Inflammatory Myofibroblastoma Tumor of Intraocular Ciliary Body: A Case Report and Literature Review

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Case Report

Keywords: Inflammatory myofibroblastoma tumor , ciliary body, immunohistochemistry

Posted Date: October 9th, 2020

DOI: https://doi.org/10.21203/rs.3.rs-86928/v1

Abstract

Background: IMTs are extremely rare in eyes. This is the first report of a IMT of the ciliary body.

Case presentation: A ciliary body tumor was found under slit lamp biomicroscopy in a 55-year-old male first diagnosed with cataract. Then this patient underwent trans-sclera resection via partial lamellar sclerouvectomy and par plans vitrectomy to remove the mass. Hematoxylin and eosin (HE) staining and immunohistochemistry findings showed that the characteristics of the tumor were consistent with inflammatory myofibroblastoma tumor (IMT).

Conclusions: we reported a rare case of intraocular IMT, which is confirmed by H&E staining, and IHC positive staining for Vimentin, Desmin and ALK, while negative staining for SMA, S-100, ki-67, CK, CD68, and calponin.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a distinctive lesion composed of myofibroblastic spindle cells with a prominent inflammatory infiltrate of lymphocytes, eosinophils and plasma cells[1]. Although IMT is relative rare, it is found more common in the lungs and can occur everywhere in the body[2]. However, few reports have announced IMTs in eyes. Here, we presente'd a case of a 55-year-old male with an intraocluar IMT of ciliary body for the first time.

Case Presentation

A 55-year-old male with no significant past ocular history presented with the chief complaint of progressively painless vision loss in the left eye over the course of twenty days without any other concomitant symptoms. The patient had no significant past medical history. His best corrected visual acuity (BVCA)in the left eye was 20/100, and the intraocular pressure (IOP) was 15 mmHg. Only an uneven opacity of the cortex in the left lens was found by slit-lamp biomicroscopy before the pupil was dilated. Therefore, the initial diagnosis of age-related cataract was made by outpatient physicians. To perform cataract surgery for the patient, the patient was refered to a surgeon. When the patient was given a dilated pupil examination, the surgeon revealed a well-marginated, ovoid soft achromatic colour mass filling the infernonasal quadrants under a slit lamp biomicroscopy. Superficial vessels could be seen on its surface and the opacity of the lens was mainly localized at the position where the mass was (Fig. 1A). A solid mass can be seen of the ciliary body from 6:30 to 8:30 by ultrasound biological microscope (UBM) examination (Fig. 1B). With ultrasonography, the tumor showed medium to high echodense and measured about 0.5-0.6-cm ovoid mass (Fig. 1C). By magnetic resonance imaging (MRI) examination, we also revealed a 0.5-0.6-cm well marginated, ovoid soft tissue mass and the signal intensity of the mass was slightly hyperintense on T1-weighted images(Fig. 2A) and markedly hypointense on T2-weighted images(Fig. 2B) and enhanced very well (Fig. 2C). Based on these findings, modified diagnosis was ciliary body neoplasm, secondary cataract of the left eye. The neoplasm squeezed the lens, influence its

metabolism, and then, lead to the opacity of the lens. Surgical intervention was performed not only to investigate the pathologic diagnosis, but also to relieve symptoms and improve vision. Instead of enucleation, sclerotomy with tumor biopsy was performed because of considering benign tumor. We performed trans-sclera resection via partial lamellar sclerouvectomy, par plans vitrectomy and silicon oil tamponade for the patient. The tumor was completely removed during the operation. Pathological examination was performed after complete removal of tumor. The light microscope showed that the tumor was composed of collagenized fibrous tissue, and in some areas, there were more smooth muscle-like cells showing tumor-like proliferation, mucus degeneration in the interstitium, and some chronic inflammatory cell infiltration in the surrounding area (Fig. 3A,D). Immunohistochemistry showed positive staining for anaplastic lymphoma kinase (ALK) (Fig. 3B,E), Desmin (Fig. 3C,F)and Vimentin(Fig. 3G,J), negative for smooth muscle actin(SMA) (Fig. 3H,K), S-100, ki-67 (Fig. 3I,L), CK, CD68, and calponin. The final diagnosis based on histology and immunohistochemistry was IMT of the ciliary body.

During the postoperative follow-up, the patient's visual acuity and eye condition were stable. At the latest follow-up, the best corrected visual acuity of this patient was 20/40 4 months after surgery.

Discussion

IMTs are mesenchymal neoplasm of intermediate biological potential with a predilection for the lung, abdomen, and pelvis of children and young adults [3]. Metastasis is a rare event in IMTs. Currently IMT is regarded as an intermediate, locally recurrent, rarely metastasizing neoplasm[4, 5].

