

Sclerochoroidal Calcification is Primarily a Scleral Condition Based on EDI-OCT

BY PATRICK W. SHIELDS, MHS, AND CAROL L. SHIELDS, MD

Sclerochoroidal calcification is an uncommon ophthalmic condition characterized by yellow-white subretinal lesions classically located in the superotemporal midperipheral region of the fundus.¹ This condition has a typical appearance, which is recognizable by indirect ophthalmoscopy as an abruptly elevated mass with overlying retinal pigment epithelial atrophy and choroidal thinning. Diagnostic evaluation using ultrasonography can confirm the presence of intrinsic calcification. Enhanced depth imaging optical coherence tomography (EDI-OCT) has provided new information on the features of sclerochoroidal calcification, demonstrating that this lesion, previously believed to be of scleral and choroidal origin, is most likely primarily of scleral origin.

We describe a patient with sclerochoroidal calcification and describe the classic EDI-OCT features of this condition.

CASE REPORT

A 64-year-old white man with a history of osteoarthritis and gastroesophageal reflux disease was referred with an asymptomatic, newly diagnosed yellow choroidal mass in the left eye suspicious for a nevus, melanoma, or metastasis. He denied having kidney problems, taking

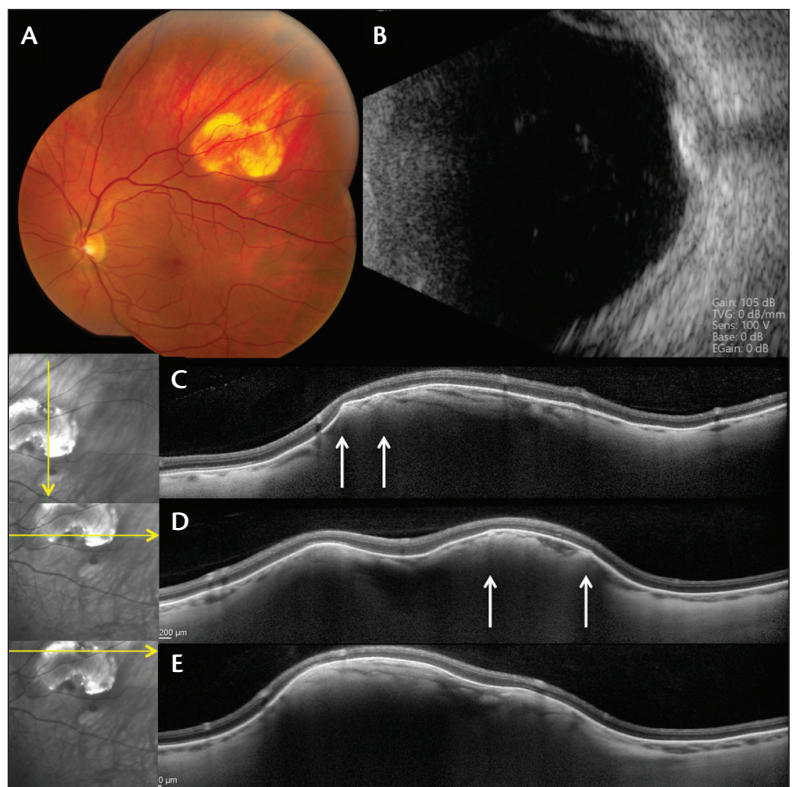


Figure. A 64-year-old white man with sclerochoroidal calcification. The geographic, yellow, deep lesion in the left eye with overlying atrophy of the retinal pigment epithelium and choroid is noted above the superior vascular arcade (A). There is dense calcification and shadowing on ultrasonography (B). On EDI-OCT, there is dramatic “rocky and rolling” elevation with no subretinal fluid. The mass arises within the sclera and compresses the choroid inward. The “rocky” pointed (arrows) or jugged surface (C,D) and “rolling”, smooth surface (E) are noted on various horizontal and vertical scans.

diuretic medications or calcium supplementation, and having ever been diagnosed with Gitelman or Bartter syndrome.

On examination, visual acuity was 20/25 in the right eye and 20/20 in the left eye. Intraocular pressures were 17 mm Hg in each eye. The anterior segment was unremarkable in each eye, and there was no calcified scleral plaque of Cogan.

