IDIOPATHIC SCLEROCHOROIDAL CALCIFICATIONS. A CASE REPORT

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SUMMARY

Aim: Sclerochoroidal calcifications (SCHC) are an uncommon benign ocular condition that occurs in elderly patients. SCHC usually manifest as multiple placoid yellow lesions in the midperipheral fundus, most often in the upper temporal quadrant.
They are asymptomatic and often discovered during routine eye examinations in a patient with good visual acuity and visual field. According to the etiology, SCHC are divided into idiopathic, metastatic and dystrophic.
Case reports: This is case report of 2 patients with idiopathic SCHC, who underwent basic eye examinations, fundus photography, optical coherence tomography, ultrasonography, fluorescein angiography, fundusautofluorescence, laboratory screening and in the second case also CT head scan.
Conclusion: The aim of this publication is to point out the typical features of SCHC and their distinction from more serious conditions that they may resemble.
Key words: sclera, choroid, calcification, calcium, osteoma

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INTRODUCTION

Sclerochoroidal calcifications (SCHC) are a rare benign ocular condition that occurs in elderly, fair-skin patients [1,2,3]. Yellow to yellow-white mono or multifocal lesions are visible on the fundus in the mid-periphery. They are placoid, flat or minimally elevated, rare tumor-like lesions with an elevation of up to 6 millimeters [2,3]. The lesions occur mostly bilaterally in all quadrants, but are most often found in the superotemporal quadrant [1,2,3].

Their borders may be sharply defined, with an irregular map-like or yellowish halo edge. The pigmentation of the calcification varies; sometimes with mild hyper or hypo-pigmented changes, even atrophy of the retinal pigment epithelium (RPE) in the SCHC area [2].

They are asymptomatic and can be easily overlooked or misinterpreted. At first look, they may imitate choroidal amelanotic melanoma, osteoma, choroidal metastasis, lymphoma or chorioretinitis [1].

Histopathologically, these are calcium pyrophosphate deposits in the sclera [1]. Based on the etiology, calcifications can be divided into primary - idiopathic, where the cause is unknown. They form in the healthy tissue of patients with no metabolic abnormalities. In addition, calcifications can be dystrophic or metastatic, according to the known cause of formation. Dystrophic calcifications have normal calcium and phosphate metabolism, but deposits are formed in damaged ocular tissues, for example caused by chronic intraocular inflammation, severe ocular trauma or chronic scleritis.

Metastatic precipitates occur when there are systemic disorders of calcium and phosphate metabolism due to calcium salt deposited in otherwise healthy tissue, such as in primary and secondary hyperparathyroidism, pseudohypoparathyroidism, vitamin D intoxication, sarcoidosis, hypophosphatemia, chronic renal failure, Bartter or Gitelman syndrome. [1,4,5,6]. After excluding dystrophic and metastatic etiology, we can diagnose idiopathic SCHC. It is important for an ophthalmologist to correctly recognize them to mitigate the risk of systemic disease or improper intervention. Lim and Goldberg describe the case of a 70-year-old patient diagnosed with choroidal metastases. The patient was treated with local radiotherapy for two years with no effect. After some time, the diagnosis was changed to idiopathic SCHC [7].

The exact pathophysiology of SCHC is unclear [3]. Examinations such as ultrasonography (USG), optical

coherence tomography (OCT), computed tomography (CT) and fluorescein angiography (FAG) help differentiate SCHC from other conditions. Laboratory examinations have categorized calcifications by their etiology as idiopathic, dystrophic and metastatic [3].

In this publication I describe case reports of two patients with idiopathic SCHC who underwent basic eye examinations, fundus photography, OCT, USG, FAG, fundusautofluorescence (FAF), head X-rays, laboratory screening and in the second case also CT brain scan. The typical features of SCHC are pointed out in this publication, including how to distinguish them from more severe conditions they may resemble.