IMTs are extremely rare in eyes, and most reported to be seen in the orbit. Intraocular IMTs appear to arise from uveal tissues and are associated with uveitis[6]. Common presentations include proptosis, ptosis, and painless loss of vision[7]. Dermarkarian et al [8]considered that these tumors were prone to invasively grow and be recurrence, so they recommended the complete removal as the first choice of treatment for this kind of tumor if possible. Usually, the diagnosis of IMTs may be delayed owing to nonspecific presenting symptoms[9]. Definitive diagnosis of IMTs relies on histopathologic examination with immunohistochemical characterization. The histopathology of IMTs are the variated product of erratic proportions of acute and chronic inflammation and fibrosis[10]. The principal components were plasma cells and spindle-shaped cells. Plasma cells were scattered throughout the lesions, often in small clusters, and predominated at the periphery. They were mature and occasionally large and multinucleated[2]. IMTs demonstrated positivity for vimentin (99%) and desmin (69%)[1]. Myofibroblasts express intermediate filaments and contain the phenotype VD, represented by cells positive for vimentin and desmin[11]. ALK expression has been documented in IMT cases[12]. The tumor has no positive response to myoglobin,cytokeratin, KP-1, CD30, and estrogen receptor[13]. This tumor can be difficult to distinguish from spindle cell tumors. The main differential diagnosis in the eye are leiomyomas and anaplastic large cell lymphomas.

Leiomyomas are rare benign tumors and commonly found in the uterus and alimentary tract[14].Light microscopically, the tumors are typically composed of interlacing bundles of spindle cells with blunt-

ended oval nuclei, moderate amounts of eosinophilic fibrillation cytoplasm, and intracellular myoglial fibrils[15]. Leiomyomas are most consistently reactive to SMA and uniformly negative for staining with ALK, S-100, and HMB-45 [16]. The tumor in our patient occurred in the ciliary body, and spindle-shaped cells were seen in H&E staining. But immunohistochemistry showed negative for staining with SMA and ALK. And this patient has chronic inflammatory cell infiltration, therefore, the diagnosis of leiomyoma is not considered.

ALK positive is not a specific manifestation of IMT. Some aspects of the ALK activation mechanism in IMT are also seen in some anaplastic large cell lymphomas(ALCLs)[1]. Light microscopically, the ALCLs are typically composed of interwoven fascicles of histiocytes and myofibroblasts mixed with chronic inflammatory cells, mostly T lymphocytes[17]. The tumor in our patient does not conform to ALCL because of the large proportion of collagenized fibrous tissue, and smooth muscle-like cells. This patient's immunohistochemical examination ki-67 (2%), cell proliferation is not active, so it is not considered as ALCL. And the primary intraocular lymphomas are mostly large B-cell lymphomas, a few are T-cell sources[18].

In our case, other possible tumors were excluded. Combined with the HE staining and immunohistochemistry results of the above patients, the diagnosis of primary intraocular inflammatory myofibroblastic tumor with ALK overexpression was considered.

However, there were several differences between this case and other typical cases of IMT reported in the past. In previous studies, IMTs demonstrated positivity for SMA (92%), because the most common phenotype of intermediate filaments expressed by myofibroblasts is phenotype VA: represented by cells positive for vimentin and [alpha]-smooth muscle actin, but there are also reports of phenotype VD[11]. This patient is in line with phenotype VD. ALK expression has been documented in 35%-60% of IMT cases[12]. ALK markers are mainly expressed as light to moderate cytoplasmic staining. In addition to the classic IMT, the literature also reported the existence of special epithelioid subtypes[19, 20]. There are few reports of intraocular IMT, so it is speculated that there may be other subtypes of IMT, and ALK stains in the nucleus.

In conclusion, we reported a rare case of intraocular IMT, which is confirmed by H&E staining, and IHC positive staining for Vimentin, Desmin and ALK, while negative staining for SMA, S-100, ki-67, CK, CD68, and calponin. As far as we know, this case of IMT is the first report of an intraocular IMT of ciliary body.

Abbreviations

IMT - Inflammatory myofibroblastic tumor
BVCA - best corrected visual acuity
IOP - intraocular pressure
UBM - ultrasound biological microscope
MRI - magnetic resonance imaging
ALK - anaplastic lymphoma kinase

SMA - smooth muscle actin
ALCLs - anaplastic large cell lymphomas

Declarations

Ethic and consent

Ethics approval and consent to participate

Availability of data and materials

All data generated or analysed during this study are included in this published article [and its supplementary information files].

Consent

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

Competing interest

The authors declare that they have no competing interests.

