OPHTALMOLOGY

CASE REPORT OF SCLEROCHOROIDAL CALCIFICATION

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ABSTRACT

Background. *Sclerochoroidal calcification is an idiopathic rare benign lesion of the sclera or choroid characterized by histological deposition of calcium pyrophosphate. Taking into consideration its similar clinical manifestations with other diseases of the sclera, the most dangerous of which are malignant, timely verification of the diagnosis with the appointment of a further observation period is important.*
The aim. *The description of a clinical case of sclerochoroidal calcification to improve the efficiency of disease detection through the use of multimodal diagnostics.*
Material and methods. *A 62-year-old patient with complaints of "bright flashes" in her left eye for the past few months, who underwent a standard complex of ophthalmological examinations, supplemented according to indications by optical coherence tomography of peripapillary nerve fibers, macular zone, B-scan, Dopplerography in color Doppler mapping mode. Auxiliary diagnostic methods were magnetic resonance imaging of the orbits and extraocular muscles, computed tomography of the orbits and a biochemical blood test.*
Results. *Considering the anamnesis, the absence of progression of complaints, the data of instrumental diagnostic methods, the absence of pathological blood flow in the area of both eyes formations, the correct diagnosis is most likely to be sclerochoroidal calcification of both eyes, despite the difficulties of the diagnostic process, which consisted in the absence of visualization of foci during ophthalmoscopy.*
Conclusion. *Sclerochoroidal calcification is of interest to practicing ophthalmologists due to the difficulties of diagnostic search and differential diagnosis with malignant neoplasms. Modern medicine has a sufficient set of instrumental and laboratory research methods for making an accurate diagnosis.*

Key words: *sclerochoroidal calcification, mineral metabolism, computed tomography*

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КЛИНИЧЕСКИЙ СЛУЧАЙ СКЛЕРОХОРИОИДАЛЬНОЙ КАЛЬЦИФИКАЦИИ

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РЕЗЮМЕ

Обоснование. *Склерохориоидальная кальцификация – это идиопатическое редкое доброкачественное образование склеры или сосудистой оболочки, характеризующееся гистологическим отложением пирофосфата кальция. Учитывая схожие клинические проявления с другими заболеваниями склеры, большую опасность из которых представляют злокачественные, имеет значение своевременная верификация диагноза с назначением дальнейшего периода наблюдения.*

Цель. *Описание клинического случая склерохориоидальной кальцификации для повышения эффективности выявления заболевания путём применения мультимодальной диагностики.*

Материал и методы. *Пациентка 62 лет с жалобами на «яркие вспышки» перед левым глазом последние несколько месяцев, которой был проведён стандартный комплекс офтальмологического обследования, дополненный по показаниям оптической когерентной томографией перипапиллярных нервных волокон, макулярной зоны, В-сканированием, допплерографией в режиме цветного доплеровского картирования. Вспомогательными методами диагностики являлись магнитно-резонансная томография орбит и экстраокулярных мышц, компьютерная томография орбит и биохимический анализ крови.*

Результаты. *Учитывая анамнез, отсутствие прогрессии жалоб, данные инструментальных методов диагностики, отсутствие патологического кровотока в области образований обоих глаз правомерным диагнозом, вероятнее всего, будет склерохориоидальная кальцификация глаз, несмотря на трудности диагностического процесса, которые заключались в отсутствии визуализации очагов при офтальмоскопии.*

Выводы. *Склерохориоидальная кальцификация представляет интерес для практикующих офтальмологов ввиду трудностей диагностического поиска и дифференциальной диагностики со злокачественными новообразованиями. Современная медицина располагает достаточным набором инструментальных и лабораторных методов исследования для постановки точного диагноза.*

Ключевые слова: *склерохориоидальная кальцификация, минеральный обмен, компьютерная томография*

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OBJECTIVES

Sclerochoroidal calcification (SCC) is an idiopathic rare benign lesion of the sclera or choroid characterized by histological deposition of calcium pyrophosphate. Morphologically, SCC is visualized as whitish-yellow foci in the thickness of the sclera, usually located at the medial periphery in the superior temporal and superior nasal quadrants, with a symmetrical location in both eyes. Often, diagnostic difficulties lie in the similar clinical manifestations of diseases such as choroidal osteoma, choroidal nevus, metastatic choroidal lesion, choroidal hemangioma, and retinal astrocytoma [1–4]. According to a study by C.L. Shields, the average age at the time of diagnosis was 69 years. It was most common in Caucasian women, with unilateral and bilateral eye lesions occurring in almost equal percentages of cases (48 and 52 % respectively). Patients usually had no complaints, and the detected calcium deposition did not lead to a decrease or loss of vision acuity, changes in the size of the focus, decalcification and associated subretinal fluid, and neovascularization for 4 years of follow-up [1, 3–6]. According to the results of foreign studies, optical coherence tomography (OCT) with an extended depth imaging module showed that the lesion foci are located in the sclera rather than being of scleral and choroidal origin, as previously believed [7–9]. Among Russian scientists, A.S. Stoyukhina confirms the fact of scleral origin of SCC in the course of studies on high-resolution OCT data with the study of deep tissues using the scan averaging function [2, 10]. In rare unique cases described by foreign scientists, SCC is accompanied by neovascular membrane formation with subsequent need for treatment with angiogenesis inhibitors [11–13]. A case of SCC with detected metabolic disorders in a 70-year-old patient with chronic renal failure and a follow-up period of 7 months is described. The results of the follow-up showed no growth of visualized foci [14].

