CASE REPORT

Bilateral Adie's Tonic Pupil and Hyperopic Shift in Idiopathic Sclerochoroidal Calcification

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ABSTRACT

Sclerochoroidal calcification is a rare but recognised ophthalmic manifestation seen mostly in elderly Caucasian individuals. The lesions, often bilateral, appear as yellow-white irregular subretinal lesions usually found along the mid-peripheral fundus. Though typically asymptomatic, sclerochoroidal calcification has rarely been associated with parafoveal involvement, choroidal neovascularisation, and serous detachment of the calcifications. Visual involvement is typically minimal, and neovascularisation is often visually insignificant. We present a rare case of sclerochoroidal calcification in a 64-year-old Caucasian female who presented with painless progressive bilateral vision loss and a hyperopic shift with subsequent development of bilateral sequential Adie's tonic pupil. To the best of our knowledge, this is the first such report in the English language literature.

Keywords: Adie's tonic pupil, hyperopic shift, sclerochoroidal calcification

INTRODUCTION

Sclerochoroidal calcification is a rare but recognised ophthalmic manifestation seen mostly in elderly Caucasian individuals.¹ The lesions, often bilateral, appear as yellow-white irregular subretinal lesions usually found along the mid-peripheral fundus. Though typically asymptomatic, sclerochoroidal calcification has rarely been associated with parafoveal involvement, choroidal neovascularisation, and serous retinal detachment.² Visual involvement is typically minimal, and neovascularisation is often visually insignificant. We present a rare case of sclerochoroidal calcification in a 64-year-old Caucasian female who presented with painless progressive bilateral vision loss and a right-sided hyperopic shift with subsequent development of bilateral sequential Adie's tonic pupil. To the best of

our knowledge, this is the first such report in the English language literature.

CASE

A 64-year-old Caucasian female with a past medical history of hypothyroidism presented in August 2010 with a 2-year history of bilateral gradual painless progressive visual loss and chorioretinal striae. She had no history of thyroid eye disease, had not been treated previously with surgery or radioactive iodine therapy, and had a normal thyroid function study 6 months ago. The patient also denied any signs or symptoms of giant cell arteritis (i.e. scalp tenderness, temporal nodularity, or jaw claudication). She was seen by an outside ophthalmologist who reported a 1-dioptre right-sided hyperopic shift (+2.75

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-0.75×144 to $+3.75 -1.75 \times 088$) from February 2010 to June 2010. An outside fluorescein angiogram (FA) was performed that showed bilateral hyperfluorescence in the temporal macula and periphery surrounding the chorioretinal folds in the arterial phase, with persistent late staining in the later phases. The remainder of the patient's past surgical, family, social, and ocular history was noncontributory. The patient's medications included only synthroid, fluoxetine, and amitriptyline.

On examination, best-corrected visual acuity (BCVA) was 20/25 OU with a refraction of $+4.00 +2.75 \times 176$ OD and $+0.50 +0.75 \times 022$ OS. The pupil was irregular and oval measuring 4×3.5 mm OD and round measuring 5 mm OS. There was inferior sector paresis OD, mild light-near dissociation OD, and a right-sided relative afferent pupillary defect (RAPD). External examination was significant for an inferior sclera show of 2 mm. Motility and intraocular pressure examinations were normal OU. Slit-lamp biomicroscopy was normal except for the iris sector paresis. The sclera showed no evidence of scleritis or edema. Dilated funduscopy examination showed extensive chorioretinal scarring, pigmentary change, peripheral geographic atrophy, and diffuse multifocal hyperplasia of the retinal pigment epithelium OU, more significant on the right than the left (Figure 1). Scattered peripheral haemorrhages were seen OU, but there was no evidence of active chorioretinitis. The cup-to-disc ratio was 0.3 OU. Humphrey visual fields showed nonspecific scatter OU, with a mean deviation of -3.86 dB OD and -2.52 dB OS. Cranial magnetic resonance imaging (MRI) was normal except for sclera thickening in the right orbit. Subsequent laboratory testing (complete blood count [CBC], basic metabolic panel [BMP], fluorescent treponemal antibody absorption [FTA-ABS] test, rapid plasma reagin [RPR], erythrocyte sedimentation rate/C-reactive protein [ESR/CRP], parathyroid hormone [PTH]) was within normal limits (with a normal calcium level) except for an elevated angiotensin-converting enzyme

(ACE) level of 77 (normal 8-52 units/L). Vitamin A levels were normal, and a subsequent paraneoplastic antibody panel was negative. An electroretinogram (ERG) was within normal limits. A chest x-ray showed no evidence of hilar lymphadenopathy, and a full-body positron emission tomography (PET) scan was normal except for a calcified indeterminate lesion in the lower pole of the left kidney with low suspicion for malignancy. A lumbar puncture (LP) was negative with normal cytology. A- and B-scan ultrasonography demonstrated bilateral plaque-like lesions with increased echogenicity, reflective surface spikes, and posterior orbital shadowing. Computed tomography (CT) of the orbits demonstrated bilateral inferolateral choroidal/scleral calcifications (Figure 2).