CASE REPORT 1

A 63-year-old male patient was referred to our department in June 2019 for a branch retinal vein occlusion (BRVO) in the right eye. His personal medical history showed him to have been treated for arterial hypertension, type 2 diabetes mellitus and hyperlipidemia. He had experienced impaired vision in his right eye for a month. The best corrected distance visual acuity was 0.5 in the right eye and 1.0 in the left eye. Intraocular pressure and anterior segment were normal. A defined optic nerve disc, irregular blood vessels, Gunn sign presents and lesion of intraretinal hemorrhages and retinal leakage in the superotemporal arcade were observed on the right eye fundus. In addition, numerous small yellow circumscribed inactive lesions of about 1-1.5 pupillary diameter (PD) in size were present in the mid-periphery of the superotemporal and nasal quadrants and prominent slightly above the level (Figure 1). The same lesions were also found in the mid-periphery of the left eye. The calcifications did not encroach upon the macular region and were not accompanied by exudation or vitreous reaction. The right-eye OCT examination showed a cystoid macular edema with central retinal thickness of 486 micrometers. The patient was indicated for anti-VEGF treatment with ranibizumab. The ultrasonography B-scan bilaterally showed numerous small placoid hyperechogenicities at the level of the retina (Figure 2). FAG showed hyperfluorescence to be present from the initial stages at the SCHC region, with no leakage (Figure 3). FAF showed hyperfluorescence in the SCHC region near the superotemporal arcade, with spots after laserphotocoagulation visible in the right eye (Figure 4). The head X-ray found no remarkable findings. The patient underwent laboratory screening to rule out a disorder of calcium and phosphate metabolism and the results were in a normal scale. We diagnosed idiopathic SCHK bilaterally and BRVO in the right eye. The patient currently received seven injections of ranibizumab and focal laserphotocoagulation of the right eye with an excellent effect. The fundus finding is stable, the patient is not being treated and has BCVA 1.0 bilaterally. Over the two-year follow-up, there was no increase in the number or size of lesions.

CASE REPORT 2

A 68-year-old female patient was referred to our clinic because of a choroidal tumor in her right amblyopic eye. The patient had no acute subjective ocular problems. Her personal medical history showed that she had been treated only for arterial hypertension. Family medical history for calcium metabolism disorder was negative. The best corrected distance visual acuity was 0.7 in the right eye and 0.1 in the left. A corticonuclear cataract was bilaterally present. No remarkable findings from intraocular pressure and the rest of the anterior segment were documented. On the right eye fundus there was a yellow-white inactive, well-defined lesions at the inferotemporal arcade about 3 PD and two small indistinct