Funding

No applicable

Authors’ contributions

HWQ pathologically examined the specimen, including H&E and IHC staining. ZZY participated in the design of the study, performed the operation, and drafted the manuscript. YH performed the image cutting and drafted the manuscript. TNT analyzed the results and helped draft the manuscript. All authors have read and approved the final manuscript.

Acknowledgements

The authors would like to thank Hui Zong performed a detailed ophthalmic examination for this patient.

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Figures

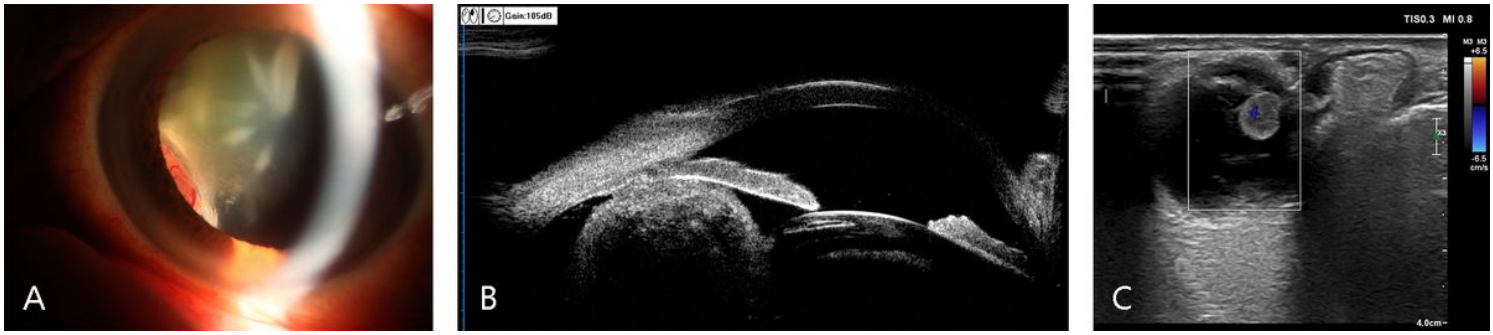

Figure 1

Under the slit lamp, superficial blood vessels can be seen on the surface, and the opacity of the lens is mainly located at the location of the mass. (A). Ultrasound biomicroscopy image show a solid mass of the ciliary body from 6:30 to 8:30 (B). The ultrasonography image shows a mass of about 0.5-0.6cm in size with medium-high echo (C).

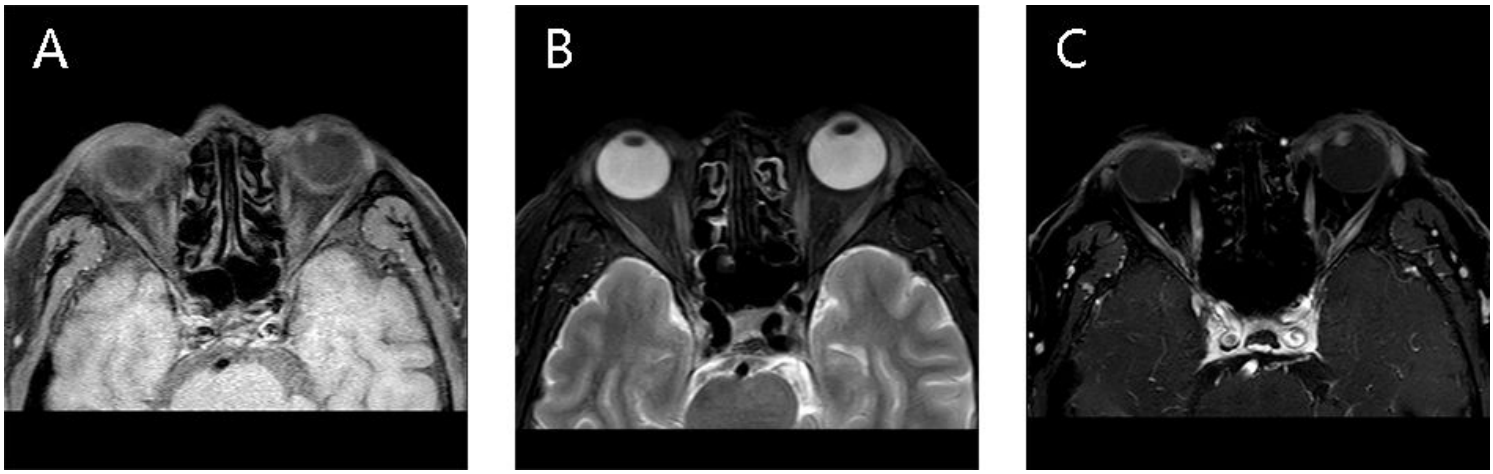

Figure 2

MRI of the eyeball and orbit. In the lens of the left eye, the signal intensity of the mass was slightly hyperintense on T1-weighted images (A) and markedly hypointense on T2-weighted images (B) and enhanced very well (C).

