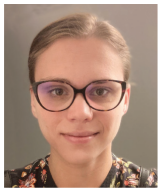
# LEIOMYOMA – CILIARY BODY TUMOR. A CASE REPORT

**Černá Karolína[1], Hlinomazová Zuzana[1], Glezgová Johana[2], Ambrosová Kateřina[3,4]**

**[1]Lexum a.s., Czech Republic**
**[2]Department of Ophthalmology, First Faculty of Medicine, Charles University, Prague, Czech Republic**
**[3]Regional Hospital Mladá Boleslav a.s., Hospital of the Central Bohemian Region, Czech Republic**
**[4]Laboratory of pathology a cytology, Unilabs pathology k.s., Laboratory Prague, Czech Republic**

*The authors of the study declare that no conflict of interests exists in the compilation, theme and subsequent publication of this professional communication, and that it is not supported by any pharmaceuticals company. The study has not been submitted to any other journal or printed elsewhere, with the exception of congress abstracts and recommended procedures.*

Submitted to the editorial board: December 13, 2023
Accepted for publication: April 24, 2024
Available on-line: June 25, 2024



MUDr. Karolína Černá
Oční klinika LEXUM Mladá Boleslav
Na Celně 885
293 01 Mladá Boleslav
E-mail: cernak@lexum.cz

## SUMMARY

**Aim:** To demonstrate a rare case of ciliary body leiomyoma in our patient
**Case report:** A 72-year-old female reported to our clinic for a preventive examination, upon which we found a dome-shaped grey-brownish mass on the retinal periphery. After completing gonioscopic and ultrasound examinations, we referred the patient to a specialist facility. Due to a finding of suspicious malignant melanoma, we completed the MRI scan and recommended enucleation of the eyeball. A histopathological examination showed a leiomyoma of the ciliary body.
**Conclusion:** The aim of this case report is to demonstrate the difficulty of intraocular leiomyoma diagnosis. Only immunohistochemical examination differentiated the tumor from malignant melanoma and determined the diagnosis of ciliary body leiomyoma. Perhaps because of the extreme rarity of this type of tumor, we often neglect to consider a diagnosis of leiomyoma.
**Key words:** leiomyoma, ciliary body, melanoma, intraocular tumor

*Čes. a slov. Oftal., 80, 2024, No. x, p.*

## INTRODUCTION

Intraocular leiomyoma is an extremely rare tumor of the iris, ciliary body and choroid [1–4]. Although this concerns a benign tumor, it is locally destructive to the intraocular structures [5]. Like other tumors in this localization, it may cause retinal detachment, secondary cataract and secondary glaucoma [2]. It most commonly occurs in young women of reproductive age [1,3,6,7]. Out of 80 described cases of uveal leiomyoma published by Ankit in 2019, 51 occurred in women [1]. The average age at the time of identification was 35.8 years, in which the average age regarding leiomyoma of the iris was 44.6 years, of the ciliary body 35.5 years and of the choroid 35.4 years [1]. Despite the fact that macroscopically this most often concerns amelanotic ovoid tumors with a smooth surface, darkly pigmented lesions are also described [1,8]. Histopathologically, the tumor is composed of intertwined bundles of spindle cells with oval nuclei, which can be distinguished from uveal melanoma by immunohistochemical examination and electron microscopy [1].

## CASE REPORT

In January 2023, within the framework of long-term observation of diabetic retinopathy, a 72-year-old patient with glaucoma and type 2 diabetes reported to our ophthalmological outpatient center in Mladá Boleslav for a regular preventive examination. The patient had been observed regularly at the ophthalmological outpatient center since 2005. In February 2013 she had been diagnosed with suspected primary open angle glaucoma, and regular preventive examinations were set for her every six months, including perimetry monitoring and control analysis of the retinal nerve fiber layer (RNFL analysis). The patient was also diagnosed with incipient senile cataract. In 2019 we diagnosed primary open angle glaucoma due to thinning in the retinal nerve fiber layer (RNFL). Other important data in the patient's per-

sonal medical history are well compensated arterial hypertension, hypothyroidism, post-stroke condition of the character of vertebrobasilar insufficiency, and a cyst on the left kidney. The patient's family anamnesis includes Hodgkin's lymphoma in her son.

