



Bilateral Sclerochoroidal Calcifications: A Case Report

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Case Report

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Abstract

Purpose: The aim of this case report is to describe a rare condition of bilateral sclerochoroidal calcification

Case Report: A 79 years old asymptomatic Caucasian female was referred to our Ocular Oncology Center of the Ophthalmic Surgery Department of Pisa on November 2021. She was in apparent good health. Funduscopy examination revealed yellow-white irregular lesions in the supero-temporal quadrant of both eyes. Best-corrected visual acuity was 20/20. B-scan and A-scan ultrasound revealed hyper-echoic solid lesions with acoustic orbital shadowing measuring 1.5 mm in thickness and 4.6 mm in diameter, on average. Enhanced Deep Imaging Optical Coherence Tomography showed a scleral lesion compatible with the presence of deposition of calcium with Table Mountain and rolling pattern. Fluorescein angiography revealed hypo-fluorescence in the arterial phase and a mild hyper-fluorescence in the venous phase with persistent late staining. Indocyanine green angiography showed hypo-fluorescence at the early, middle, and late phases. Systemic investigation of calcium and phosphate metabolism was performed and the patient didn't display any underlying systemic disorder.

Conclusion: It is important to know and recognize this condition. Although most cases of sclerochoroidal calcifications may be idiopathic, systemic laboratory workup must be performed to exclude calcium-phosphate metabolic disorder that can be associated to these ocular signs. Given the rare possible development of choroidal neovascularization, regular imaging check-up is recommended.

Keywords: Retina; Sclerochoroidal Calcification; Sclera; Tumor

Introduction

Sclerochoroidal calcifications is a rare, benign condition characterized by yellow-white flat or minimally elevated lesions typically located in the supero-temporal post-equatorial area or along the temporal arcades [1]. It is often discovered in older white asymptomatic individuals during a routine fundus examination. There is a slight female predisposition (approximately 60% of cases) [2]. The lesions are commonly symmetric and bilateral [3]. This condition can

be idiopathic or associated to systemic calcium disorder [4]. Clinically, it can be mistaken for choroidal osteoma, choroidal metastasis, amelanotic choroidal nevus, choroidal lymphoma or choroiditis [5]. Fluorescein angiography, optical coherence tomography and ultrasonography may help in confirming the diagnosis. Clinical prognosis can vary depending on systemic associations. Visual prognosis is generally good even if in rare cases choroidal neovascularization (CNV) may occur [6,7].

Case Report

A 79 years old asymptomatic Caucasian female was referred to our Ocular Oncology Center of the Ophthalmic Surgery Department Pisa on November 2021. She was in apparent good health. Fundusoscopic examination revealed yellow-white irregular lesions in the supero-temporal quadrant of both eye (Figures 1 & 2).

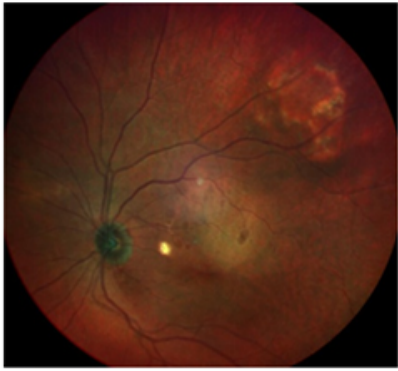


Figure 1: Fundus photographs of the right eye.

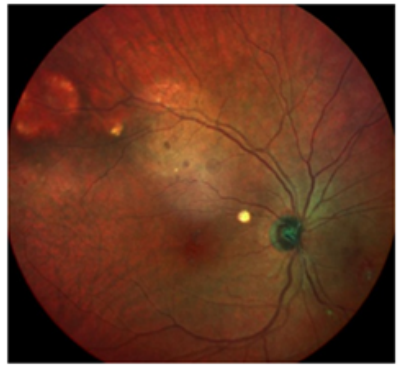


Figure 2: Fundus photographs of the left eye.