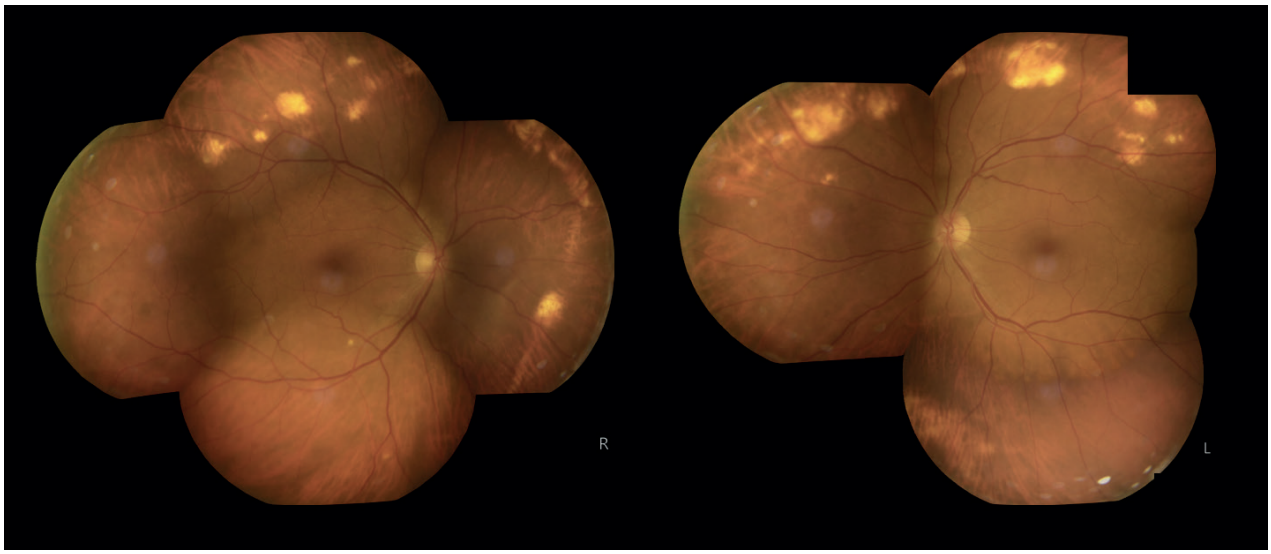

Figure 1. Fundus photography (right eye after antiVEGF therapy) – in middle periphery upper temporal and nasal quadrant are sclerochoroidal calcifications

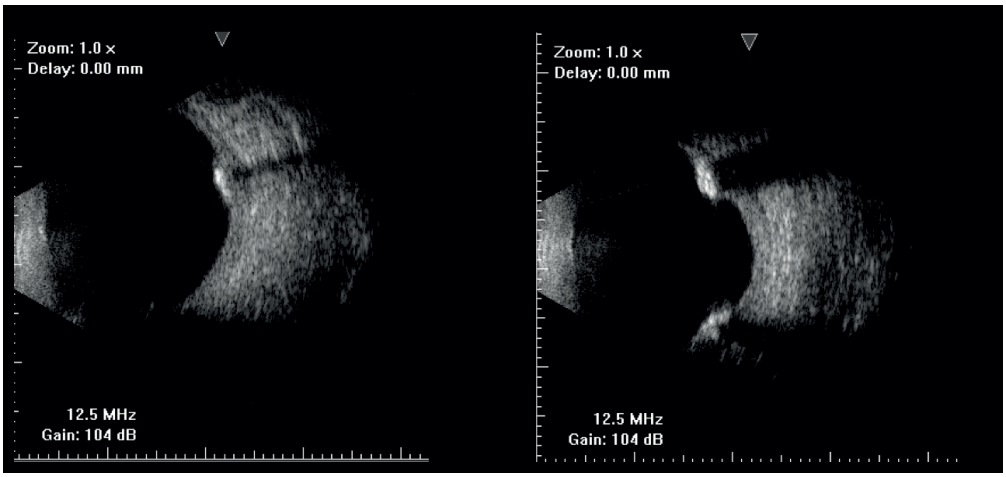

Figure 2. USG B-scan – bilateral numerous small placoid hyperechogenities are seen in the level of retina, which promotes slightly above

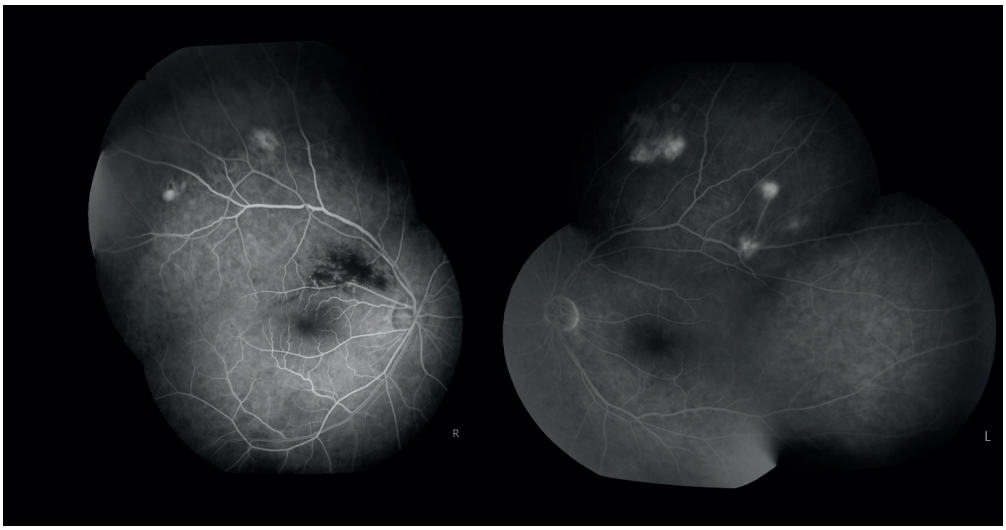

Figure 3. Fluorescein angiography – hyperfluorescence in the place of calcification from initial stages and the blockade of fluorescence due to venous occlusion