To date the patient has been without subjective ocular complaints, best corrected visual acuity in the right and left eye is 0.8 for distance vision, and 0.8 for near vision with addition of +2.50 Dsf. Intraocular pressure measured by non-contact tonometry was 20 mmHg in the right eye and 24 mmHg in the left eye. Control RNFL analysis detected a bilateral reduction in the RNFL and in the ganglion cell layer. Examination on a slit lamp revealed incipient senile corticonuclear cataract, on the ocular fundus in mydriasis the papilla manifested wide glaucomatous excavation C/D 0.7 bilaterally and peripapillary pigment margin, macula without lesion changes, in the right eye the retinal periphery was without lesion changes and in the left eye a dome-shaped grey-brownish mass was present on the retinal periphery, without secondary collateral retinal detachment and without hemorrhages. The patient was subsequently referred to the Lexum eye clinic in Mladá Boleslav for a more detailed examination.

On 27 January 2023 we additionally performed a gonioscopic examination on the patient, with a finding of a medium-wide markedly pigmented angle, without a pathological formation, and an examination using a Goldmann lens, with a finding of a dome-shaped grey--brownish mass in the region of the ciliary body, and an ultrasound examination (Figure 1), which confirmed a solid lesion temporally behind the iris, without collateral retinal detachment, with a size of 8.9 x 4.7 mm. On anterior segment optic coherence tomography (AS-OCT) we did not identify the solid lesion. We referred the patient for an examination at the Ocular Oncology Center at the General University Hospital in Prague due to a conclusion of suspected melanoma of the ciliary body.

On 8 February 2023 the patient underwent a consultation examination at that center. Pronounced episcleral blood vessels were described in the left eye nasally, as well as a lens with incipient corticonuclear cataract without sector opacity, and in mydriasis on the retinal periphery temporally between nos. 2 and 4 a projecting dome-shaped lesion of dark brown color. Upon performance of transillumination, a shadow was visible in the left eye fixed on the ciliary body in the superior temporal quadrant. An ultrasound A+B scan showed a markedly projecting lesion of rather lower internal reflectivity, diagonally identified for a localization of approximately 7 mm, and a flatter lesion of less than 3 mm, out of which a unilocular lesion of approximately 6 mm was growing. The conclusion of the examination was suspected malignant melanoma of the ciliary body in the left eye, with a recommendation for the soonest possible appointment for magnetic resonance (MR) of the brain and eye sockets, focusing on the left orbit, and measurement of the solid lesion in the left eye.

MR examination of the brain and orbits was performed on three levels, in prepared turbo spin echo (TSE), inversion recovery (IR)-TSE, diffusion-weighted imaging (DWI), fast or turbo spin echo FSE/TSE, gradient echo sequences (GRE) natively.

Finding: Central structures of the brain without dislocation. Cerebral ventricular system commensurately wide, cerebral aqueduct with through passage. The subarachnoid spaces were supratentorially, primarily frontally and parietally widened. Wider subarachnoid spaces also in the surrounding area of the brain hemispheres. Brain structures of usual configuration. In the white matter of both brain hemispheres primarily subcortically in the frontal and parietal lobes, multiple lesions of gliosis individually up to 10 mm. No lesion changes of a different character or inferotemporal lesions were identified. The right maxillary sinus was reduced, filled with chronic mucosal changes, on the left a small polyp, otherwise the paranasal sinuses and mastoid cavities were airy, inner ear canals slim, hypophysis not enlarged. Orbits of normal configuration. Commensurate finding on right. On the left within the region of the ciliary body a spin-