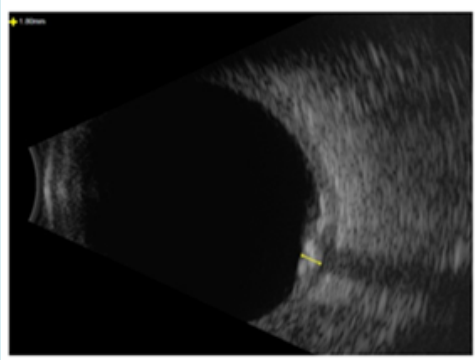


Figure 3: B-scan ultrasound.

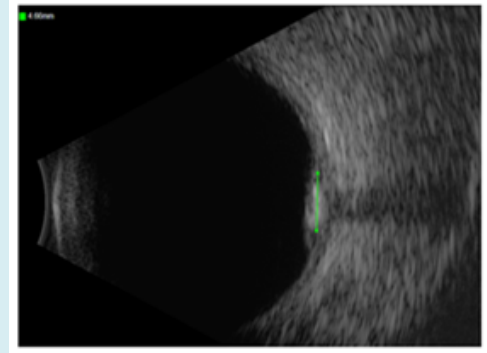


Figure 4: B-scan ultrasound.

Best-corrected visual acuity was 20/20. B-scan and A-scan Standardized Echography was performed with a 20 and a 10 MHz probe and revealed hyperechoic solid lesions with acoustic orbital shadowing measuring 1.5 mm in thickness and 4.6 mm in diameter, on average (Figures 3 & 4).

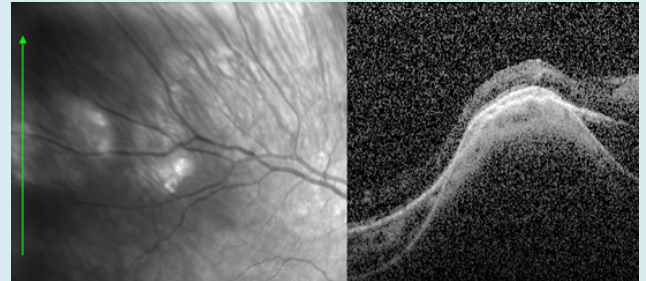


Figure 5: EDI-OCT of the right eye table mountain pattern.

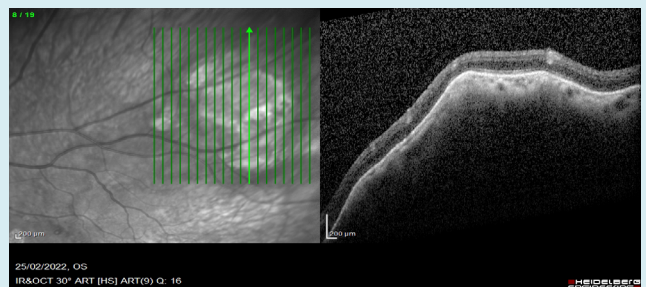


Figure 6: EDI-OCT of the left eye rolling pattern.

Enhanced Deep Imaging Optical Coherence Tomography (EDI-OCT) showed scleral lesion compatible with the presence of deposition of calcium with table mountain and rolling pattern (Figures 5 & 6).

Fundus auto fluorescence showed hyper auto-fluorescence in the areas of the lesions annular pattern on

the right eye and bright speckles on the left eye (Figures 7 & 8).



Figure 7: Fundus auto fluorescence of the right eye annular pattern.



Figure 8: Fundus auto fluorescence of the left eye bright speckles.



Figure 9: Fluorescein angiography and Indocyanine green angiography of the right eye.

Fluorescein angiography revealed hypo fluorescence in the arterial phase, and a mild hyperfluorescence in the venous phase with persistent late staining. Indocyanine green angiography showed hypofluorescence at the early, middle, and late phases (Figures 9 & 10).

