Case Report

Stones, bones, groans, thrones, and psychiatric overtones: Systemic associations of sclerochoroidal calcification

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Sclerochoroidal calcification (SCC) is a frequent masquerader of choroidal melanoma with important systemic associations such as hyperparathyroidism and parathyroid adenoma. Herein, we describe a case of a 67-year-old male who presented with an amelanotic choroidal lesion in the right eye (OD) and a history of kidney stones. Ultrasonography showed the lesion to be flat and calcified OD. Incidentally, a subclinical calcified plaque was also found in the fellow eye. Optical coherence tomography showed an elevated suprachoroidal mass in a table mountain configuration OD and flat configuration left eye, consistent with type 4 and type 1 SCC. The patient was referred for metabolic testing to rule out the underlying electrolyte imbalance and was found to be normal.

Keywords: Eye, hyperparathyroidism, parathyroid adenoma, sclera, sclerochoroidal calcification

Introduction

Sclerochoroidal calcification (SCC) is characterized by calcium deposition within the sclera giving rise to flat or protruding yellow-white amelanotic lesions in the fundus.[1,2] SCC can masquerade as various intraocular tumors and is frequently misdiagnosed as choroidal metastasis or choroidal osteoma. The confusion with choroidal osteoma stems from the presence of calcification within both tumors; however, the clinical features differ in that osteoma affects young patients whereas SCC affects the elderly.[3] Furthermore, choroidal osteoma is typically flat and peripapillary in location with linear opacities within the choroid on optical coherence tomography (OCT).[4] In contrast, SCC appears more often in the superotemporal equator as a nodular elevation with shadowing on OCT emanating from the sclera outward.[1]

Most cases of SCC require no treatment, since calcium deposition is often stable and vision loss does not generally occur.[1] SCC is more often idiopathic, but in some cases, calcification could be indicative of an underlying imbalance in calcium/phosphate metabolism. Such imbalances have rarely been associated with renal dysfunction due to Gitelman syndrome and Bartter syndrome, and more frequently hyperparathyroidism.[1] Hyperparathyroidism manifests clinically with renal stones, bone pain, abdominal pain, polyuria, and psychiatric symptoms such as depression, anxiety, insomnia, cognitive dysfunction, and even coma.[1] This collection of symptoms has led to the popular mnemonic device of "stones, bones, groans, thrones, and psychiatric overtones."[1]

Herein, we report a recent case of a 67-year-old male with kidney stones referred for multifocal yellow-white tumors in the choroid and found to have SCC.

Case Report

A 67-year-old white male was referred for a small amelanotic choroidal tumor in his right eye (OD) found on routine examination and suspected to be choroidal nevus or metastasis. Medical history revealed primary open angle glaucoma, kidney stones of unspecified composition, high cholesterol, and surgical

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removal of his gallbladder. On examination, visual acuity was 20/20 in both eyes. Intraocular pressure was 14 mmHg in the OD and 13 mmHg in the left eye (OS), and both eyes showed mild nuclear sclerosis on slit-lamp examination.

Fundus examination of the OD revealed a poorly defined yellow deep nodular lesion superotemporally, appearing to reside in the choroid [Figure 1a]. This lesion was dense on ultrasonography with acoustic shadowing consistent with calcification. Although the clinical examination of the left fundus was unremarkable, ultrasonography confirmed flat dense lesions superotemporally with calcification and orbital shadowing. Both eyes were consistent with SCC.

OCT revealed scleral thickening and shadowing in both eyes, associated with compression of the overlying choroid, more so the OD than the OS. These lesions are consistent with SCC and are distinguished from other choroidal tumors by its suprachoroidal location, preserved overlying choroidal and retinal structures, and thinning or the choroid at its margins.[5] Furthermore, they can be classified as type 4 or "table mountain" formation in the OD and type 1 or "flat" in the OS.[5] A clinical diagnosis of SCC was made, and the patient was referred for testing of serum calcium, phosphorus, magnesium, and potassium as well as parathyroid hormone. This, along with the renal stones, suggested the underlying calcium disequilibrium.

Discussion

The diagnosis of SCC is often overlooked. In one series of 118 consecutive patients, a correct referring diagnosis was made in only 5% of the cases.[1] Further, there have been reported cases of SCC erroneously treated with radiotherapy for fear of a more serious diagnosis such as melanoma.[6] Hence, the correct diagnosis of SCC is important to avoid unwarranted treatment that could result in visual compromise.

