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Case Report

Sclerochoroidal Calcifications: Case Presentation and Review

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Abstract

Sclerochoroidal calcifications are an important entity to recognize on ophthalmoscopy. Although uncommon, idiopathic, and primarily benign, they are commonly misdiagnosed as malignant tumors. Here, we present a case of an individual with sclerochoroidal calcifications with associated imaging findings in the setting of elevated serum parathyroid hormone levels. We also review background information, pathology, pathogenesis, differential diagnoses, diagnostic approach, management, and prognosis. It is crucial that patients with this finding have subsequent clinical imaging and laboratory workup to exclude other serious ocular and systemic diseases.

Keywords: Sclerochoroidal Calcification; Retina; Choroid; Sclera; RPE Atrophy

Abbreviations

SCC: Sclerochoroidal Calcifications; EDI-OCT: Enhanced Depth Imaging Optical Coherence Tomography; CNVM: Choroidal Neovascularization Membranes

Introduction

Sclerochoroidal calcifications (SCC) are primarily uncommon, incidental, benign, and idiopathic clinical findings on ophthalmoscopy [1,2]. The entity appears most often as multiple, discrete, whitish-yellow subretinal placoid lesions along the superior or inferior vascular arcades in asymptomatic middle age or older white individuals [1,3]. This report describes a case of SCC with associated fundus imaging, enhanced depth imaging optical coherence tomography (EDI-OCT), fluorescein angiography, B-scan, and a review of the literature.

Case Presentation

A 62 year-old asymptomatic man with hypertension and elevated parathyroid hormone of 103 (normal 14-72 pg/mL) presented with bilateral, yellow, deep, depigmented subretinal lesions in posterior poles of both eyes along the superotemporal arcades typical of sclerochoroidal calcification (Figure 1A and 1B). EDI-OCT further corroborated the calcifications in the scleral tissue with thinning of the overlying choroid (Figure 1C). Fluorescein angiography showed areas of hyperfluorescence at the sites of sclerochoroidal calcifications. B-scan revealed flat, calcified lesions with high A-scan reflectivity (Figure 1D). Lab testing revealed normal serum calcium, calcitonin, magnesium, and potassium levels. The patient is undergoing evaluation for subclinical hyperparathyroidism.

Discussion and Conclusion

Sclerochoroidal calcifications can be found in the mid-peripheral retina as solitary small homogenous yellow lesions, or occasionally as areas of focal RPE atrophy surrounded by a yellow halo [1]. They are usually found at the insertion of recti muscles [2]. They can present as single or multiple lesions, unilateral or bilateral, and flat or elevated [4]. SCC are typically found in both eyes. In cases where they have been associated with systemic conditions, they may occur in much younger individuals [1].

Pathology

Studies using both energy-dispersive X-ray analysis and Ramen vibrational microspectroscopy have confirmed that SCC are deposits of calcium pyrophosphate dihydrate in the sclera [5,6].

Pathogenesis

SCC can be dystrophic, metastatic, or predominantly, idiopathic calcifications. Dystrophic calcification is characterized by normal calcium-phosphorus metabolism, and generally occurs in the setting of severe ocular trauma or chronic intraocular inflammation. Metastatic calcification refers to abnormal calcium-phosphorus metabolism with subsequent deposition of calcium in normal tissues [1,2,7]. Cases are considered idiopathic only when the two former pathologies have been ruled out through ocular history and laboratory work-up.

Differential diagnoses

Although they are widely reported in the literature, they have often been misdiagnosed as amelanotic choroidal nevi, choroidal melanomas, choroidal metastases, or intraocular lymphomas [2]. Other conditions such as chorioretinitis, regressed retinoblastoma, retinal astrocytic hamartoma, choroidal hemangioma, choroidal osteoma, and eccentric macular degeneration can appear clinically similar to SCC but can be differentiated through classic ophthalmoscopic features by an experienced clinician [1].

Diagnostic approach

B-scan ultrasound usually reveals an echogenic plaque in the sclera, with A-scan ultrasound demonstrating a high spike at the site of the lesion with low amplitude echoes posteriorly [1]. A very small detachment of the sensory retina may be present directly over the lesion, although extended retinal detachment has not been observed. A few reports have highlighted the presence of choroidal neovascularization associated with SCC [2,8-10].