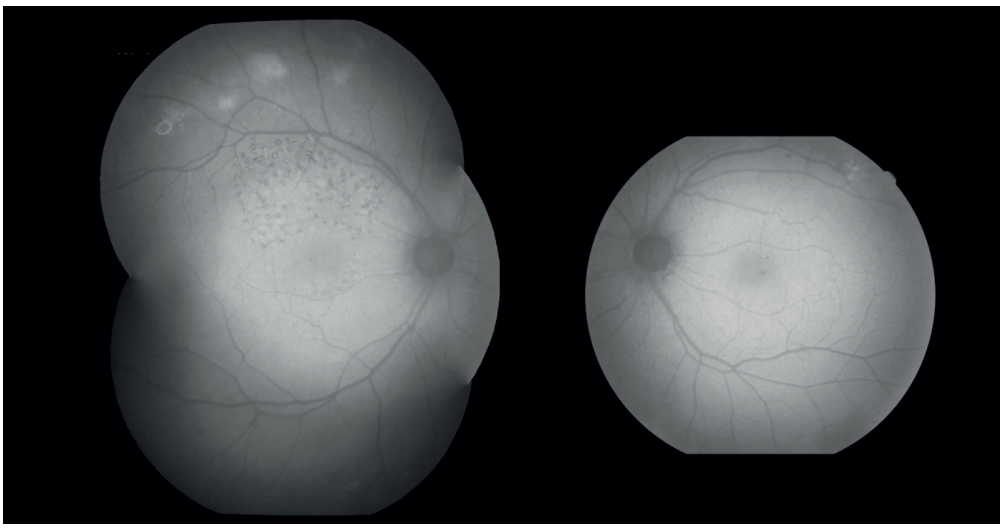

Figure 4. Fundusautofluorecsence – hyperfluorescence in the place of calcifications, in the right eye there are spots after focal laserphotocoagulation

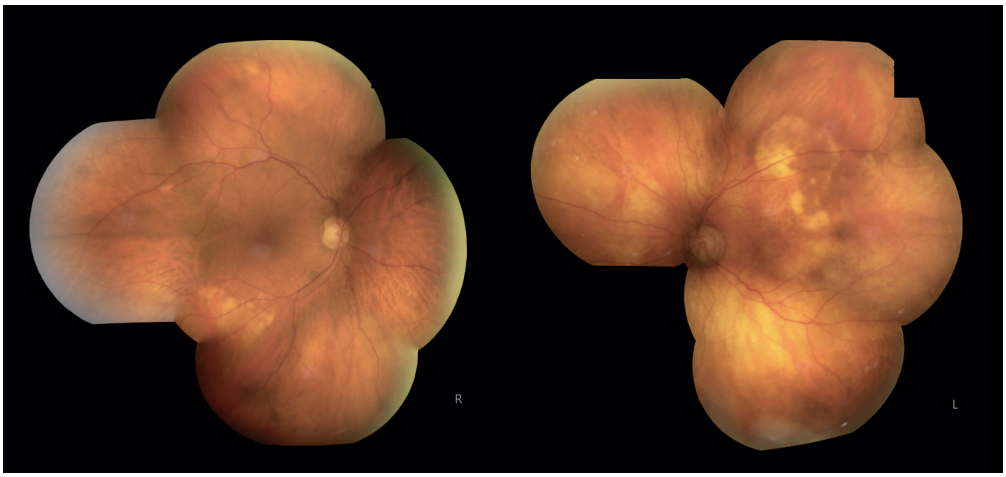

Figure 5. Fundus photography – yellow irregular inactive lesions are seen bilaterally, it the left eye lesion partially reach the macula