Serial follow-ups in March 2011 and November 2011 showed no interval changes in the patient's ophthalmic examination. In October 2012, an outside optometrist noticed progression of the right-sided hyperoptic shift and her refraction on exam was $+6.00 -2.00 \times 094$ OD and $+0.75 -0.59 \times 097$ OS. BCVA was 20/25 OD and 20/30 OS. Pupils measured 4 mm in the dark and were sluggishly reactive with light-near dissociation OU. The right pupil remained irregular, with a segment of hypoactivity and surrounding vermiform movement consistent with tonic pupil. Topical pilocarpine 0.1% was administered and confirmed the presence of bilateral Adie's tonic pupil.

DISCUSSION

The differential diagnosis for amelanotic dystrophic calcifications in the mid-peripheral fundus includes entities such as choroidal osteoma, choroidal metastases, choroidal nevus, ocular large-cell lymphoma, and choroidal granuloma.³ In our patient, workup for neoplasia was negative and there was no evidence of sarcoidosis or ocular tuberculosis. The FA showed findings typical of sclerochoroidal calcification, including mild to moderate hypofluorescence in the

FIGURE 1. Fundus photograph of right (A) and left (B) eye demonstrated well-circumscribed plaque-like areas of sclerochoroidal calcification, right significantly more affected than left, with chorioretinal scarring and deeper radial chorioretinal folds present in the right eye (arrow).

FIGURE 2. Ultrasonogram demonstrating sclerochoroidal calcification. B-scan ultrasonogram (top) with overlying A-scan (bottom) demonstrating an hyperechoic area of sclerochoroidal calcification with posterior shadowing (arrows), with corresponding high reflectivity by A-scan typical of sclerochoroidal calcification (arrowheads) in the right eye (A) and left eye (B). Axial (C) and sagittal (D) computed tomographic (CT) scans showing bilateral calcified areas in the sclerochoroidal interface, significantly more pronounced in the right orbit (arrows).

arterial phase and persistent late staining in the venous phase.¹ Ultrasonography showed echodense placoid lesions at the scleral-choroidal interface with acoustic orbital shadowing, also characteristic of calcification seen in sclerochoroidal calcification.² Although choroidal osteoma can present similarly, the patient's age, race, bilateral presentation, and fundus appearance were all more consistent with the diagnosis of sclerochoroidal calcification.

Idiopathic sclerochoroidal calcification is a typically benign condition thought to be a manifestation of dystrophic calcification in patients with normal calcium and phosphorus homeostasis.³ Lesions often occur at or near sites of stress, with calcification occurring most commonly at the insertion sites of the oblique muscles. The lesions are bilateral in most cases and may be almost flat (i.e. plaque-like) or raised (i.e. tumour-like) in appearance with the overlying retina not typically involved. Prior reports have recommended routine follow-up for patients with sclerochoroidal calcification due to an increased risk of neovascularisation and serous retinal detachment.³

Though not previously shown to be associated with sclerochoroidal calcification, hyperopic shifts have been associated with a variety of rapidly growing choroidal lesions, including choroidal hemangioma, choroidal melanoma, choroidal lymphoma, posterior scleritis, and Harada's syndrome.^{4,5} Mechanical factors likely contributed to this patient's subsequent development of bilateral sequential Adie's tonic pupil

as well, with the postganglionic parasympathetic fibres along their course in the suprachoroidal space as the probable site of disturbance. Damage to the short ciliary nerves, branches of the ciliary ganglion carrying parasympathetic impulses to the iris sphincter muscle and ciliary body, and subsequent aberrant regeneration is the primary mechanism in the development of tonic pupil. These nerves pierce the sclera in the posterior aspects of the globe. There was no evidence of choroidal ischaemia in this patient's initial evaluation, although follow-up perfusion studies were not conducted. Though rare, there exist cases of tonic pupil in the literature in patients with extensive chorioretinal disturbance.⁶ Although it was possible that our patient developed Adie's tonic pupil independent of her sclerochoroidal calcification, this was unlikely given the sequential development of bilateral tonic pupil in context of this patient's rapid hyperopic shift and bilateral dystrophic calcifications. Furthermore, our patient had no history of eye trauma or recent infections and workup for more common aetiologies (i.e. diabetes, syphilis, temporal arteritis, systemic inflammatory disorders) was negative.

We believe that ophthalmologists should be aware that sclerochoroidal calcification, a relatively "benign" condition, can be associated with progressive hyperopic shift and, rarely, tonic pupil. In patients above the age of 60 who present with tonic pupil, prompt workup is required to rule out common aetiologies, including giant cell arteritis.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Note: Figure 1 of this article is available in colour online at <http://informahealthcare.com/oph>.

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