Computed tomography will also show a plaque at the level of the sclera. The lesions autofluorescence. Fluorescein angiography reveals a relative hypofluorescence in the arterial phase, minimal hyperfluorescence in the venous phase, progressive hyperfluorescence in the recirculation phase, and maximal hyperfluorescence in the late phases. This is likely due to greater staining in the affected sclera compared to the adjacent normal sclera [1].

Indocyanine green shows moderate hypofluorescence in the vascular filling phases, with mild later hyperfluorescence [1].

EDI-OCT images of SCC have revealed thinning or absence of the overlying choroid (mean choroidal thickness of 28 microns) [4,11,12]. The anterior surface has an undulating rocky (38%) or rolling (62%) appearance with moderate reflectivity (77%) and an optically bright anterior band (23%) [11]. Another OCT feature of SCC is the presence of a dense posterior shadowing cast from the anterior surface of the lesions. However, what differentiates SCC from choroidal neoplasms is the scleral-based origination and the absence of exudative signs in the overlying retina commonly seen with choroidal melanoma or metastasis [4].

A recent study investigated a large cohort of 67 eyes with clinically determined SCC and evaluated the specific EDI-OCT features and correlated them with clinical appearance and thickness. They proposed a four-group classification scheme based on the mountain-like configurations of each one: Type 1 “flat”, Type 2 “rolling”, Type 3 “rocky-rolling” and Type 4 “table mountain”. Type 1 was most commonly the thinnest and with the greatest preservation of the overlying choroid and RPE. The type 3 “rocky and rolling” SCC caused the most extreme thinning of the overlying choroid and retina with greater abnormalities in the RPE, ELM, and ellipsoid regions in the apical portions of the SCC, and type 4 “table mountain” had the largest basal diameter but with preserved overlying retinal structures [12].

A novel wide field of view swept-source OCT system has also been utilized to differentiate SCC from other peripheral retinal pathology [13].

Further management and prognosis

From an ocular perspective, most SCC are not near the fovea and therefore do not cause any visual disturbances or impairments. Even in rare cases of parafoveal involvement, no effective treatment protocols have been established. Visual prognosis generally remains excellent [1]. Prognosis may be guarded in individuals with renal failure secondary to the aforementioned systemic associations. A few cases of choroidal neovascularization, a rare complication, associated with SCC have been reported in the literature. Three of these cases with choroidal neovascularization membranes (CNVM) were treated successfully with argon laser photocoagulation with no reported recurrences, while the fourth

patient's CNVM spared the central visual acuity, remained stable, and was left untreated during the one year follow-up [2,8-10].

A large retrospective study of 179 eyes demonstrated that 79% of patients who are worked up after an initial preliminary diagnosis of SCC demonstrate normal calcium-phosphorus metabolism and no associated systemic or ocular causes for dystrophic or metastatic calcification [7,14]. Several large retrospective case series have shown that in the other 21%, however, several conditions associated with calcium-phosphorus dysregulation, such as hyperparathyroidism, pseudohypoparathyroidism, vitamin D toxicity, sarcoidosis, hypophosphatemia, calcium pyrophosphate dihydrate deposition disease, and chronic renal failure have been associated with metastatic SCC [1,2]. SCC has also been shown to be clinical features of Bartter and Gitelman syndromes, two types of primary renal tubular hypokalemic metabolic alkalosis [15,16]. Such patients may be asymptomatic and possibly remain undetected unless specific tests for renal tubular function are performed. These diagnoses can be treated quite effectively [17] and therefore it is imperative that patients who present with these lesions receive a complete workup before the calcifications are deemed idiopathic in origin.

Due to the less common but potentially treatable systemic associations described above, these patients must be worked up. Therefore, the following serum levels are recommended for patients suspected to have SCC: thyroid hormone, parathyroid hormone, calcitonin, BUN, and creatinine. Blood and urine should be checked for potassium, calcium, magnesium, phosphorus, and vitamin D levels [7].

In summary, although SCC is a benign ophthalmic finding, clinical imaging and laboratory workup are essential to exclude other serious ocular and systemic diseases.

Financial Disclosures

None.

Proprietary Interest

None.

Conflicts of Interest

None.

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