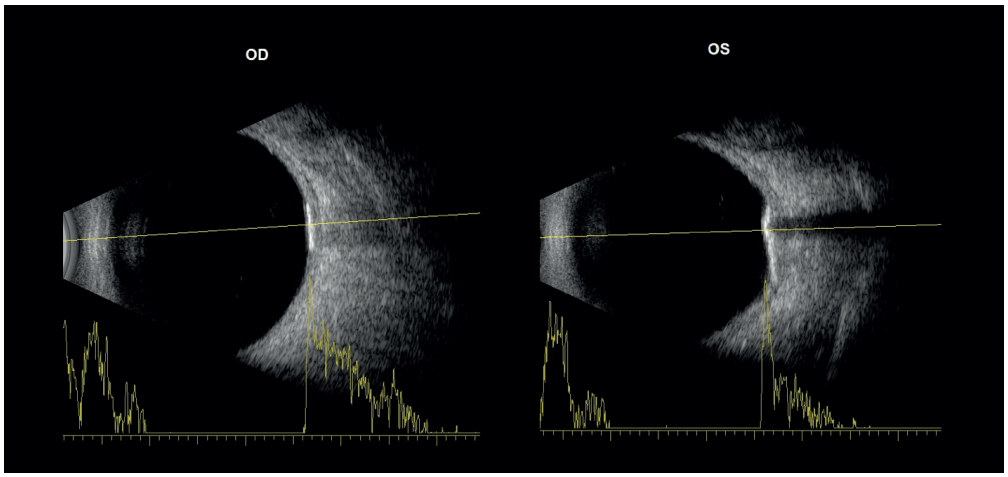

Figure 6. USG B-scan – hyperechogenic plaques at the level of retina with orbital shadowing, A scan – spike at the lesion level and a decrease amplitude behind the lesion

yellowish lesions of about ½ PD at the superotemporal arcade. The left eye fundus showed irregular yellowish confluent lesions at the superotemporal arcade, measuring 1-2 PD, that partially extend the macular region (Figure 5). An ultrasonography examination of both eyes showed hyperechogenic placoid lesions at the level of the retina with orbital shadowing (Figure 6). The A-scan was dominated by a spike at lesion level and an amplitude drop behind the lesion.

An OCT macular scan of right eye showed vitreomacular traction syndrome and a macular hole in the left eye. The lesions in the left eye partially encroaching upon the macula made it possible for OCT to be performed through the lesion (Figure 7). Choroidal elevation was present in the lesion area with normal retinal thickness. There were early to late stages of hyperfluorescence imaged in the FAG at the region of the lesions, with no later leakage (Figure 8).

The brain CT scan found bilateral intraocular calcifications at the level of the sclera and hyperostosis of the frontal and parietal bone with suspected hyperparathyroidism (Figure 9). All laboratory results were on standard scale. Calcium and phosphorus metabolism disorders were excluded and the diagnosis by us of idiopathic SCHC came from comprehensive ocular and laboratory testing.

This has been the only described case so far of SCHC affecting the macula with a macular hole present. No surgical therapy of the macular hole was recommended because of the amblyopia and the absence of subjective ocular discomfort. Cataract surgery has been planned for the patient and she stays with us for follow-up ocular fundus imaging and an ultrasonography examination.

DISCUSSION

Goldstein and Miller first described in 1982 the yellowish deposits in a patient with hyperparathyroidism