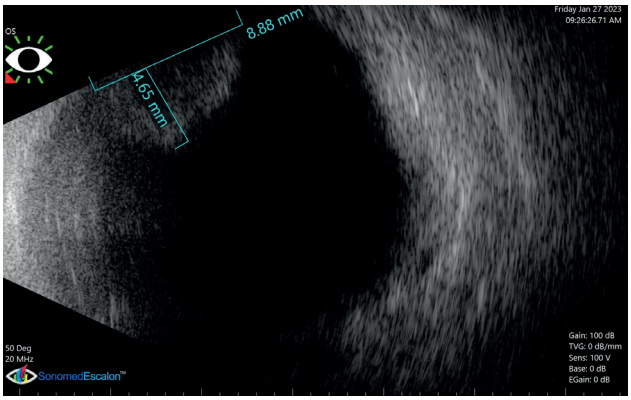



**Figure 1.** Ultrasound image with prominent spherical lesion in the upper temporal quadrant

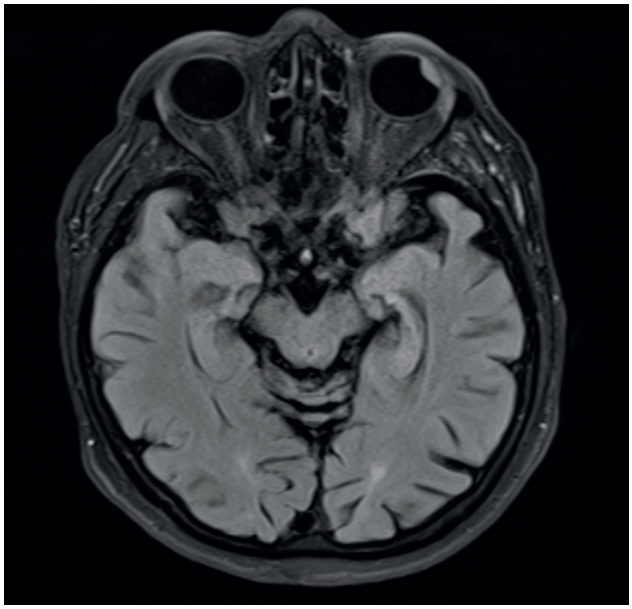


**Figure 2.** MRI with spindle-shaped lesion in the superior temporal quadrant of the left bulb

**Figure 3.** Enucleated bulbus with prominent lesion temporally

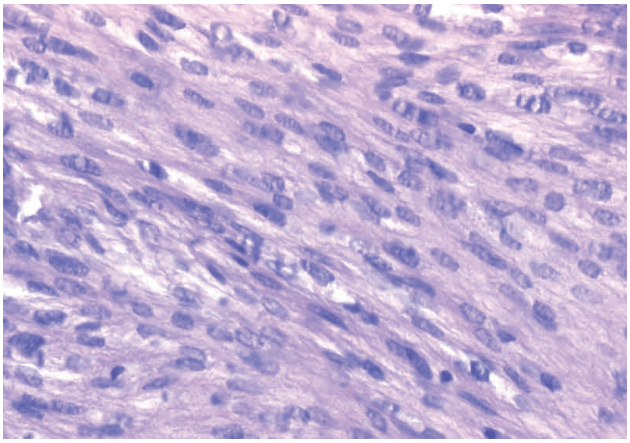


**Figure 4.** Microscopic image of tumor composed of the spindle cells in short parallel bundles

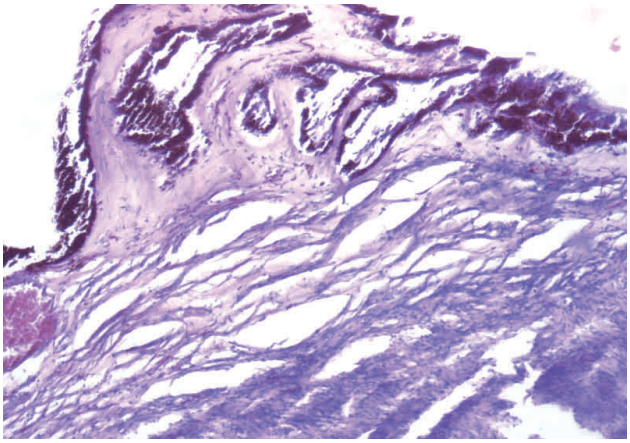


**Figure 5.** Microscopic image of well circumscribed tumor of choroid and ciliary body

dle-shaped lesion with a size of 11x5x10 mm was present in the superior temporal quadrant (Figure 2). The lesion had a medium signal in T2 and T1W, manifesting partial restriction of diffusion. The lesions did not manifest periocular growth. The surrounding structures, glandula lacrimalis and oculomotor muscles were of normal width, course, signal. Optic nerves, sheaths and chiasm with normal finding. Conclusion: Lesion – melanoma of the ciliary body within the superior temporal quadrant in the left eye without infiltration of the surrounding area. Cortical atrophy of the brain. Mild atrophy of cerebellum. Multiple lesions of vascular gliosis in white matter of both hemispheres. Examination native, the patient refused a contrast agent.

The finding on magnetic resonance was consulted with the Ocular Oncology Center at the General University Hospital in Prague. Due to the size of the tumor, enucleation was recommended in the patient's locality of residence. Because the patient refused the administration of a contrast agent and radiopharmaceuticals we did not indicate PET/CT.