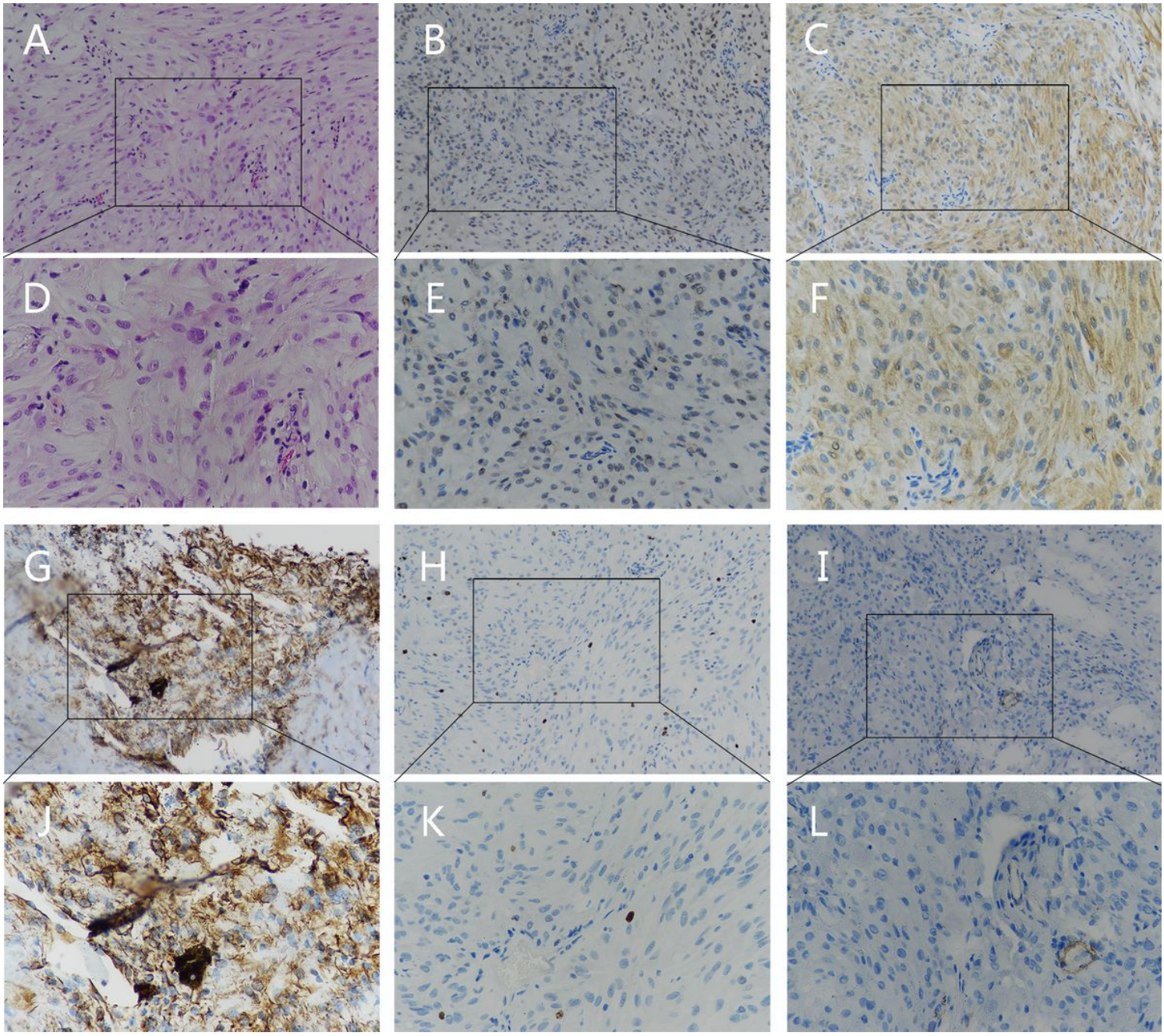

Figure 3

HE staining and IHC staining for ALK, Desmin, Vimentin, ki-67 and SMA. HE staining shows smooth muscle-like cells and some chronic inflammatory cell infiltration in the surrounding area(A 20×,D 40×) The positive staining images of ALK, Desmin and Vimentin are shown in (B 20×,E 40×), (C 20×,F 40×) and (G 20×,J 40×), respectively. While negative staining images of ki-67 and SMA are shown in (H 20×,K 40×) and (I 20×,L 40×), respectively.