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

Mona Abouzaid1, Ahmed Al-Sharefi1, Satish Artham1,2, Ibrahim Masri2, Aja Kotagiri2 and Ashwin Joshi1

1Department of Diabetes and Endocrinology, 2Sunderland Eye Infirmary, South Tyneside and Sunderland NHS Foundation Trust, Sunderland, UK, and 3Department of Endocrinology and Diabetes, North Tees and Hartlepool Hospitals NHS Foundation Trust, Hartlepool, UK

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
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. 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

### 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.

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.
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 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, pseudohyopoparathyroidism, 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,
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).

References

1 Lim JJ & Goldberg MF. Idiopathic sclerochoroidal calcification: case report. *Archives of Ophthalmology* 1989 107 1122–1123. (https://doi.org/10.1001/archopht.1989.01070020188015)
2 Schachat AP, Robertson DM, Mieler WF, Schwartz D, Augsburger JJ, Schatz H & Gass JD. Sclerochoroidal calcification. *Archives of Ophthalmology* 1992 110 196–199. (https://doi.org/10.1001/archopht.1992.01080140052025)
3 Pepe J, Cipriani C, Sonato C, Raimo O, Bianmonte F & Minisola S. Cardiovascular manifestations of primary hyperparathyroidism: a narrative review. *European Journal of Endocrinology* 2017 177 R297–R308. (https://doi.org/10.1530/EJE-17-0485)
4 Shields CL, Hasanreisoglu M, Saktanasate J, Shields PW, Seibel I & Shields JA. SC: clinical features, outcomes and relationship with hypercalcaemia and parathyroid adenoma in 179 eyes. *Retina* 2015 35 547–554. (https://doi.org/10.1097/IAE.0000000000000450)
5 Goldstein BG & Miller J. Metastatic calcification of the choroid in a patient with primary hyperparathyroidism. *Retina* 1982 2 76–79. (https://doi.org/10.1097/00006982-198202000-00003)
6 Cohen SY, Guyot-Sionnest M & Puech M. Choroidal neovascularization as a late complication of hyperparathyroidism. *American Journal of Ophthalmology* 1998 126 320–322. (https://doi.org/10.1016/s0002-9394(98)00160-3)
7 Kobune M, Neda H, Mogi Y, Hirayama Y, Sakamaki S, Watanabe N, Kohgo Y & Niitsu Y. Ectopic choroidal calcification of the eyes of a patient with parathyroid adenoma. *Internal Medicine* 1993 32 57–59. (https://doi.org/10.2169/internalmedicine.32.57)
8 Honavar SG, Shields CL, Demirci H & Shields JA. Sclerochoroidal calcification: clinical manifestations and systemic associations. *Archives of Ophthalmology* 2001 119 833–840. (https://doi.org/10.1001/archopht.119.6.833)