On 21 March 2023 the patient was admitted to the local department of ophthalmology at the Regional Hospital in Mladá Boleslav for enucleation of the left eyeball under general anesthesia. On 23 March 2023 enucleation of the eyeball was performed using the standard procedure, with insertion of an orbital implant with a diameter of 20 mm and a cross node of the rectus muscles above the implant. The procedure took place without complications. Postoperatively the patient applied antibiotic oitment with corticoid (bacitracin zinc, neomycin sulfate, hydrocortisone acetate) to the eye socket 4–5 x per day for a period of 2 weeks. On 27 March 2023 the patient was in a generally well stabilized condition and was discharged to go home. Within the framework of the preoperative examination we also added X-ray of the heart and lungs, which was without any pathological finding. With regard to general screening incorporating ultrasound examination of the abdomen, mammograph, gynecological examination, X-ray of heart and lungs and sampling of tumor markers we awaited the results of the pathological examination.

The pathological finding is demonstrated in the region of the ciliary body and anterior choroid by a temporally whitish tumor with a diameter of 12 mm and a thickness of 7 mm (Figure 3).

Microscopically, the ciliary body shows a spindle cell fascicularly arranged tumor without cytological atypia and without increased mitotic activity. (Figure 4). The tumor was well bordered and without signs of invasive growth and angioinvasion (Figure 5). Immunohistochemically in the tumor cells negative s100, HMB45 and Melan-A (Figure. 6). Immunohistochemically in the tumor cells strongly positive actin and h-caldesmon (Figure 7). Proliferation activity in staining Ki67 was low (less than 5 %). The conclusion of the pathological examination was leiomyoma of the ciliary body. Because leiomyoma is a benign tumor, we did not perform any further general examinations.

Postoperatively the patient's development was without complications, locally we retained a combined preparation in the form of cream for 3 weeks, and due to foamy secretion also for one further week antibiotic oitment with tobramy-