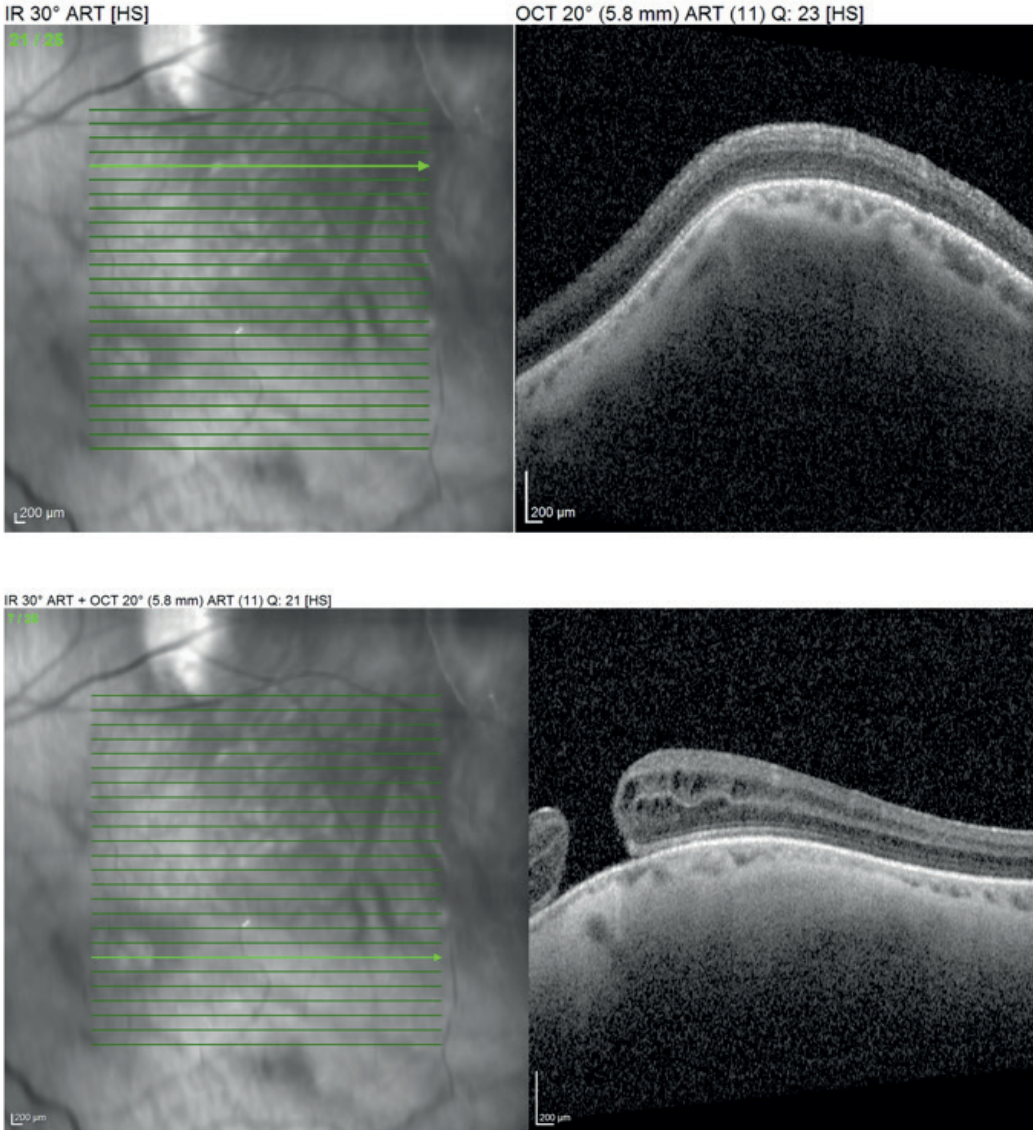

Figure 7. OCT B-scan of left eye – elevation of the choroid with normal retinal thickness in the place of the lesion (upper scan), macular hole (lower scan)

