THE RETINA PARTNERS

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Case of the Month – April 2021

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A 69-year-old male was referred for evaluation of “bilateral elevated lesions.” He denied any ocular complaints. He had a history of orthopedic surgeries for his back and neck, but was otherwise in good health. He was taking Gabapentin 300 mg PO QD for pain and Aspirin 81 mg PO QD.

Visual acuity was 20/20 in the right eye (OD) and 20/20 in the left eye (OS). Intraocular pressures were 12 OD and 10 OS. Pupils were equal, round, reactive, and there was no relative afferent pupillary defect. Slit lamp exam was unremarkable aside from 2+ nuclear sclerotic cataract in both eyes. On dilated fundus exam, there were multiple, yellow, elevated subretinal deposits superior to the arcades in each eye. There was no subretinal fluid or orange pigment. On fluorescein angiography there was early hypofluorescence with late hyperfluorescent staining corresponding to the subretinal deposits. B-scan ultrasonography conveyed hyperechoic foci with posterior shadowing, suggestive of calcifications.

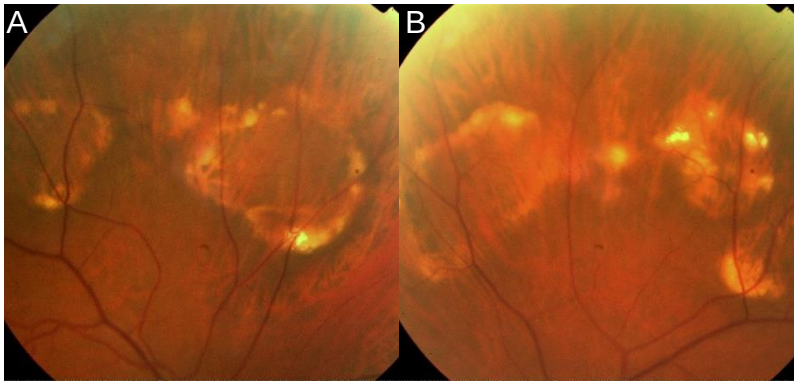

Figure 1: Fundus photographs of the (A) right eye and (B) left eye showing multiple, yellow elevated subretinal deposits superior to the arcades consistent with sclerochoroidal calcifications.

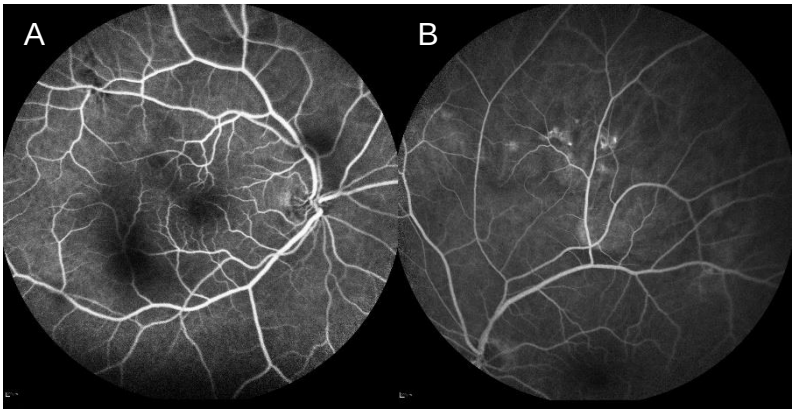

Figure 2: Fluorescein angiography of the (A) right and (B) left eyes showing early hypofluorescence (A) and late hyperfluorescent staining (B) of the sclerochoroidal calcifications. There is no secondary neovascularization noted.

Clinical Course:

The patient was counseled that his findings were consistent with sclerochoroidal calcifications and observation was recommended. The findings were communicated with his Primary Care Physician and a systemic laboratory workup for hyperparathyroidism and hypomagnesemia were recommended. The patient was reassured that sclerochoroidal calcifications are typically idiopathic, non-progressive, and systemic laboratory workups are often normal. The patient was given a follow-up appointment in 3 months.

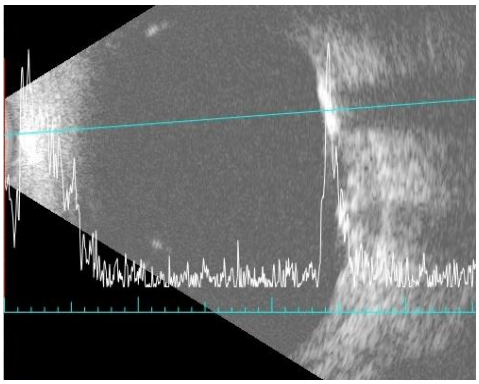

Figure 3: B-Scan ultrasonography of the right eye showing hyperechoic foci with posterior shadowing, consistent with calcifications. There is high internal reflectivity noted on the superimposed A-scan.