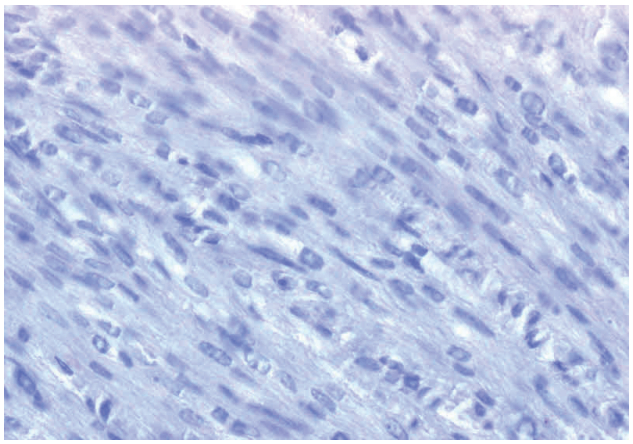


**Figure 6.** Microscopic image of tumor with negativity of HMB and MelanA markers

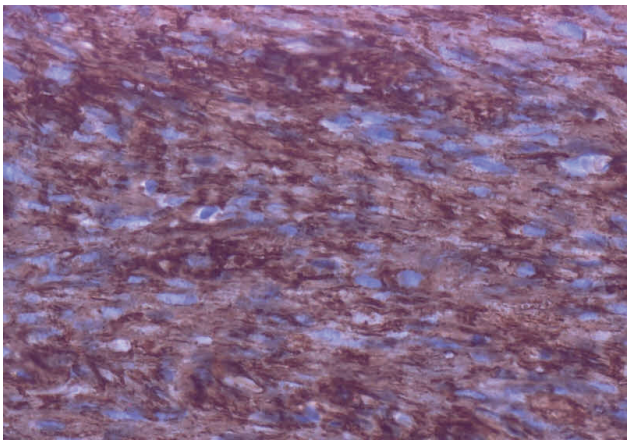


**Figure 7.** Microscopic image of tumor with positivity of smooth muscle actin (SMA) and h-caldesmon markers

cin 1x per day supplemented with rinsing of the eye socket with boric acid. Due to the pacific finding, the production of a glass prosthesis was ordered for the patient.

The patient is now 5 months after enucleation, under observation with cataract in the right eye, due to thinning in the RNFL her antiglaucoma medication was increased to a fixed combination of timolol-latanoprost. Our present objective is to ensure appropriate timing for cataract surgery on the patient's right eye, and to monitor the postoperative condition of the eye socket, which is now calm, without pathological secretion or fissures.

## DISCUSSION

Clinically it is difficult to differentiate leiomyoma from malignant melanoma [1,2,6–10]. In addition to examination on a slit lamp, fundus in mydriasis and gonioscopy, the core examination modality in the diagnosis of intraocular tumors is ultrasound examination [11–13]. An A-scan provides us with information about the internal echogenicity of the lesion, while we use a B-scan for measuring posteriorly located lesions [12]. Ultrasound biomicroscopy (UBM) is of essential importance for the diagnosis of lesions within the region of the ciliary body with an iris component, in differentiating them from tumors of the iris and peripheral choroidal tumors [1,11–14]. On an A-scan, a typical image of uveal melanoma is a high first peak and low to medium reflectivity of the tumor [13,15]. In the case of leiomyoma high, medium and low reflectivity has been described, and as a result ultrasound examination may not differentiate leiomyoma from uveal melanoma [1]. In comparison with AS-OCT, which is fast and does not require direct contact with the eye, a disadvantage of UBM is the time demand factor, the necessity for performance of the examination by an experienced doctor and direct contact with the eye [14]. On the other hand, a negative factor of AS-OCT is the lower penetration of the pigmented tissue [14]. Transscleral transillumination is another noninvasive diagnostic examination which may assist us in determining the characteristics of an anterior uveal tumor [1,3].

Fine-needle aspiration biopsy is indicated especially in unclear cases [16]. It is performed using a 25–30G needle both sclerally and transvitreally, and so its potential complications include hemorrhage, iatrogenic ocular affliction and the risk of seeding of the tumor [1,17,18]. A negative cytological finding of malignity should not be considered unequivocal evidence excluding the possibility of malignity [18].

