Case Report

Ciliary Body Leiomyoma

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Keywords
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Abstract
The report aims to present the case of intraocular leiomyoma. We conducted a case study on a patient who presented with an intraocular tumour. After examination, including magnetic resonance, positron emission tomography with computed tomography, B-scan, we performed surgery – enucleation of the eye globe with histological verification of tumour mass. Histological analysis of enucleated eyes proved intraocular leiomyoma. Leiomyoma is a rare intraocular tumour, which is clinically challenging to recognize; therefore, histological confirmation is most often required.

Introduction
Intraocular leiomyoma is a rare benign tumour. Leiomyomas are more often found in the uterus or gastrointestinal tract. However, there have been reported cases of leiomyomas in ophthalmology, often localized in the ciliary body, choroid, or iris [1, 2]. Leiomyomas, although classified as benign tumours, are locally destructive to intraocular structures. In most cases, they occur in the anterior part of the uvea and clinically manifest similarly to uveal melanoma [3]. As a result of its rarity and clinical presentation, this kind of tumour is regularly diagnosed after enucleation by histopathologic analysis of tumour tissue.
Case Presentation

A 54-year-old female patient referred from the outpatient ophthalmology department was examined in March 2021. In the past, she underwent surgery due to uterine myomas. She planned to have cataract surgery but, due to the COVID pandemic, she cancelled her appointment. Subjectively patient was complaining of progressive worsening of vision in the right eye and did not have any other symptoms. Her visual acuity at first examination was 20/160 in the right eye and 20/50; +1Dsph 20/20 in the left eye. We conducted the objective examination with slit-lamp biomicroscopy to establish the anterior segment of the eye without visible pathology. However, the investigation with mydriatic pupil discovered a double lobe mass of grey colouring stretching from h 11 to h 1, reaching almost to the centre of the pupil. Because of this suspect find, we proceeded with ultrasonography of the right eye and found the hyperechogenic focus measuring 13.68 × 12.98 mm, filling a substantial part of the vitreous cavity (shown in Fig. 1). We concluded the anatomical location of the lesion in the anterior part of the uvea, which we later confirmed by magnetic resonance imaging (MRI).

We proposed that the patient undergo MRI and positron emission tomography with computed tomography to establish the severity of the lesion. The result of the MRI in March 2021 confirmed the presence of a tumour in the ciliary body in the right eye, measuring 17 × 10 mm without signs of extrascleral proliferation. In T2-weighted images appeared as hypointense4 and in T1-weighted images appeared as isointense (shown in Fig. 2). Conclusion of positron emission tomography with computed tomography established metabolic active intraocular tumour growth in the right eye, classified as primary tumour, as there was no pathologically increased accumulation of 18F-fluoro-2-deoxy-D-glucose in the scans, concluded as without the nature of possible dissemination of underlying disease (shown in Fig. 3). Due to the severity of tumour volume over 0.8 cm3 and low visual acuity, we recommended radical surgical therapy in the form of enucleation of the eye to the patient. The pathologist in histopathological analysis analysed the enucleated eye globe. The result was of tissue of the eyeball with a tumour in the area of the ciliary body. The tumour was well-demarcated, consisted of bundles of uniform, slender spindle cells, often vacuolated cytoplasm, without increased mitotic activity, cells stained greyish-yellow when stained by van Gieson method, showed positivity for HS actin, S100−, GFAP−, CD57−, melan-A−, Ki67+ <1%, calponin+, desmin+, CD56+, neuron-specific enolase+. Histopathological analysis closed the matter as leiomyoma of the ciliary body likely of mesectodermal origin due to positivity of CD56 and neuron-specific enolase, shown

Fig. 1. Image of ultrasonography of the right eye. Hyperechogenic focus measuring 12.68 × 13.98 mm filling most of the intrabulbar cavity.
Discussion

