Sclerochoroidal Calcification Leads To Diagnosis Of Primary Hyperparathyroidism And Parathyroid Adenoma
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Abstract:
A 62 year old Caucasian male presents with an incidental finding of an elevate retinal lesion. Ophthalmic ancillary testing is consistent with sclerochoroidal calcification. Subsequent metabolic work up and diagnostic imaging reveal a parathyroid adenoma.

I. Case History
1) Patient demographics: 62 year old Caucasian male.
2) Chief complaint: Patient presents to clinic for a comprehensive eye exam. No ocular or visual complaints.
3) Ocular history: Cataract surgery OU, mild hyperopia
4) Medical history: Multisystem vascular disease including DM x 6 years, HTN, hyperlipidemia, and arteriosclerosis. Patient also has a history of hypothyroidism.
5) Medications: Levothyroxine and insulin

II. Pertinent findings
1) Clinical: Presenting BCVA was 20/25 sc OU. Intraocular pressures, extra ocular motilities, confrontation fields, and pupillary testing were all normal. Slit lamp examination revealed no calcium deposition on the cornea or conjunctiva. Dilated fundus exam revealed a 3DD pale, elevated lesion 1.5DD superotemporal to the ONH OS. There was no choroidal neovascularization present, nor did the patient have any other anomalous retinal or ONH findings. B-scan ultrasonography confirms dense hyper-reflectivity of the lesion with shadowing. Further, eccentric fixation 5-line raster optical coherence tomography confirms steep elevation of the sclerochoroidal region underlying an intact retina. OCT imaging reveals “mountain-like” elevation of the chorioretinal area, a finding unique to sclerochoroidal calcific lesions.
2) Laboratory Studies: Following presentation in the eye clinic, parathyroid hormone, calcitonin, calcium, magnesium, phosphorous, potassium, and alkaline phosphate levels were ordered. Parathyroid hormone was significantly elevated to 124.9pg/ml (normal range 29-79 pg/ml). Calcium levels were also mildly elevated to 10.3 mg/dl on two occasions (normal range 8.4-10.0 mg/dl).
3) Radiology studies: Ultrasonography of the parathyroid region localizes an enlargement of the left lobe side to 4.2x1.2x1.4 cm (for reference, the right side measured 2.0x2.0x1.8 cm). Parathyroid immunoassay with radioactive$^{99m}$Tc-MIBI is positive with abnormal uptake on the right glad side.

III. Differential diagnosis
1) Primary/leading: The most commonly cited cause of sclerochoroidal calcification is an imbalance in the metabolic rates of calcium, potassium, and magnesium. Often, patients with sclerochoroidal calcifications are on diuretics for hypertension and excrete excess electrolytes into their urine, resulting in elevated calcium levels and deposition of calcium in the retina. This phenomenon is known as Pseudo-Bartter Syndrome and is relatively benign. The patient in this
case report is hypertensive but was not taking diuretic medication at the time of nor before the diagnosis. Therefore, it is very unlikely the condition results from electrolyte wasting in urine. Less commonly, the sclerochoroidal calcifications are associated with metabolic alkalosis in patients with renal failure secondary to tubular hypokalemia. In extremely rare circumstances (and in this case), sclerochoroidal calcification is the direct result of hypercalcemia secondary to hyperparathyroidism with or without an associated parathyroid adenoma. Ancillary ophthalmologic testing performed during the eye exam strongly suggests the presence of a calcific deposit. A full endocrinology consultation including blood specimen, ultrasonography, and radioactive immunoassay with $^{99m}$-Tc-MIBI confirm the presence of primary hyperparathyroidism and parathyroid adenoma. Such a wealth of correlative data leaves little ambiguity in the diagnosis.

2) Others: Sclerochoroidal calcifications may be confused for choroidal metastases, choroidal osteomas and choroidal nevi. In a study conducted at the Oncology Service at Wills Eye Hospital, 27 patients were ultimately diagnosed with sclerochoroidal calcification. Of these 27 patients, the referring diagnoses were 26% choroidal metastasis and 11% choroidal nevus. In this case, the patient has no history of cancer; therefore, metastasis can be ruled. Due to the elevated, dense nature of the lesion in question and its persistence under red-free light, choroidal nevus is an incorrect diagnosis.

IV. Diagnosis and discussion
1). Elaborate on the condition: Metastatic calcification is a process describing inappropriate calcium deposition in normal tissue throughout the body. Sclerochoroidal calcification pertains to metastatic calcium deposition primarily involving the sclera, choroid, and rarely the retina. It is believed that the sclera becomes involved first, often leaving the normal fundus appearance unchanged. Progression of the condition is indicated by choroidal involvement and atrophy of the RPE, as well as other retinal layers. According to Hasanreisoglu et al., sclerochoroidal calcific lesions have a characteristic “mountain-like, table top” pattern on enhanced depth OCT. The shape is unique to this condition. In very progressed cases, small serous retinal detachments and choroidal neovascular membranes may ensue due to extensive chorioretinal degeneration. Vision is typically unaffected and the patient visually asymptomatic unless serous RD or CNVM compromise the macula.

2) Expound on unique features: Sclerochoroidal calcification is typically of an idiopathic or benign etiology. Rarely are such lesions the result of primary hyperparathyroidism or a parathyroid adenoma. A clinical chart review of 118 patients and 179 eyes showed only 5 participants with an associated parathyroid adenoma. In this case, the patient had no outward signs or symptoms typically associated with hyperparathyroidism, such as polyuria, muscle weakness, or bone pain. A dilated retinal examination incited suspicion and ultimately lead to the diagnosis and treatment of progressed systemic disease.

V. Treatment, management
1) Treatment and response to treatment: The ocular and visual complications of sclerochoroidal calcification and parathyroid disease are minimal. Vision is typically unaffected and the patient
asymptomatic. However, the patient will undergo routine dilated fundus examinations to monitor the size of the lesion, serous detachments, choroidal neovascular membranes, and/or deposition of conjunctival and corneal calcium. The patient is currently being co-managed by an endocrinologist and is being considered for surgical removal of the parathyroid adenoma.

2) Research/ Literature review/ Bibliography: References are available below and include detailed findings of all supportive data.

VI. Conclusion
A differential diagnosis of sclerochoroidal calcification must be considered in the presence of pale, elevated retinal lesions. B-scan, OCT, and blood work targeting parathyroid hormone, calcitonin, calcium, magnesium, phosphorous, potassium, and alkaline phosphate levels should be performed. Anomalies on these tests warrant a consultation with endocrinology for continuity of care.

References


Image 1: Fundus photography showing location and elevation of retinal lesion
Image 2: Eccentric 5-Line Raster OCT showing characteristic “mountain-like” elevation of sclerochoroidal area