Fundus examination of the right eye was unremarkable. In the left eye, there was an amelanotic mass, deep to the retina, measuring 4.5 mm x 3 mm in diameter (Figure). The choroidal vessels appeared draped over the mass. Ultrasonography disclosed a calcified mass within the posterior wall of the globe with dramatic elevation measuring 2.7 mm in thickness. On EDI-OCT, the mass was clearly depicted within the sclera, showing an abruptly elevated, jutting surface topography that pushed the choroid inward and elevated both the choroidal and retinal tissue. There was no subretinal fluid.

Based on these findings, the diagnosis favored sclerochoroidal calcification.

The patient was observed without treatment. Metabolic evaluation revealed normal levels of serum calcium at 9.1 mg/dL (normal 8.4–10.2), potassium at 3.9 mmol/L (normal 3.6–5.0), phosphorus at 3.4 mg/dL (normal 2.5–4.6), and magnesium at 2.1 mg/dL (normal 1.8–2.5).

DISCUSSION

Sclerochoroidal calcification is a recently recognized asymptomatic fundus abnormality occurring most often in older individuals and is characterized by benign, yellow-white lesions.^{1,2} These lesions are located deep to the retina and were previously presumed to occupy the sclera and choroid, hence the term sclerochoroidal calcification. According to the literature, it was believed that the earliest signs of calcification occurred in the sclera and eventually involved both the sclera and choroid.^{3,4}

EDI-OCT images show that sclerochoroidal calcification is localized primarily within the sclera with secondary compression of the choroid. Fung et al analyzed 17 cases of sclerochoroidal calcification using EDI-OCT.⁵ The mean age of patients was 74 years.⁵ The lesions were most often superotemporal (n = 12), were yellow in color (n = 17), and had a mean diameter of 3.5 mm. On EDI-OCT, the lesions were all in the sclera with choroidal compression or absence (n = 17). The authors described the surface topography of this condition as “rocky and rolling.” The “rocky” topography showed abrupt elevation, often with a pointed and jutting surface and generalized widening of the scleral wall. The “rolling” topog-

raphy showed more gentle elevation with a smooth surface undulation. Some cases showed both rocky and rolling surface.

Systemic workup should be performed for all patients with sclerochoroidal calcification to screen for calcium-phosphate metabolic abnormalities and to evaluate for renal tubular hypokalemic metabolic alkalosis as a primary disease (ie, Gitelman syndrome, Bartter syndrome) or a secondary disease (eg, from diuretic medication). The workup should include measurements of serum and urine levels of calcium, potassium, phosphorus, and magnesium, as well as serum levels of parathyroid hormone and calcitonin.¹ Occasionally, parathyroid adenoma with excessive serum calcium elevation has been found following recognition of sclerochoroidal calcification.⁶

CONCLUSION

EDI-OCT can pinpoint sclerochoroidal calcification within the sclera as a “rocky or rolling” appearance. This finding questions previous terminology presuming the calcification to be within both the sclera and the choroid. On EDI-OCT, the calcification appears precisely as “scleral calcification.”⁷ ■

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Carol L. Shields, MD, is the co-director of the Ocular Oncology Service, Wills Eye Hospital, Thomas Jefferson University, Philadelphia. She is a member of the Retina Today Editorial Board. Dr. Shields may be reached at carol-shields@gmail.com.

Patrick W. Shields, MHS, is a medical student at the Sidney Kimmel Medical School at Thomas Jefferson University.



- Hundle R, Turaka K, Shields CL. Sclerochoroidal calcification resembling choroidal metastasis. *Retina Today*. July/August 2011;57-58.
- Sivalingam A, Shields CL, Shields JA, et al. Idiopathic sclerochoroidal calcification. *Ophthalmology*. 1991;98:720-724.
- Shields JA, Shields CL. Sclerochoroidal calcification. The 2001 Harold Gifford Lecture. *Retina*. 2002;22:251-261.
- Schachat AP, Robertson DM, Mieler WF, et al. Sclerochoroidal calcification. *Arch Ophthalmol*. 1992;110:196-169.
- Fung AT, Arias JD, Shields CL, Shields JA. Sclerochoroidal calcification is primarily a scleral condition based on enhanced depth imaging optical coherence tomography. *JAMA Ophthalmol*. 2013;131(7):960-963.
- Choi JY, Bianciotto C, Shields JA, Shields CL. Sclerochoroidal calcification in a patient with chronic hypercalcemia from undiagnosed parathyroid adenoma. *Retin Cases Brief Rep*. 2009;3:431-433.
- Honavar S, Shields CL, Demirci H, Shields JA. Sclerochoroidal calcification: Clinical manifestations and systemic associations. *Arch Ophthalmol*. 2001;119:833-840.