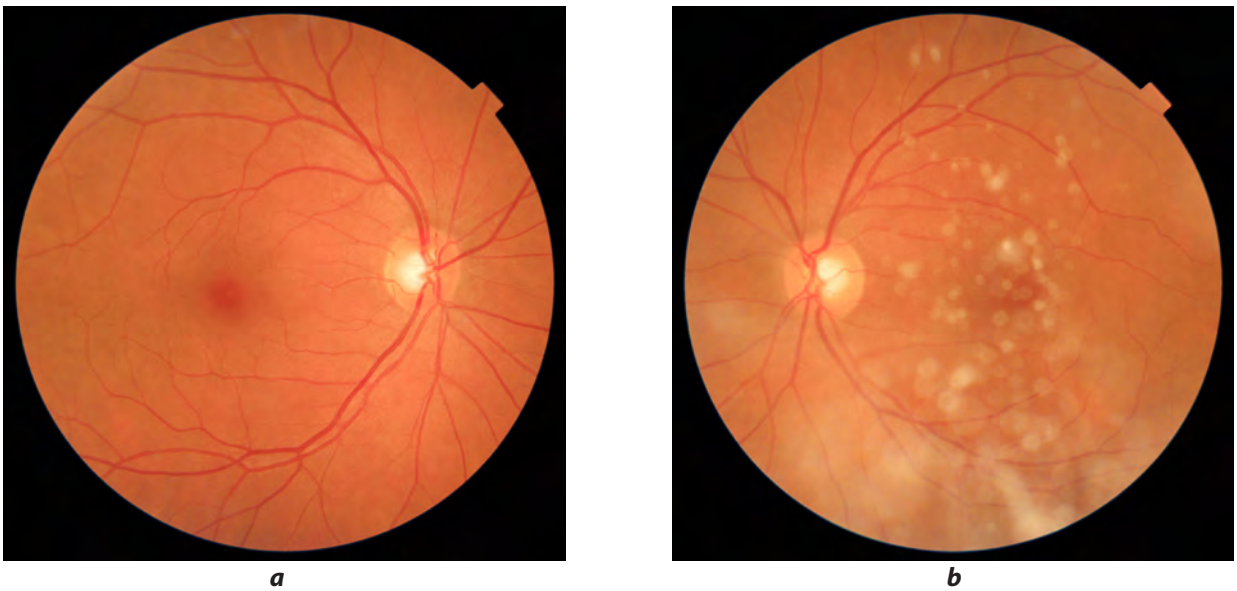
THE AIM

Description of a case report of SCC to improve the detection of sclerochoroidal calcification by using multimodal diagnosis.

MATERIALS AND METHODS

A 62-year-old female patient came to the Orenburg Branch of the Eye Microsurgery Institution in October 2021 with complaints of "bright flashes" in her left eye that had been bothering her for several months.

Medical history: she was registered with iris nevus of the left eye at the place of residence. At her follow-up examination in April 2021, the patient underwent ultrasound of the orbit and duplex ultrasound of the ophthalmic artery territory using the GE Logiq e machine. Along the posterior contour of the left eyeball at the edge of the optic nerve a focal choroideal lesion was detected with a length of 4.2 mm and a prominence of 2.1 mm of hyperechogenic homogeneous structure, with clear and irregular contours. Within the focal area there were single arterial and venous vessels with linear blood flow velocity (LBFV) in arteries 3.1 cm/s, RI – 0.50, in veins – up to 8.3 cm/s. The patient underwent the control examination in October 2021. The results with regard to the foci in the left eye showed no negative dynamics. When performing duplex ultrasound of the right eye in the right orbit, a focus of similar characteristics was found, 4.6 × 1.5 mm in size, with LBFV in arteries up to 16.1 cm/s, in veins – up to 7.9 cm/s. The ophthalmic artery (OA) hemodynamics was asymmetric, with some hemodynamic predominance in the central retinal artery (CRA) in the right orbit, in OA – in the left orbit – sufficient with markedly increased tone.