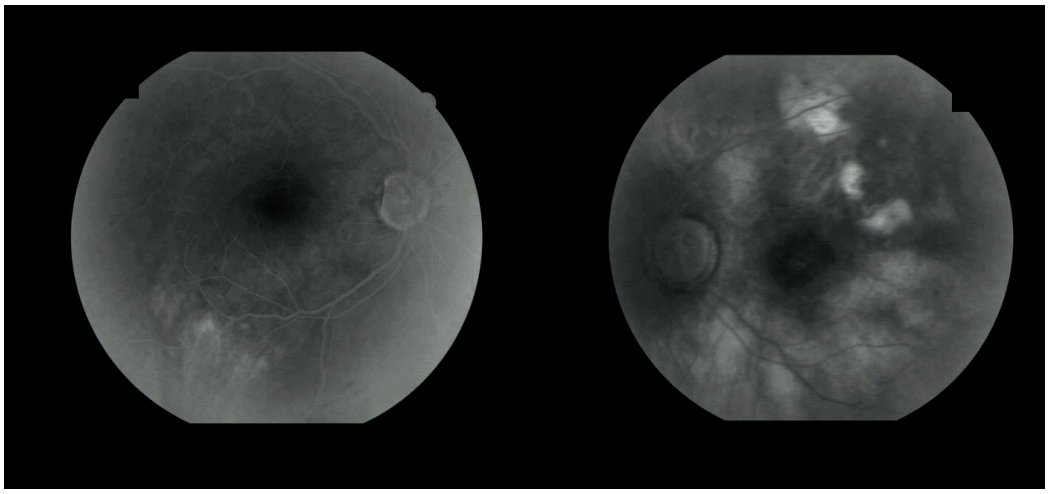

Figure 8. Fluorescein angiography shows hyperfluorescence during initial to late stage in place of calcifications

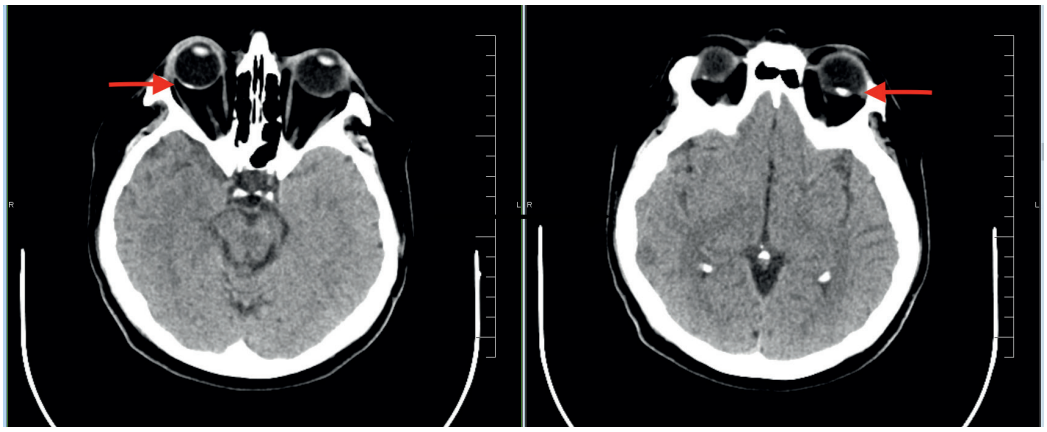

Figure 9. Brain CT scan – arrows indicate bilateral sclerochoroidal calcifications

as metastatic calcifications [8]. In 1991, idiopathic SCHC were first defined when seven patients were analyzed and found to have calcifications. They had had no abnormalities in calcium-phosphate metabolism and the calcifications were located in an otherwise healthy eye [9].

The exact pathogenesis of SCHC is unclear [3]. One theory is that the calcification is a result of a chronic traction on the sclera at the point of attachment to the oblique muscles. This is how Cogan senile scleral plaques are formed, they are grey areas of the sclera in front of the retraction of the horizontal rectus muscles. The plaque's characteristic grey color is due to hyaline degeneration of the sclera and the translucency of the uvea. These plaques calcify over time [2].

Shields et al. analyzed various parameters in a large group of 179 eyes among 118 patients with SCHC. The mean age of the patients when they were diagnosed was 69 years. 60% were female and 98% of the patients had fair skin. SCHC had unilaterally occurred in 48% of the cases and bilaterally in 52% of them.

The patients were primarily diagnosed with choroidal nevus (20%), choroidal melanoma (13%), choroidal lymphoma (10%), metastasis (5%) and idiopathic SCHC (5%), while choroidal osteoma was suspected in 3% of the patients and in 43% the diagnosis was unclear. SCHC were idiopathic in 79% and secondary in 21% of the patients. Secondary SCHC were most often caused due to hyperparathyroidism, parathyroid adenoma, Gitelman or Bartter syndrome. The OCT showed focal choroidal atrophy in 35% of the lesions and retinal pigment epithelium atrophy in 49% of them. There was no visual enlargement, decalcification, or vision decline from SCHC during the four-year follow-up. No patient ever developed subretinal fluid, hemorrhages or choroidal neovascular membranes (CNV). Cogan plaque was present in 14% of the patients [4].