MR plays an essential role in the detection of a tumor, determination of its character, progression, planning of treatment and monitoring the effect of treatment [13]. The basic MR protocol in the diagnosis of uveal melanoma contains T1 and T2 weighted scans before and after the administration of the contrast agent (gadolinium) [19]. Typical uveal melanoma is hyperintense on a T1 weighted scan, and hypointense on a T2 weighted scan [19]. Leiomyoma is isointense to hyperintense on a T1 weighted scan and hypointense on a T2 weighted scan [5]. Upon suspicion of a metastatic process and in the case that we do not know the primary lesion of the tumor, biochemical examination is indicated, as well as X-ray of the heart and lungs, ultrasound of the abdomen, examination of thyroid gland hormones, skin examination, tumor markers, full-body computer tomography (CT), fibroscopy of the stomach, colonoscopy, skeletal scintigraphy, in women a gynecological examination and mammography, in men a prostate examination [15]. In the case of a negative result then PET or SPECT [15]. Uveal melanoma is the most common primary intraocular tumor, which metastasizes most often in the liver, then in the lungs and bones [13]. The goal of treating uveal melanoma is to preserve the eyeball, its functions and to prevent metastatic dissemination [13]. We select the therapeutic procedure according to the stage of the pathology. The approach to treatment should be multidisciplinary. The multidisciplinary team should contain an ophthalmologist, radiologist and specialist in radiation oncology [13,20]. We select the treatment according to the stage of the pathology [20,21]. Current options for the treatment of uveal melanoma are regular observation, transpupillary thermotherapy, brachytherapy using radionuclides Ru106 and I-125, teleradiotherapy and surgical treatment from eyeball preserving pro-

cedures to enucleation and exenteration of the orbit in the case of advanced tumors with extrabulbar spread [20,21].

Within differential diagnostics, we also have to take into consideration metastasis of breast carcinoma and bronchogenic carcinoma, schwannoma, neurofibroma, melanocytoma, adenoma and adenocarcinoma of the epithelium of the ciliary body, medulloepithelioma and choroidal hemangioma [7,10]. We must then take melanoma and schwannoma into consideration also within microscopic differential diagnostics [22]. Metastases are the most common intraocular tumor with typical localization on the posterior pole of the choroid [23]. This frequently concerns multiple lesions, in 20–40% of cases bilateral flat lesions of a yellowish, creamy or less commonly orange color associated with the presence of subretinal fluid [2,23]. By contrast with melanoma, on an ultrasound A-scan intraocular metastasis is typical in its higher internal reflectivity [23]. The most common primary lesion is breast carcinoma, followed by bronchogenic carcinoma [2,23,24]. Intraocular schwannoma is an extremely rare benign tumor of the ciliary body and choroid, less often of the iris [25]. In clinical practice, as with leiomyoma it may be mistaken for amelanotic melanoma, and is only differentiated from uveal melanoma by immunohistochemical examination [25]. Uveal neurofibromas are highly associated with neurofibromatosis, and concern diffuse lesions of the choroid [2]. Melanocytoma is a benign pigmented lesion most frequently in close proximity to the optic nerve, the method of treatment is monitoring [26,27]. Adenomas and adenocarcinomas of the epithelium of the ciliary body are very rare solid tumors, which are often mistaken for melanoma and treated with brachytherapy or enucleation [2,28,29]. Medulloepithelioma is a unilateral tumor occurring in childhood age, the average age at the time of diagnosis is 5 years and therapy is usually enucleation [2,30]. Choroidal hemangioma is an amelanotic red-orange mass of dome-shaped or placoid appearance, and occurs most frequently on the posterior pole [14,31]. In contrast with melanoma, with which it may be confused, on an ultrasound A-scan it is typical in its higher internal reflectivity, washout phenomenon upon angiography of the eye with indocyanine green [14,31,32].