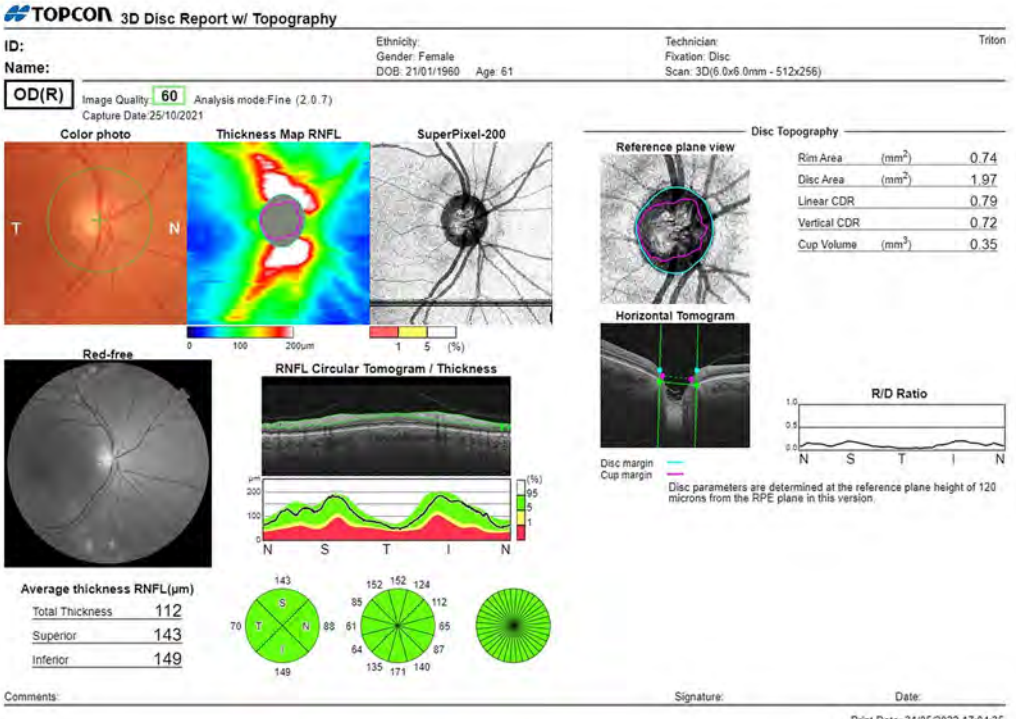


a ***b***

FIG. 1.
Ophthalmoscopic picture of the fundus: ***a*** *– right eye;* ***b*** *– left eye*

When the patient came to the Orenburg Branch of the Eye Microsurgery Institution, the visual acuity of the right eye was 0.8 with hyperopic complex correction of best corrected visual acuity (BCVA) to 1.0, and the left eye was 0.7 with hyperopic complex correction to 1.0.

Ophthalmoscopy: right eye without features, left eye – flat iris nevus at 8 o'clock position, pseudoexfoliative syndrome, vitreous degeneration by the type of asteroid hyalopathy (Fig. 1). Examination with the Goldmann three-mirror lens in the right eye from 7 to 9 o'clock postitions, retinoschisis with the necessity for retinal laser photocoagulation was detected in the extreme periphery; the extreme periphery of the left eye was unremarkable.



a

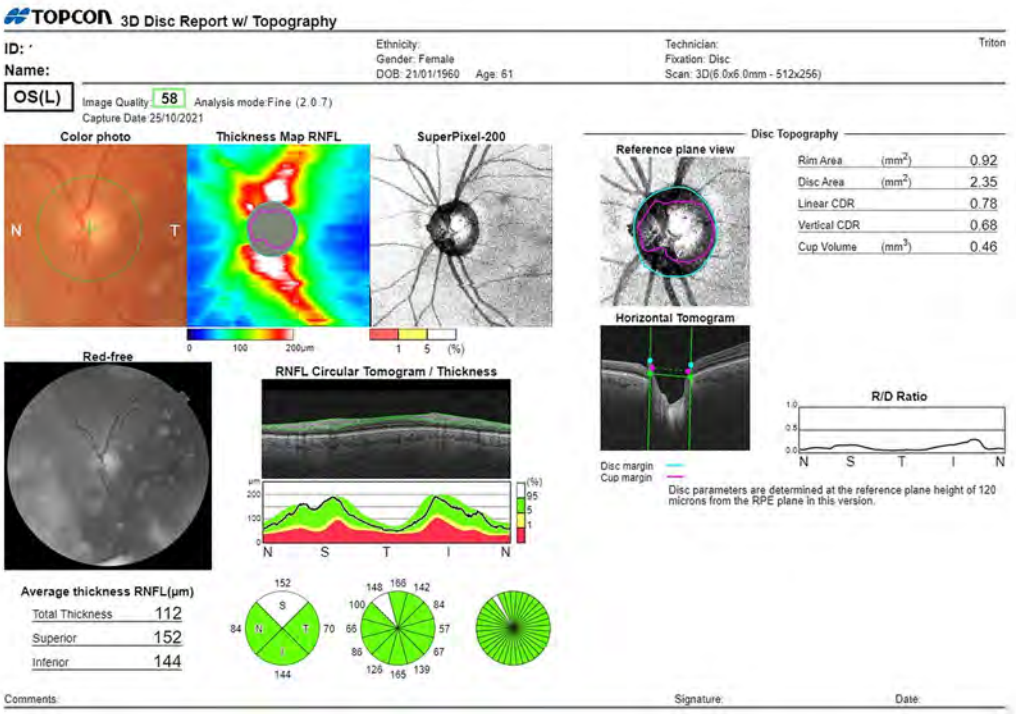


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FIG. 2.
OCT of peripapillary nerve fibers: ***a*** *– right eye;* ***b*** *– left eye*

The patient underwent OCT of peripapillary nerve fibres (Topcon DRI OCT Triton), macular zone (Spectralis HRA + OCT (Heidelberg), B-scan (Accutome B-scan Plus) and Doppler ultrasound (GE Logiq e) in colour Doppler imaging (CDI) mode.

