A rare ophthalmic condition associated with primary hyperparathyroidism (PHPT): sclerochoroidal calcification (SC)

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Summary

An 82-year-old male with a proven diagnosis of primary hyperparathyroidism (PHPT) was found to have bilateral changes in the fundi during a routine eye examination which were consistent with SC. In this report, we discuss the link between SC and PHPT and question the need for prospective observational studies to establish the true association between these conditions. Though screening PHPT patients for SC might not be justified/warranted given the benign course of the latter, patients with SC need to be assessed for PHPT, as the former may be the first clue to an underlying treatable systemic disease.

Learning points:

- Sclerochoroidal calcifications (SCs), though rare and harmless, could be associated with an underlying systemic disease, such as primary hyperparathyroidism (PHPT).
- Biochemical screening for hypercalcaemia is a simple, cheap and widely available tool that could facilitate an identification of undiagnosed PHPT in patients with SC.
- A joint care by endocrinologists and ophthalmologists is warranted for those patients, as thorough investigations and long-term follow-up plans are crucial.

Background

Sclerochoroidal calcification (SC) is an uncommon asymptomatic condition that is often discovered as an incidental finding during a routine eye examination. It is ordinarily believed to be idiopathic but can be the first hint to an underlying systemic metabolic disorder such as primary hyperparathyroidism (PHPT). Recognition of SC needs to be followed by careful screening for any systemic diseases which sometimes prove curable or at least, controllable, in the long term.

Case presentation

An 82-year-old patient was referred to the endocrinology clinic due to hypercalcaemia. His past medical history included dyslipidaemia, osteoarthritis and cognitive impairment. He lived alone, and his medications included atorvastatin, paracetamol and donepezil. He had no personal history of kidney stones or any fractures. His endocrine evaluation confirmed the diagnosis of biochemical PHPT (Fig. 1) with subsequent localisation of a 7×6mm left inferior parathyroid adenoma by

Figure 1
Right eye fundi image.

ultrasonography, high-resolution computed tomography (CT) and technetium sestamibi scan of the neck. An ultrasound of the abdomen showed no nephrocalcinosis but a bone densitometry scan confirmed a diagnosis of osteoporosis which was mostly affecting the distal radius bilaterally. Given the patients other morbidities, his PHPT was treated conservatively with monitoring of his biochemistry after a multidisciplinary team discussion. Bisphosphonates therapy was added with a prospective plan to initiate calcimimetics if his hypercalcaemia to worsen in the future.

Eight months after his PHPT diagnosis, he was referred by the optometrist to the ophthalmology department after noticing asymptomatic pale lesions in both fundi. The appearance was deemed classical of SC, which were likely to be related to PHPT-induced hypercalcaemia.

Right and left eye fundi images are given in Figs 1 and 2.

Figure 2
Left eye fundi image.

Table 1 Biochemistry results table.

Parameter	Value
Corrected calcium	2.83 mmol/L (2.20–2.60)
Parathyroid hormone	13.9 pmol/L (1.1–6.9)
Phosphate level	0.66 mmol/L (0.8–1.5)
Vitamin D	77.3 nmol/L (50–175)
Creatinine	78 µmol/L (60–105)
eGFR	83 units (90–120)
Urinary calcium:creatinine ratio	0.2

Investigation

Ultrasonography, high-resolution CT and technetium sestamibi scan of the neck confirmed 7 × 6 mm left inferior parathyroid adenoma.

An ultrasound of the abdomen showed no nephrocalcinosis but a bone densitometry scan confirmed a diagnosis of osteoporosis which was mostly affecting the distal radius bilaterally. The biochemistry results are given in Table 1.

Auto fluorescence photographs of his right and left fundi showed areas of hyperauto fluorescence (Figs 3 and 4) corresponding with the pale lesions seen on the colour pictures (Figs 1 and 2).

The ultrasound B-scan of both eyes shows multiple hyperechoic deposits in the posterior ocular coats, persisting at low gain (Fig. 5). These are causing significant shadowing consistent with calcium deposits. The lesions appear to be posterior to the muscle insertion. There is no associated retinal detachment.

CT scan of the orbit showed bilateral SC not involving the extraocular muscles (Fig. 6).

Optical coherence tomography (OCT) scanning through the areas of calcification (here at the level of the supratemporal area of the right eye) confirms that the

Figure 3
Auto fluorescence photographs of the right fundi showed areas of hyperauto fluorescence.

Figure 4

Auto fluorescence photographs of the left fundi showed areas of hyperauto fluorescence.

abnormality is subretinal and located at the level of the choroid (Fig. 7).

Treatment

Long-term biochemical monitoring of his raised calcium was arranged by the endocrine clinic. Long-term follow-up in the ophthalmology clinic regarding the SC was not required once imaging and relevant investigations have been completed.

Outcome and follow-up

In this particular case, he is under conservative management with regular follow-up in the endocrinology clinic.

Discussion

SC is usually recognised as unilateral or bilateral yellowish placoid lesions often discovered as an incidental finding in

Figure 5

Ultrasound B-scan of both eyes shows multiple hyper echoic deposits in the posterior ocular coats, persisting at low gain.

Figure 6

CT scan of the orbit shows bilateral sclerochoroidal calcification not involving the extraocular muscles.

asymptomatic older white individuals (1). It is ordinarily believed to be idiopathic (2). However, several conditions associated with abnormal calcium-phosphorus metabolism such as PHPT can be found in patients with SC.

Other associations may include sarcoidosis, pseudohypoparathyroidism, vitamin D intoxication, hypophosphatemia and chronic renal failure.

In addition, it can be associated with primary renal tubular hypokalaemic metabolic alkalosis syndromes for example, Bartter and Gitelman syndromes (2).

The exact prevalence of SC in patients with PHPT is not clearly concluded. In one study involving 30 post-menopausal women with PHPT (3), there was no report of SC. This was a small-sized study with several limitations, and it is difficult to be sure if SC was actively looked for in those patient with PHPT. On the other hand, other studies reported a relation between SC and systemic diseases, such as PHPT. Shield *et al.* (4) described 119 patients with SCs in which 9/33 had hyperparathyroidism and 5/33 had a confirmed parathyroid adenoma. In addition to that, SCs were observed in several case series as the first observation leading to a diagnosis of an underlying hypercalcaemia and/or PHPT (5, 6, 7). Large prospective observational studies may be required to establish the true association between both conditions.

Indeed, SC can pose a clinical challenge in diagnosis as it may be misdiagnosed as a choroidal metastasis,

Figure 7

Optical coherence tomography (OCT) scanning through an area of calcification at the level of the supratemporal area of the right eye.

amelanotic choroidal nevus, melanoma or choroidal lymphoma resulting in unwarranted intervention (8). Fluorescein angiography and ultrasonography may help in confirming the diagnosis.

Finally, while it would be difficult to draw a solid conclusion if the association between PHPT and SC in this patient was causality or merely an association, this case highlights the need for collaboration between different specialties (in this case between endocrinology and ophthalmology) in approaching patients with SC and PHPT. Identifying these lesions promptly helps with the management of underlying systematic disorders. Moreover, all patients with SC should be screened for any treatable systemic associations before categorising and prognosticating the diagnosis as idiopathic (8).

Declaration of interest

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this case report.

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Patient consent

Written informed consent has been obtained from the patient.

Author contribution statement

M A, A A, S A, I M, A K and A J all were involved in the patients care and contributed equally to the production of this report. M A wrote the initial draft of this case report with the help of S A, I M and A K. The draft was revisited by A A and A J with further corrections. M A finalised the draft and submitted its final version after its review by the other authors.

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