## CONCLUSION

The objective of this case report was to demonstrate the difficulty of diagnosing intraocular leiomyoma. Several clinical and imaging examinations were performed, and all pointed to a diagnosis of melanoma of the ciliary body. Diagnosis of intraocular metastasis was excluded by clinical and ultrasound examination. It was only immunohistochemical examination that differentiated the tumor from malignant melanoma and confirmed a diagnosis of leiomyoma of the ciliary body. Despite the fact that these two tumors cannot be clinically differentiated, we should consider a diagnosis of leiomyoma in the case that we find the tumor in younger patients, and especially women. In the case of leiomyoma the localization of the tumor is more frequently in the region of the ciliary body and the periphery of the choroid, and affects the supraciliary and suprachoroidal space [9]. By contrast, melanoma is more commonly located centrally and affects the stroma [9]. Upon suspicion of a malignant intraocular lesion it is necessary to perform multimodal imaging and to ensure close cooperation of an ophthalmologist, radiologist and radiation oncologist.

Acknowledgments: Thanks to MUDr. Kateřina Zelenková, FEBO, who was also an operating surgeon, for providing photography of the enucleated eyeball.

## REFERENCES

1. Tomar AS, Finger PT, Iacob CE. Intraocular leiomyoma: Current concepts. Surv Ophthalmol. 2020 Jul;65(4):421-437.
2. Sadda SR, Wilkinson CP, Wiedemann P, Schachat AP, editors. Ryan's Retina. Volume 3/ editor-in-chief SriniVas R. Sadda, MD (Professor of Ophthalmology, Doheny Eye Institute, David Geffen School of Medicine, University of California, Los Angeles, Los Angeles, CA, USA). Sixth edition. London New York Oxford Philadelphia St Louis Sydney: Elsevier;2018. 1821 p.
3. Shields C, Shields J, Remmer M, Kaliki S, Eagle R. Giant leiomyoma of the ciliary body. Oman J Ophthalmol. 2014;7(2):81.
4. Biswas J, Kumar SK, Gopal L, Bhende MP. Leiomyoma of the Ciliary Body Extending to Anterior Chamber. Surv Ophthalmol. 2000 Jan;44(4):336-342.
5. Plesníková P, Jurenová D, Lysková D, Furdová A, Valášková J, Babál P. Ciliary Body Leiomyoma. Case Rep Ophthalmol. 2022 April 5;13(1):259-264.
6. Kim JM, Hall LB, Elia M, Ehrlich MS, Materin MA, Sinard JH. Acute Presentation of Mesectodermal Leiomyoma of the Ciliary Body. Ocul Oncol Pathol. 2017;3(4):304-309.
7. Fernandez de Castro JP, Syed NA. Clinicopathologic Correlation: Ciliary Body Leiomyoma. EyeRounds.org. Sept. 9, 2009; Available from: http://webeye.ophth.uiowa.edu/eyeforum/cases/64-Leiomyoma-Clinicopathologic.htm
8. Tomar AS, Fam A, Finger PT, Iacob CE. Doppelgänger dilemna: Leiomyoma versus uveal melanoma. AM J Ophthalmol Case Rep. 2021 Jun;22:101040.
9. Oh KJ, Kwon BJ, Han MH et al. MR imaging findings of uveal leiomyoma: three cases. AJNR Am J Neuroradiol. 2005 Jan;26(1):100-103.
10. Zhou N, Wei W, Xu X. Pathology features and the recults of treatment of two cases of posterior choroidal leiomyoma. BMC Ophthalmol. 2020 Dec;20(1):198.
11. Salmon JF, Kanski JJ. Kanski's clinical ophthalmology: a systematic approach. 9th edition. Erscheinungsort nicht ermittelbar: Elsevier; 2020. p.941.
12. Kadaika A, Zhang J, Yao X, Zhou Q, Heiferman MJ. Ultrasound in ocular oncology: Technical advances, clinical applications, and limitations. Exp Biol Med. 2023 May 22;248(5):371-379.
13. Foti PV, Travali M, Farina R, et al. Diagnostic methods and therapeutic options of uveal melanoma with emphasis on MR imaging-Part I: MR imaging with pathologic correlation and technical considerations. Insights Imaging. 2021 Jun 3;12(1):66.
14. Medina CA, Plesec T, Singh AD. Optical coherence tomography imaging of ocular and periocular tumours. Br J ophthalmol. 2014 Jul 1;98(Suppl 2):ii40-46.
15. Kuchynka P. Oční lékařství. 2.vyd. Praha:Grada;2016.936. Czech.
16. Rusňák Š, Hecová L, Kasl Z, Sobotová M, Hauer L. Uveal Melanoma Biopsy. A review. Cesk Slov Oftalmol. 2020 Aug 30;76(6):247-252.
17. Frizziero L, Midena E, Trainiti S, et al. Uveal Melanoma Biopsy: A Re-