According to OCT data of peripapillary nerve fibres, the thickness of peripapillary nerve fibres of the right eye is within normal limits (Fig. 2a), thickened in the upper segment of the left eye (Fig. 2b).

According to OCT data of the macular zone of both eyes, the retinal thickness is within normal limits, the pigment epithelium is preserved (Fig. 3).

The B-scan of the right eye (Fig. 4a) revealed single threads of low echogenicity in the vitreous body. A high echogenicity inclusion with Hmax = 0.81 mm, 2.17 mm in length is scanned in the middle periphery in the membrane thickness at 12 o'clock meridian. According to the B-scan of the left eye (Fig. 4b), there are many clumps of high echoicity in the vitreous body (the "golden rain" type of degeneration). At 12–1:30 meridian in the membrane thickness at the middle periphery a high echoicity inclusion giving a shadow with Hmax = 0.92 mm, extent 2.33 mm, is scanned.

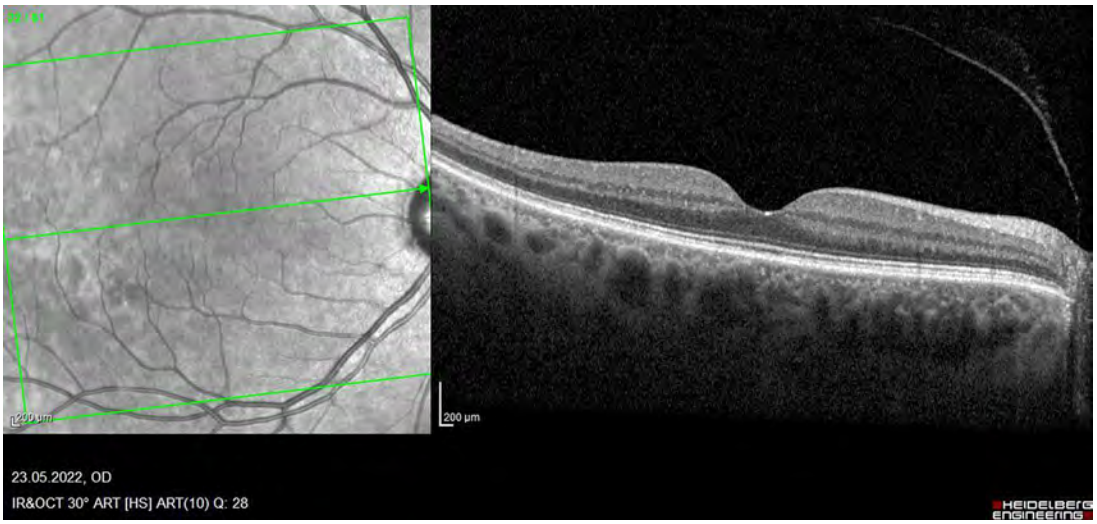
According to the Doppler ultrasound of both eyes (Fig. 5), a high echogenicity lesion with acoustic shadow (3.9 × 1.9 mm – right eye, 3.9 × 1.1 mm –left eye) with no signs of blood flow in the CDI mode was scanned in the upper anterior segment in the membrane thickness (Fig. 6).

At the follow-up examination in January 2022 (4 months later), the size of the lesion in the right eye had increased to 4.3 × 2.1 mm, in the left eye – no negative dynamics, still without signs of blood flow.

The patient was recommended to consult an endocrinologist and a biochemical blood test was scheduled to search for mineral metabolism disorders (Table 1).

According to the examination results, no mineral metabolism disorders were found, and no endocrinological pathology was detected.

In January 2022, the patient underwent focal retinal laser photocoagulation of the retinoschisis zone at 7–9 o'clock of the right eye at the Orenburg Branch of the Eye Micro-



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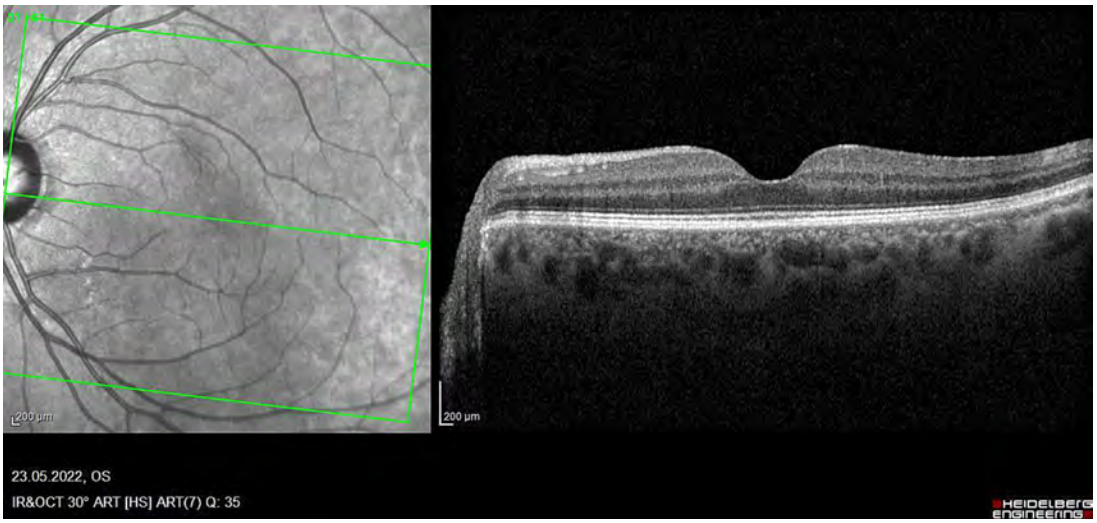

