deposits nor drusen were seen, however, extensive subtotal exudative retinal detachment was noted. The ultrasound biomicroscopy scan showed an iso-dense echoic lesion, which fully transmitted light during light transillumination test. Fine needle aspiration biopsy from the tumor by the ocular oncologist ruled out CB melanoma, which was highly suspected in her case based on clinical presentation by lack of expression of HMB45 and Melan-A. The differential diagnosis included glomus tumor, and angioleiomyoma but no definitive diagnosis was reached. The tumor continued to grow rapidly over the ensuing two weeks with further deterioration of her right eye vision to light perception. The ultimate decision by her treating

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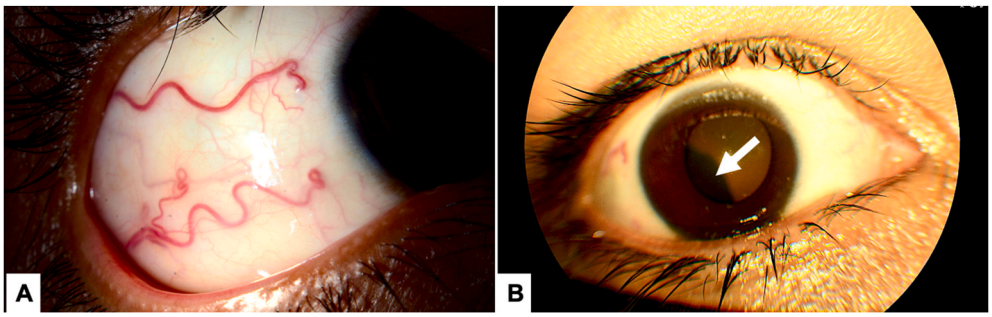

Fig. 1. 1A: The clinical appearance of the dilated scleral sentinel vessels in the temporal conjunctiva of the patient's right eye at the initial presentation.**1B:** The ciliary body pigmented mass seen through a dilated right pupil (White arrow).

oncologist was to perform enucleation of her right non-seeing globe, which was accepted by the patient and an informed consent was obtained from her for the procedure. The procedure was successfully conducted and was well tolerated by the patient. The enucleated globe showed a large CB mass extending to the adjacent choroid posteriorly with subtotal retinal detachment. Histopathologically, a well-defined CB tumor mass was observed to be arising from adjacent normal smooth muscle (Fig. 2a). The tumor consisted of polygonal and spindle-shaped cells with eosinophilic cytoplasm, round to oval nuclei, and inconspicuous nucleoli. The cells showed fibrillar appearance and intervening dilated vessels (Fig. 2b). Mitotic figures were rare (1 per 10 high-power fields), and the proliferative index using Ki67 was 1%. The tumor cells expressed smooth muscle IHC markers: Desmin, smooth muscle actin (SMA), muscle specific actin (MSA), h-caldesmon, and Calponin in addition to progesterone receptors (Fig. 2c–f). Focal weak reactivity was observed with CD56 and neurofilament protein. The tumor cells remained unreactive to the melanoma markers mentioned above as well as to epithelial and neuroendocrine markers. The cells were also not reactive to estrogen receptors marker. Based on the morphology of the tumor, the cellular appearance, and the IHC characteristics, the diagnosis of mesectodermal leiomyoma of the CB was made. The patient management plan included a custom-made prosthesis for the right socket, and close regular follow up in the same eye center where the enucleation, diagnosis and further management was performed. She has been followed up now every six months for 3 years with clean socket upon external examination by the ophthalmic surgeon, good cosmetic outcome, and no recurrence.

3. Discussion

Mesectodermal leiomyoma of the CB is rare and has been shown to arise from multipotential neuroectodermal cells resulting in a combined neurogenic and myogenic morphology with occasional negative staining to neural markers. However, positive staining with CD56, which is identified in the nervous system as neural cell adhesion molecule has been reported and was also observed in our case [5]. Quhill et al. have interestingly reported the expression of the tumor cells in 3 cases of CB mesectodermal leiomyoma to 2 sex steroid hormone receptors: progesterone and androgen receptors (but not to estrogen receptors) and suggested their role in the pathogenesis of such a tumor [6]. The tumor in our case similarly demonstrated reactivity to progesterone -but not estrogen- receptors.