Discussion:

Sclerochoroidal calcifications are depositions of calcium in the sclera and/or the choroid, which are rare, and are most commonly primary or an idiopathic finding. They can also occur secondary to hypercalcemia, or in association with Gitelman or Bartter syndromes. Sclerochoroidal calcifications typically present as multiple foci, in both eyes, superiorly or superotemporally, most commonly from the vascular arcades to the mid-periphery. The differential diagnosis includes choroidal osteomas, choroidal metastases, choroidal nevus, and amelanotic melanoma. In contrast to sclerochoroidal calcifications, choroidal osteomas are typically solitary lesions, unilateral, and juxtapapillary.

In one single-center retrospective series of 27 patients with sclerochoroidal calcifications, the mean age was 70, all patients were white, and the finding was noted incidentally on routine examination. In this cohort, 59% of patients had unilateral findings and visual acuity was 20/50 or better in 97%. There was a mean of 2 foci of sclerochoroidal calcifications, and 53% of foci were yellow and 42% were yellow-white. Mean diameter on ultrasonography was 2.6 mm with a mean thickness of 1.1 mm; 58% of foci were post-equatorial, 39% were along the temporal vascular arcades, and 56% were in the superotemporal quadrant. All lesions remained stable in size and shape with a mean follow-up of 38 months, though one patient developed a secondary choroidal neovascular membrane. One of 19 patients (5%) was found to have primary hyperparathyroidism, while 6 of 13 were found to have hypomagnesemia (46%). Four patients (31%) were diagnosed with Gitelman syndrome.

Sclerochoroidal calcifications are most commonly mistaken for a choroidal osteoma or choroidal metastasis. While there are certain features on clinical examination that help differentiate sclerochoroidal calcifications from other entities, it is also important to take a thorough history of present illness, past medical history, and to carefully review the patient's medications. Several conditions with abnormal calcium-phosphorus metabolism can cause secondary sclerochoroidal calcifications, including hyperparathyroidism, pseudohypoparathyroidism, vitamin D toxicity, sarcoidosis, hypophosphatemia, chronic renal failure, and renal tubular hypokalemic metabolic alkalosis syndromes such as Bartter and Gitelman syndromes. These patients can be asymptomatic and sometimes have normal calcium-phosphorus metabolism; in these cases, the diagnosis can only be made by specific renal tubular function tests.

Bartter and Gitelman syndromes are both autosomal recessive renal tubular disorders of sodium-chloride transport. Patients have a primary renal tubular hypokalemic metabolic alkalosis. In Bartter syndrome, patients usually present in childhood with polyuria, polydipsia, vomiting, dehydration, and failure to thrive. Patients have hypokalemic metabolic alkalosis, increased potassium excretion, and normal to high levels of urine calcium excretion. There may be an associated hypomagnesemia. It is thought that the primary defect in classic Bartter syndrome is in the ascending limb of the loop of Henle, owing to a molecular defect in the sodium-potassium-chloride cotransporter or the basolateral chloride channel. Patients with Gitelman syndrome typically have milder signs and symptoms and present older in age. They may present with fatigue, muscle weakness, and spasms of the hands and feet. Hypocalciuria and hypomagnesemia are commonly noted. In Gitelman, there is a mutation in the sodium-chloride cotransporter of the distal nephron with a genetic defect at 16q13.

Findings consistent with sclerochoroidal calcifications on dilated fundus examination should be communicated to the patient's Primary Care Physician. It can be helpful to also communicate the numerous, albeit rare, systemic diseases that can be associated with this finding.

Take Home Points: Sclerochoroidal Calcifications

- Sclerochoroidal calcifications usually present as discrete yellow-white lesions in asymptomatic white patients of mean age 70. There is minimal risk for vision loss, unlike with choroidal osteomas.
- Most cases are idiopathic, but a minority can be due to systemic disorders involving hypercalcemia, hyperparathyroidism, renal metabolic conditions, or abnormal calcium-phosphorus metabolism.
- Findings should be communicated with the patient's Primary Care Physician who may consider further evaluation for systemic disease.


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