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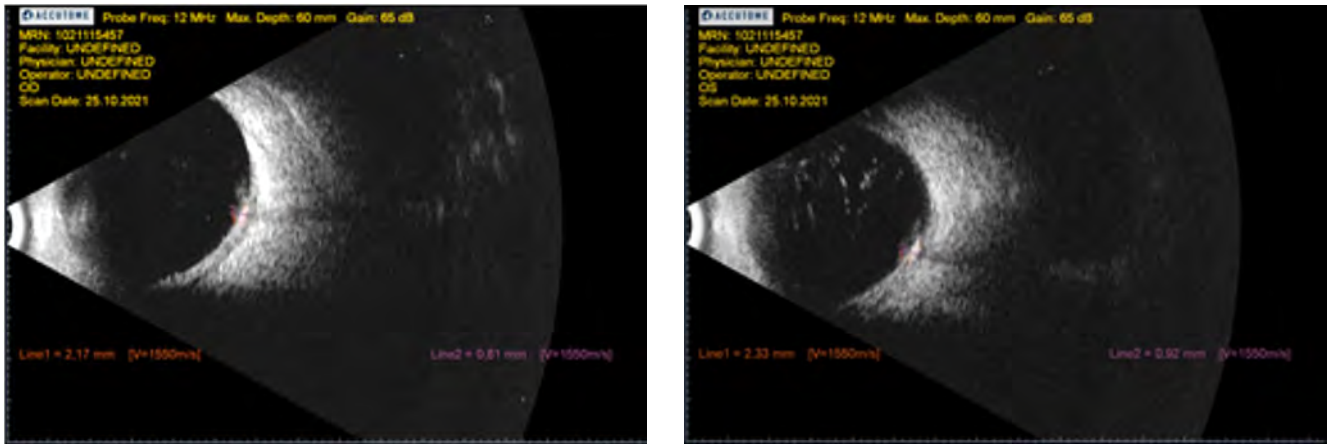
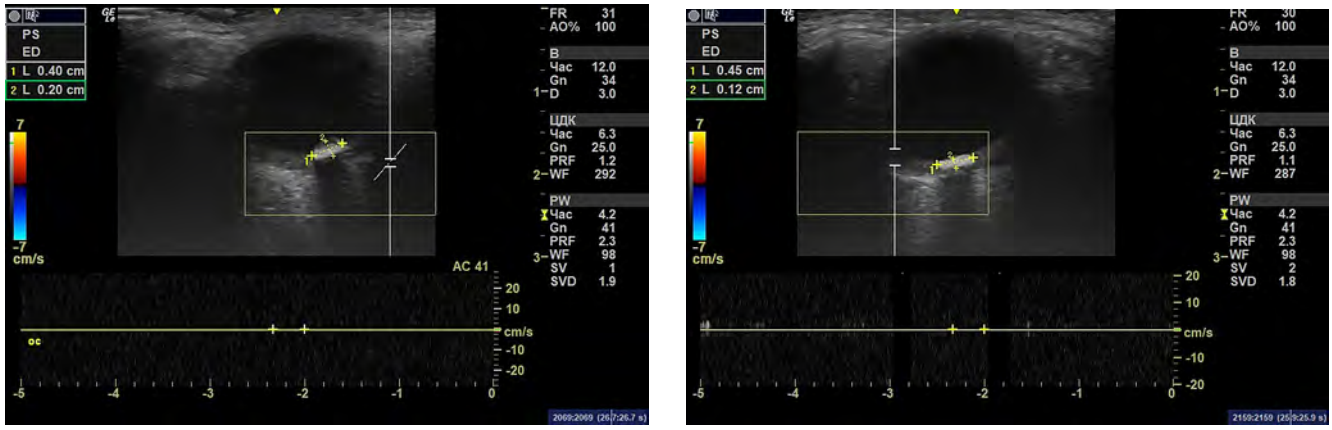
FIG. 3.
*OCT of the macula: **а** – right eye; **б** – left eye*