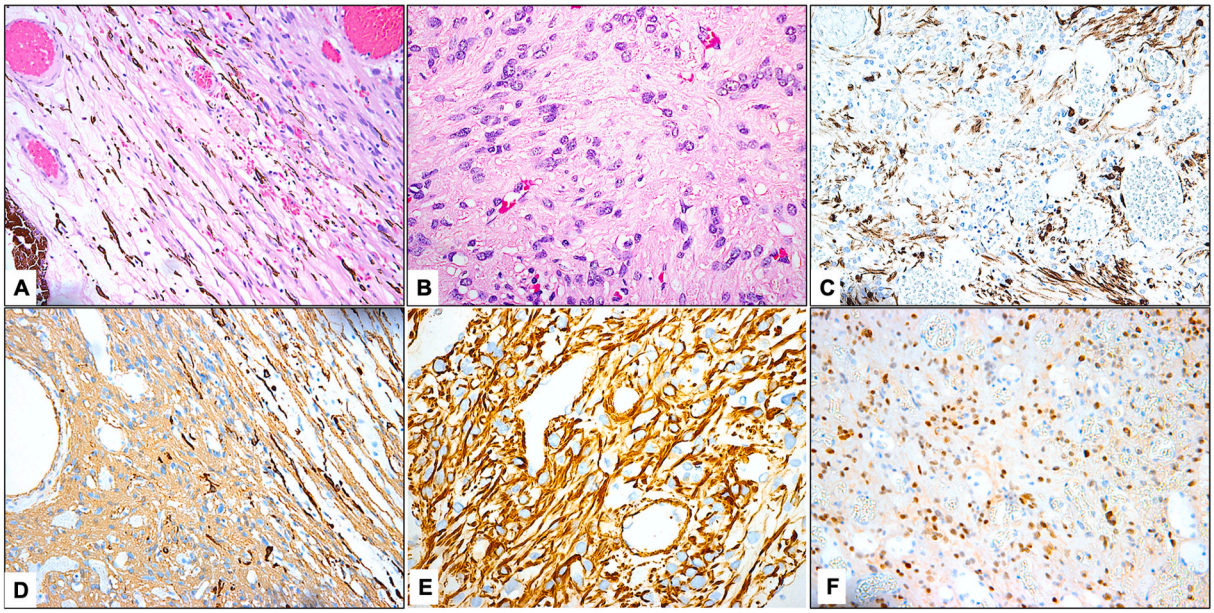

Fig. 2. 2A: The histopathological appearance of the ciliary body (CB) tumor following enucleation, which appears to be arising from normal CB smooth muscle (Original magnification x200 Hematoxylin and eosin). **2B:** The higher power appearance of the tumor cells with fibrillary appearance (Original magnification x400 Hematoxylin and eosin). **2C:** The tumor cells expressing one of the immunohistochemical muscle markers used to confirm the diagnosis in this case (Original magnification x400 Desmin smooth muscle actin). **2D:** The tumor cells originating from CB smooth muscles, both of which are expressing smooth muscle marker (Original magnification x200 smooth muscle actin). **2E:** Another confirmation of the expression of tumor cells to muscle markers (Original magnification x400 h-caldesmon). **2F:** The CB tumor cells staining with sex hormone marker showing positive expression (Original magnification x400 Progesterone).

Clinically, it is difficult to differentiate CB leiomyoma from uveal melanoma, and many reported cases stress on the necessity for tissue diagnosis confirmation by histopathology and IHC staining. However, authors who have attempted to summarize previously reported cases of mesectodermal leiomyoma have concluded few hints that might aid in the clinical diagnosis, such as predilection of the tumor for females in their reproductive age and light transillumination (as in our patient) [3,5]. Even though this tumor is known to be slow growing, it can reach a large size resulting in retinal detachment, lens displacement, and glaucoma with loss of vision thus ending by enucleation similar to what happened in our case [3]. However, local complete resection of smaller tumors has been preferred over observation along with long-term follow up to detect any recurrence [2,3]. We do support local surgical excision of such lesions if presenting early to avoid complications and to salvage the globe, which unfortunately was not possible in our case because of a relatively late presentation.

4. Conclusion

We describe the rare occurrence of a rapidly growing large CB mesectodermal leiomyoma resulting in loss of vision that necessitated enucleation. The diagnosis based on the fine needle aspiration biopsy alone was challenging. This might not have affected the outcome in our case because of the late presentation of the patient and the further complicated rapid growth of the tumor. However, early detection and recognition of this rare tumor would allow globe salvage. The fact that the tumor in our case expressed reactivity to sex steroid hormone by IHC staining similar to few other reported cases is interesting. Further studies to investigate the response of this tumor to hormonal therapy might be also warranted.

Ethical approval

Case reports do not require Ethical approval in our institution.

This case report was prepared in accordance with the ethical standards and the Helsinki Declaration. No trial of new drugs or therapy is applicable in this case.

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Author contribution

First author: Literature review, Histopathological images, Manuscript writing/editing for submission and the corresponding author.

Second author: Clinical diagnosis of the case and manuscript review.

Third author: Histopathological examination and final tissue diagnosis.

Registration of research studies

1. Name of the registry: This is a case report, which does not require a registry.

2. Unique Identifying number or registration ID:

3. Hyperlink to your specific registration (must be publicly accessible and will be checked):

Guarantor

Dr. Hind Manaa Alkatan.

Consent

A written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Provenance and peer review

Not commissioned, externally peer reviewed.

Declaration of competing interest

This case report was prepared in accordance with the ethical standards and the Helsinki Declaration. No trial of new drugs or therapy is applicable in this case. Case reports do not require Ethical approval in our institution. A general written informed consent was taken from the patient, which includes permission for anonymous use of information and photos for reporting. The authors have no conflict of interest to declare in relation to this case report. This manuscript was not submitted to any other journal.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2020.11.067.

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