Lesions remain stable and are not becoming enlarged [4]. However, a case of SCHC with a discovery of new lesions and enlargement of existing calcifications over 10 years has been described [10].

The most critical examinations for making a diagnosis are: OCT, USG, CT scans, FAF and FAG [1]. Despite the current nomenclature, enhanced depth imaging (EDI) an OCT revealed calcifications primarily arising from the sclera and merely putting pressure on the adjacent choroid [12]. Large lesions can stress not only the choroid but also the retina. In some cases, an OCT will show reduction of the outer nuclear layer, disruption of the outer limiting membrane, and RPE changes [11].

An ultrasonography B-scan typically images SCHC. They appear as solid lesions with high echo and posterior orbital shadow. The A-scan has a typical high spike around the lesion and low echo behind. A fundoscopy may sometimes not spot subclinical lesions and only ultrasonography will reveal them. An ultrasound is also useful for monitoring over time the size of the SCHC. They appear as hyperdense lesions at the level of the sclera that resemble bone in density on a CT scan. Hyperfluorescence appears at the site of SCHC and in staining at the later stages with no leakage on the FAG in the venous phase. An FAF will describe areas of hypo or hyperfluorescence, depending on the degree of RPE damage. The hypothesis is that calcification will chronically burden the choriocapillaris, causing RPE atrophy [12].

Although these calcifications are mostly idiopathic, they may be associated with abnormal calcium and phosphor metabolism. Laboratory testing of serum calcium, phosphate, parathyroid hormone, vitamin D, urea and creatinine is recommended to exclude systemic diseases.

SCHC can imitate a variety of diseases [2,3]. These lesions originate in the sclera, unlike choroidal osteoma, amelanotic nevus, melanoma, lymphoma, granuloma and metastasis. [11].

It is typical for SCHC to have no symptoms of exudation. Most choroidal malignant melanomas are pigmented but, at first glance, unpigmented amelanotic melanoma may resemble SCHC. Patients with SCHC are asymptomatic, whereas patients with melanoma develop recurrent macular

degeneration and, in the later stages, a decrease in vision caused by the development of serous neuroretinal ablation, exudative retinal detachment, or secondary glaucoma. In addition, malignant melanoma tends to be more elevated, has no distinct borders, has a lower ultrasound echo and is more vascularized on an FA. Choroidal osteoma is an ossified, solitary and juxtapapillary tumor that is benign, 75% unilateral and yellow-orange in color. It occurs mainly in young, healthy women aged 20-30 years with fair skin and is characterized by slow growth. Calcium and phosphorus metabolism disorders play no role in osteoma. Unlike SCHCs, exudative retinal detachment and CNV are complications. In ultrasonography, it has an even more pronounced echo comparable to the bone [12]. Breast and lung tumors most commonly metastasize to the choroid. Choroid metastases tend to be closer to the macular landscape and are accompanied by a neuroretinal ablation [2,3].

Although SCHC are mostly asymptomatic and require no treatment, follow-up is recommended. The prognosis is good as long as the lesions do not affect the macular area. There are a few articles describing vision loss from central macular deposits, CNV formation or peripheral scotomas, reported as complications of SCHC [3].

CONCLUSION

Idiopathic sclerochoroidal calcifications are a rare benign ocular condition. They typically occur in patients aged 50–80 years. The calcifications are asymptomatic and were discovered incidentally during a routine fundus examination in a patient with good visual acuity and visual field. These are precipitated calcium salts in the sclera with the oppression of the adjacent choroid. They appear mostly bilaterally, in the mid-periphery of the superotemporal quadrant [1,2,3]. Ultrasonography is most critical in the differential diagnosis, where highly echogenic plaques with orbital shadowing are seen [4]. There are no signs of exudation around the SCHC. Excluding dystrophic and metastatic etiology, idiopathic SCHC can be diagnosed. Idiopathic SCHC do not require any treatment. It is advisable to monitor these patients once a year and monitor the number and size of the lesions using fundus photography and ultrasonography. If the lesion is close to the macula, it is ideal to perform an OCT and Amsler grid is recommended because of the risk of CNV. In the case of metastatic and dystrophic SCHC, the primary cause must be treated [2].

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