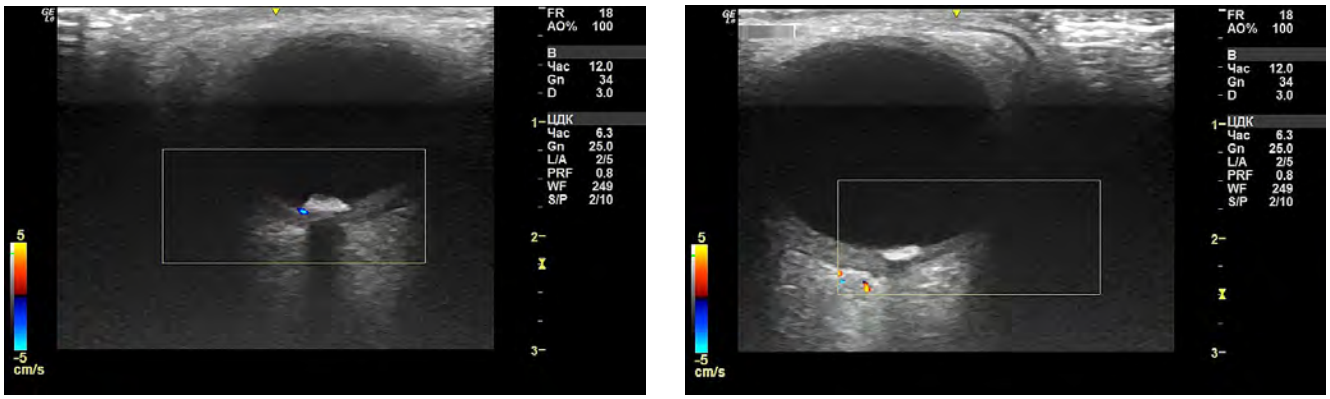
a ***b***

FIG. 4.
B-scan: ***a*** *– right eye;* ***б*** *– left eye*



a ***b***

FIG. 5.
Dopplerography: ***a*** *– right eye;* ***б*** *– left eye*



a ***b***

FIG. 6.
Dopplerography in the color duplex mapping mode: ***a*** *– right eye;* ***б*** *– left eye*

surgery Institution. She was also recommended to undergo computed tomography (CT) of the orbits at her place of residence.

The patient returned for a follow-up examination in May 2022. She presented no complaints. At the time of examination, the BCVA of the right eye was 1.0, the BCVA of the left eye was 0.7 with previous correction to 1.0. Due to the impossibility to perform the prescribed CT examination at her place of residence, the patient provided magnetic resonance imaging (MRI) data of the orbits and extraocular muscles on a Siemens Magnetom Essenza 1.5 T in T1 Vibe Fs Ttra mode with a slice thickness of 1 mm.

TABLE 1

THE RESULTS OF A BIOCHEMICAL ANALYSIS OF THE BLOOD OF THE EXAMINED PATIENT, INDICATING THE REFERENCE VALUES

Study	Result	Unit	Reference values
Calcium	2.37	mmol/L	2.20–2.55
Potassium	4.1	mmol/L	3.5–5.1
Sodium	141	mmol/L	136–145
Chlorine	106	mmol/L	101–110
Magnesium	0.88	mmol/L	0.66–1.07
Organic phosphorus	1.16	mmol/L	0.74–1.52
Parathyroid hormone	6.8	pmol/L	1.45–10.41
Glomerular filtration CKD-EPI Creatinine	95	ml/min/1.7 m^2	> 60
25-OH vitamin D	32	ng/ml	< 10 ng/ml – severe deficiency; < 20 ng/ml – deficiency; 20–30 ng/ml – insufficiency; 30–100 ng/ml – adequate level; > 150 ng/ml – possible toxic effect

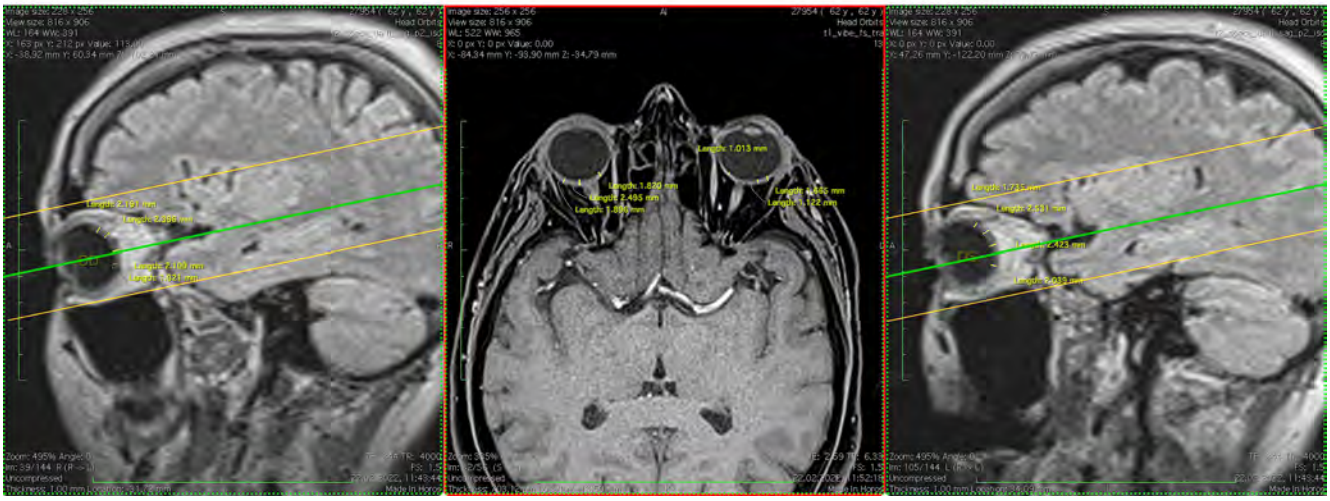


FIG. 7.