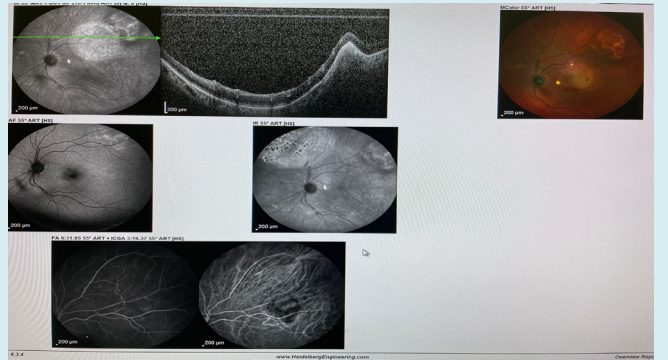


Figure 10: Fluorescein angiography and Indocyanine green angiography of the left eye.

Systemic investigation of calcium and phosphate metabolism was performed and the patient didn't display any underlying systemic disorder. The diagnosis of idiopathic sclerochoroidal calcification was made. At twelve month follow-up examination the lesions were stable with no lesion enlargement or decalcification or choroidal neovascularization.

Discussion

Sclerochoroidal calcification is an uncommon benign condition that may have an idiopathic, dystrophic or metastatic pathogenesis [3,8]. This condition is primary in 79% of cases and secondary in 21% [9]. Indeed dystrophic calcification can occur after chronic intraocular inflammation and ocular trauma [10]. In metastatic calcification the deposition of calcium in normal tissues is secondary to abnormal calcium-phosphorus metabolism displayed in chronic renal disease, hyperparathyroidism, hypervitaminosis D, hypophosphatemia and sarcoidosis [11-13]. Bartter's syndrome and Gitelman's syndrome are two rare conditions that can be associated with sclerochoroidal calcifications. Both are autosomic recessive renal tubular disorders of sodium-chloride transport. Bartter Syndrome generally occurs in childhood, with polyuria, polydipsia, vomiting and dehydration. These patients present hypokalemic metabolic alkalosis, increased potassium excretion, and normal-to-elevated levels of urine calcium excretion. The Gitelman syndrome is characterized by a later clinical presentation with fatigue, muscle weakness, and episodes of tetany. Hypocalciuria and hypomagnesaemia are the prominent features [14]. It is important to determine the

presence of these diseases because electrolyte abnormalities and potential cardiac arrhythmias can be fatal without appropriate management [15]. Calcium deposition was shown to be confined to the sclera [16]. Hasanreisoglu, et al. has classified sclerochoroidal calcification into four categories based on enhanced depth imaging optical coherence tomography. According to anterior surface topography of the mass, they described four patterns: flat, rolling, rocky-rolling and Table Mountain [17]. Based on this classification in our case the lesions showed a rolling pattern with a thinning of the overlying choroid at the apex on the left eye and a table mountain with a preserved overlying choroid on the right eye. Choroidal neovascularization in rare cases may complicate the course of sclerochoroidal calcification as reported by Battaglia Parodi, et al. that recently described two cases treated by intravitreal injection [18]. In literature are described about 70 cases. It is possible that the pathogenesis is related to the retinal pigment epithelium compression and this can lead to the development of choroidal ischemia in the area of the lesions with subsequent development of choroidal neovascularization [19]. In our case no neovascularization was displayed.

Conclusion

It is important to know and recognize this clinical entity. Although most cases of sclerochoroidal calcifications may be idiopathic as in the patient described in this case report, systemic laboratory workup must be performed to exclude calcium-phosphate metabolic disorder or primary renal tubular hypokalemic metabolic alkalosis syndromes (e.g. Gitelman syndrome, Bartter syndrome) that can be associated to these ocular signs. Moreover, given the rare possible development of CNV, a regular imaging check-up is recommended.

Funding

None.

Conflicts of Interest

The authors declare no conflict of interest, financial or otherwise.

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