The most common presenting symptom of leiomyoma in patients is blurred vision, but leiomyomas are slow-growing tumors, and symptoms are proportional to tumor size. In some cases, leiomyomas present without clinical symptoms, and the diagnosis of tumor mass in a patient is coincidental [4]. Most leiomyomas occur in relatively young patients, primarily women. Leiomyomas are considered smooth, amelanotic rubbery mass, in some cases covered by pigmented layer and mostly of ovoid shape [5]. Ciliary body tumors are challenging to diagnose; there are many options to consider, from malignant melanoma to benign leiomyoma. The other lesion, such as medulloepithelioma, hemangiopericytoma, or schwannoma, is very rare, even though they should be considered in clinical differential diagnostics of amelanotic tumors. Ciliary body schwannoma was documented by Küchle et al. [6] and Goto et al. [7]. Medulloepitheliomas are primarily seen in children, and histopathologically the tumor is classified as teratoid or nonteratoid based on elements it contains. The systemic prognosis of such tumors is favorable, but with extrascleral extension, the chance of metastasis spread is higher [8]. Typically, it is impossible to distinguish between uveal melanoma and leiomyoma.
Both tumours share light microscopic appearance. Therefore, immunohistochemistry or transmission electron microscopy after enucleation is needed [9, 10]. Leiomyoma appears in T1-weighted images isointense to hyperintense and has exhibited marked signal enhancement with gadolinium.
In T2-weighted images, leiomyomas exhibit low signal. In cases of uveal melanoma, the tumour appeared hypointense in T2-weighted images and hyperintense in T1-weighted images. Due to the relatively similar appearance, we cannot confirm the diagnosis and choose a less invasive treatment plan based only on MRI[11]. Fine needle aspiration biopsy while a useful method for diagnostics can lead to tumour seeding and subsequently to metastatic growth [12].

Leiomyomas are composed of intracellular myoglial fibrils, interlacing bundles of spindle cells with oval nuclei and a moderate amount of eosinophilic fibrillary cytoplasm [1]. Uveal leiomyomas are known to be divided into two groups, those that are of mesodermal origin and those that are of mesectodermal origin [13]. The smooth muscle filaments stain with muscle-specific actin, desmin, vimentin, h-caldesmon, and α-smooth muscle actin [14]. Leiomyomas are uniformly reactive to smooth muscle actin and muscle-specific actin and constantly negative for staining with melanoma-specific markers. They can have positive staining with S-100, neuron-specific enolase and CD-56 and negative staining for melanoma-specific markers such as HBM-45 and Melan-A [15]. There are many treatment options in onco-ophthalmology according to diagnosis, clinical presentation, the prognosis of vision retention, bulbus preservation, metastatic disease.

Due to difficulty of differentiating between leiomyoma and uveal melanoma, the most common treatment option is enucleation or radiation. An enucleation is a radical form of treatment for tumours, with tissue availability for histopathological confirmation of disease. The downside of such radical treatment lies in permanent loss of vision and the need for monocular precautions. However, depending on tumour sizes, it is the only logical way to proceed in treatment and bring the patients better quality of life. There is a possibility of simply observing the tumour and in time when the mass starts to progress in size and clinical symptoms manifest in a patient, the oncologist may proceed to conservative way of treatment such as radiation that may slow the growth. Radiation is globe preserving treatment, depending on the type of radiation there are side effects such as skin excoriation, cataract, radiation retinopathy, neovascular glaucoma, and others [11].

In some cases, radiation treatment may in years led to enucleation because of the side effects of the treatment. Leiomyomas are benign non-life-threatening tumours. However, they are slow growing, locally destructive, and may cause exudative retinal detachment or other ocular complications such as neovascular glaucoma. In the end, the prognosis for vision and ocular retention is determined by size, location, and progression of leiomyoma.

**Statement of Ethics**

This article does not include results of experimental investigations on humans; the presented research complies with guidelines for human studies. We confirm that the patient provided written, informed consent to publish the case details and images. The consent form is included in medical record of the patient. Ethical approval is not required for this study in accordance with local or regional guidelines.

**Conflict of Interest Statement**

The authors have no conflict of interest to declare.

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Author Contributions

Paulina Plesníková: formal analysis, investigation, and writing – original draft preparation; Denisa Jurenová: formal analysis, investigation, and writing; Darina Lysková: formal analysis, investigation, and writing; Alena Furdová: supervision, resources, investigation, and analysis; Jela Valášková: analysis, resources, and review of the manuscript; Pavel Babál: investigation, analysis, and resources. All authors made significant contributions to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis, and interpretation, or in all these areas; took part drafting, revising, or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

Data Availability Statement

All data supporting reported case can be found in this article.

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