view. Cancers. 2019 Jul 30;11(8):1075.
18. Singh AD, Biscotti CV. Fine needle aspiration biopsy of ophthalmic tumors. Saudi J Ophthalmol. 2012 Apr 1;26(2):117-123.
19. Solnik M, Paduszyńska N, Czarnecka AM, et al. Imaging of Uveal Melanoma-Current Standard and Methods in Development. Cancers. 2022 Jan;14(13):3147.
20. Foti PV, Travali M, Farina R, et al. Diagnostic methods and therapeutic options of uveal melanoma with emphasis on MR imaging-Part II: treatment indications and complications, Insights Imaging. 2021 Jun 4;12(1):67.
21. Rusňák Š, Hecová L, Kasl Z, Sobotová M, Hauer L, Therapy of Uveal Melanoma. A Review. Cesk Slov Oftalmol. 2021 Aug 30;77(1):1-13.
22. Grossniklaus HE, Eberhart CG, Kivelä TT. WHO classification of tumours of the eye. 4th ed. Lyon: International agency for research on cancer;2018. (Worlh health organization classification of tumours).
23. Konstantinidis L, Damato B. Intraocular Metastases-A Review. Asia-Pac J Ophthalmol. 2017 Apr;6(2):208.
24. Jindal A, Kapatia G, Gupta G. Ocular metastasis from renal malignacies - A comprehensive review. Indian J Ophthalmol. 2023 Oct;71(10):3281.
25. Yu Y, Cheng Y, Wang K, Sun K, Shen D, Liang J. Intraocular schwannoma: A case series of 3 patients. Oncol Lett. 2019 Jan;17(1):1274-1278.
26. Reidy JJ, Apple DJ, Steinmetz RL, et al. Melanocytoma: nomenclature, pathogenesis, natural history and treatment. Surv Ophthalmol. 1985;29(5):319-327.
27. Shields JA, Demirci H, Mashayekhi A, Eagle RC, Shields CL. Melanocytoma of the optic disk: A review. Indian J Ophthalmol. 2019 Dec;67(12):1949-1958.
28. Pecorella I, Ciocci L, Modesti M, Appolloni R. Adenoma of the non-pigmented ciliary epithelium: A rare intraocular tumor with unusual immunohistochemical findings. Pathol - Res Pract. 2009 Dec 15;205(12):870-875.
29. Mansoor S, Qureshi A. Ciliary body adenoma of non-pigmented epithelium. J Clin Pathol. 2004. Sep;57(9):997-998.
30. Peshtani A, Kaliki S, Eagle RC, Shields CL. Medulloepithelioma: A triad of clinical features. Oman J Ophthalmol. 2014;7(2):93-95.
31. https://fyra.io. Retina Today. Bryn Mawr Communications; [cited 2024 March 13]. Choroidal Hemangioma Masquerading as Central Serous Chorioretinopathy. Available from: https://retinatoday.com/articles/2012-apr/choroidal-hemangioma-masquerading-as-central-serous-chorioretinopathy
32. Karimi S, Nourinia R, Mashayekhi A. Circumscribed Choroidal Hemangioma. J Ophthalmic Vis Res. 2015;10(3):320-328.