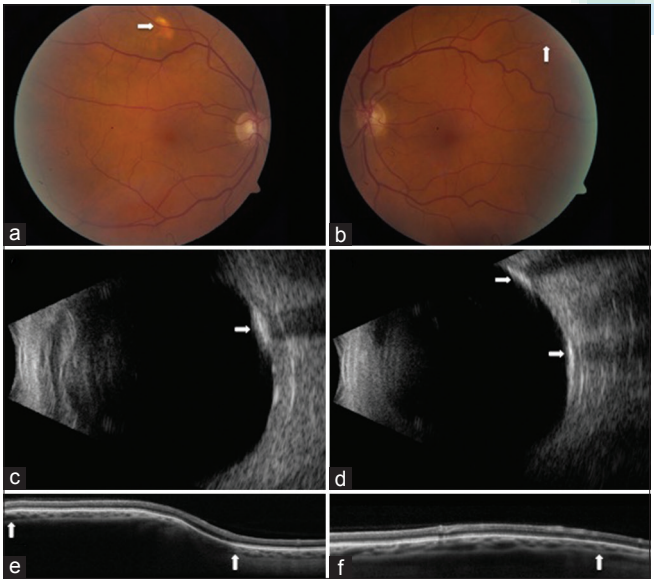

Figure 1: A 67-year-old male presented with an amelanotic lesion in the right eye along the superotemporal quadrant (a). Fundus examination of the left eye was unremarkable (b), but arrow points to the site of ultrasonographically detected deep calcification. Ultrasonography of the right eye (c) depicted a flat hyperechoic lesion with shadowing consistent with calcification, and this was similarly found superotemporally in the left eye (d). Optical coherence tomography showed suprachoroidal hyporeflective lesions consistent with sclerochoroidal calcification in a table mountain formation in the right eye (e) and a flat formation in the left eye (f)

The occurrence of SCC is often isolated without systemic associations. The initial reports on SCC demonstrated that this condition is rarely associated with primary hyperparathyroidism, hypomagnesemia, Gitelman syndrome, and Bartter syndrome.[7-10] A more recent update, however, provided evidence that association with electrolyte imbalance in the setting of hormonal abnormalities could occur more often than expected. Our team reported parathyroid adenoma in 15% of the patients with SCC and hyperparathyroidism in 27% of those who underwent systemic testing.[1] The parathyroid gland is composed primarily of chief cells that synthesize and release parathyroid hormone to regulate calcium and phosphate metabolism. Parathyroid adenoma is caused by abnormal proliferation of chief cells leading to elevated parathyroid hormone (primary hyperparathyroidism), while the secondary hyperparathyroidism is more often caused by renal dysfunction with elevated serum calcium requiring compensation by the parathyroid gland. Our patient manifested with two features of metabolic calcium dysfunction including renal stones and SCC, both potentially related to systemic hypercalcemia, possible renal dysfunction, and secondary hyperparathyroidism.

Primary renal tubular hypokalemic metabolic alkalosis syndromes such as Bartter and Gitelman syndromes are more serious conditions associated with SCC.[7-10] These renal abnormalities constitute a group of similar autosomal recessive disorders of sodium chloride transport causing renal tubular hypokalemic metabolic alkalosis.[11] Clinically, Bartter syndrome presents in childhood with polyuria, polydipsia, vomiting, frequent dehydration, and failure to thrive.[11] Gitelman syndrome is characterized by a milder onset in late childhood or early adulthood, with clinical symptoms including fatigue, muscle weakness, and carpopedal spasm.[11] These symptoms are in addition to the classical presentation of hypokalemia and metabolic alkalosis. Patients with these syndromes are at a high risk for vascular incident with the use of anesthesia.[11] Therefore, it is important to be aware of these syndromes in addition to hyperparathyroidism in the differential diagnosis when considering SCC. Both of these renal abnormalities are treatable, and symptoms are reversible with proper treatment. Bartter syndrome is primarily treated with potassium supplementation and prostaglandin synthase inhibitors, with the main goal of correcting hypokalemia.[11] Gitelman syndrome is treated with oral magnesium pyrrolidone carboxylate supplementation.[11] The mechanism of SCC in relation to Bartter and Gitelman syndromes is not yet understood.

In summary, SCC is a benign calcified tumor seen in older adults that is visually insignificant. Although treatment is not required, blood tests are necessary to check for electrolyte imbalance and hormonal abnormalities in relation to calcium, phosphorus, magnesium, and potassium metabolism.

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Conflicts of interest

There are no conflicts of interest.

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