MRI of thickening of the sclera

MRI revealed MR signs of uneven thickness of the sclera of both eyeballs, inhomogeneous structure (Fig. 7) with point-like inclusions (Fig. 8).

As a final method of examination for the most accurate visualization of calcification foci in the sclera, CT of the orbits on a Philips Mx-16 device in bone and brain modes with a slice thickness of 1 mm in sagittal, frontal and vertical planes was chosen.

According to the CT findings, almost symmetrical areas of calcification located at 11 o'clock, at a distance of about 6 mm from the optic disc (OD) of the right eye are determined in the posterior hemispheres of the orbits and the sclera structure (Fig. 9) the size of the calcificates is 2.3 × 4 × 1.6 mm.

A focus measuring 3.8 × 3.7 × 1.5 mm was detected at a distance of 6.5 mm from the OD of the left eye during 12 o'clock CT in the posterior hemispheres of the orbits of the left eye (Fig. 10).

These calcificates have a maximum density of about +200 HU, repeat the course of the sclera, without bulging beyond its outer contour, with minimal bulging towards the chorioidea on both sides, their contours are clear, slightly irregular on the left side. A questionable, but not excluded, forming small clumpy calcificate about 1 mm in diameter was found at the border of the anterior and posterior OS hemispheres at 3 o'clock. There were no signs of other areas of pathological density in the sclerae.

On the next Doppler ultrasound in May 2022, the size of the lesion in the right eye was negative, and in the left eye it was 4.3 × 5 mm with no signs of blood flow.

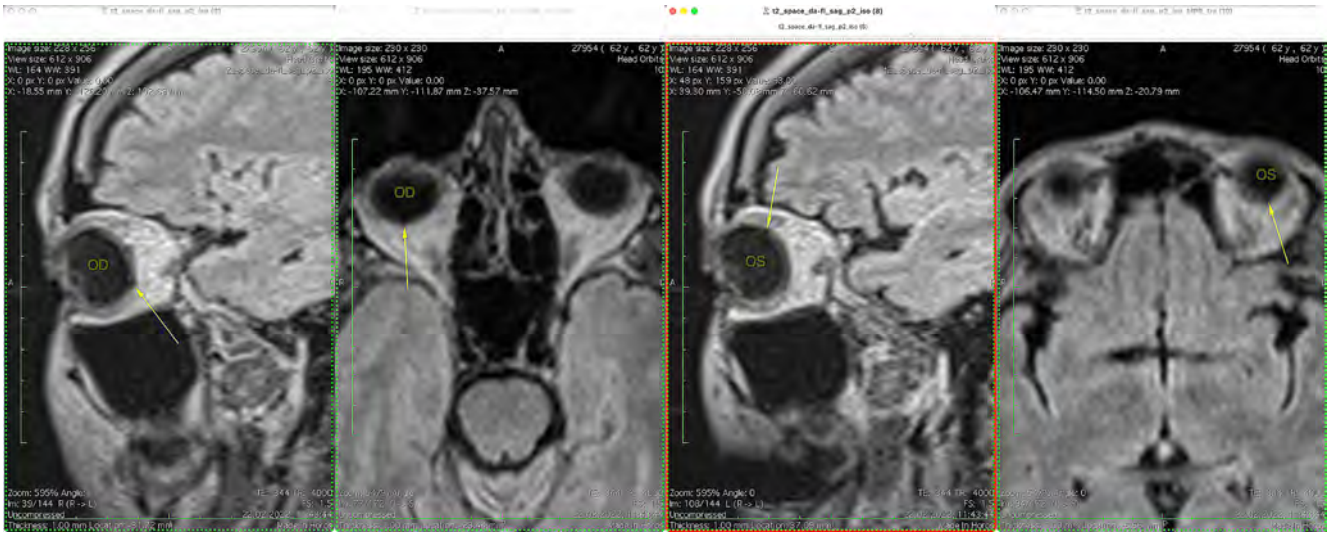


FIG. 8.
MRI of inclusions suspicious of calcifications

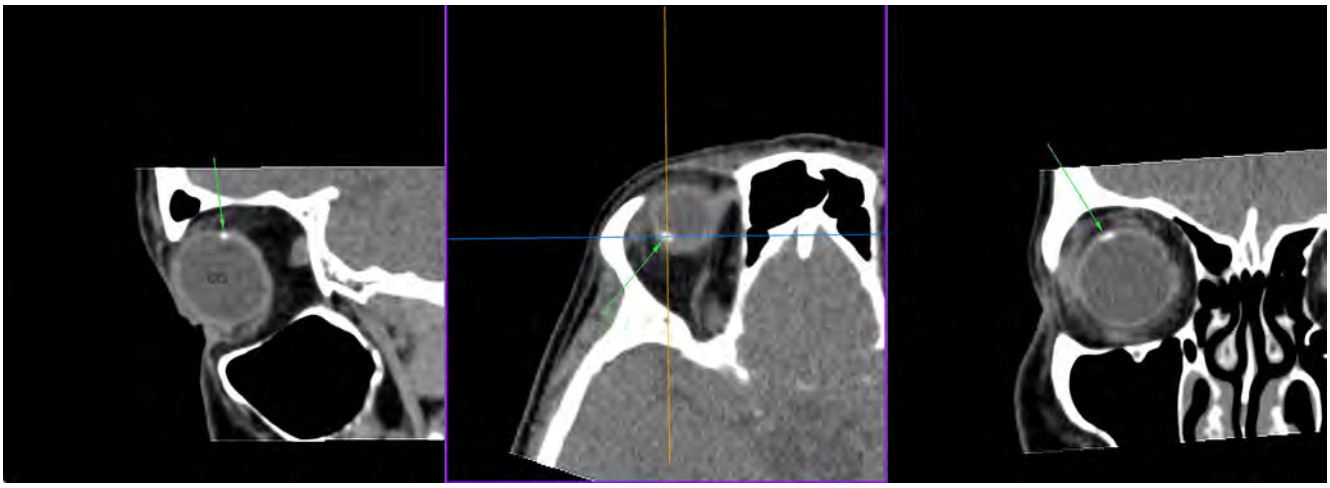


FIG. 9.
CT of calcifications in the structure of the sclera of the right eye

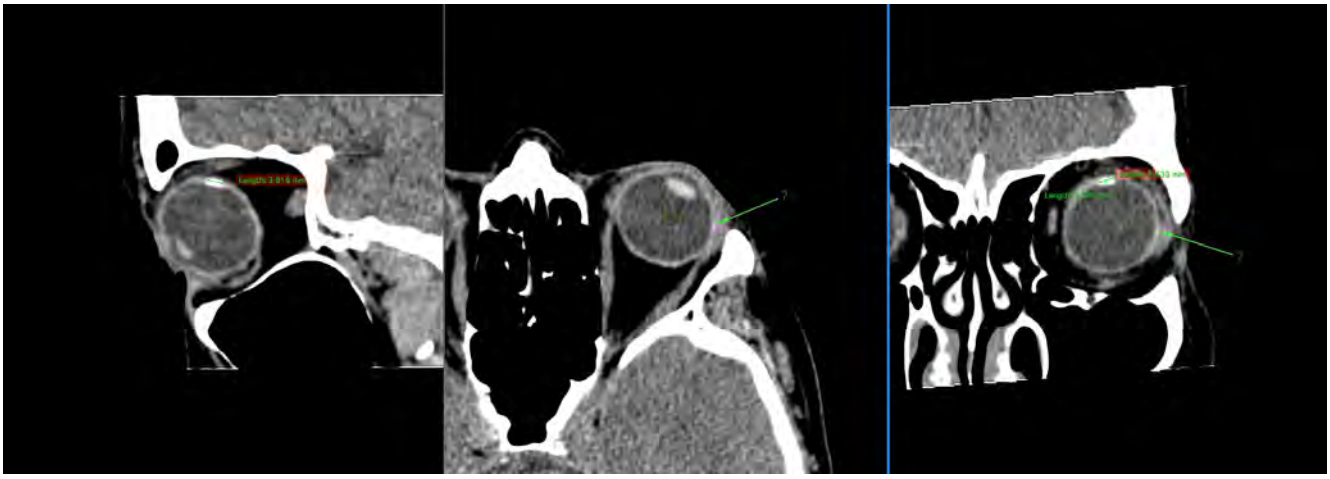


FIG. 10.
CT of calcifications in the structure of the sclera of the left eye

RESULTS AND DISCUSSION

Considering the anamnesis, the absence of progression of complaints, the data of instrumental diagnostic methods, the absence of pathological blood flow in lesion areas of both eyes according to Doppler ultrasound in CDI mode, the correct diagnosis is most likely to be sclerochoroidal calcification of both eyes, despite the difficulties of the diagnostic process, which consisted in the absence of visualization of foci during ophthalmoscopy. Taking into account the follow-up period of 9 months and the growth of the lesions by 0.4 × 0.2 mm and 0.4 × 3.9 mm according to Doppler ultrasound, the patient was recommended dynamic follow-up in 3 and 6 months with control biochemical blood test. Thanks to the above-described algorithms of diagnostic search of Russian and foreign colleagues, today the diagnosis of SCC for ophthalmologists together with CT-diagnosticians is a feasible task, which is confirmed by the result of the work done.

CONCLUSION

SCC is an idiopathic benign disease of the sclera that is difficult to diagnose. Diagnosis is made on the basis of patient's medical history, instrumental and laboratory tests. Given the uniqueness of the disease, as well as the difficulties in diagnosis due to its similarity to other malignant diseases of the sclera and vasculature, the correct diagnosis, determination of management methods, prescription of appropriate treatment and establishment of a follow-up period are of great importance. Modern medicine has a sufficient set of instrumental and laboratory methods of examination to make an accurate diagnosis of patients with SCC.

Conflict of interest

The authors of this article declare the absence of a conflict of interest.

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