CASE REPORT

Bilateral central serous chorioretinopathy secondary to endogenous hypercortisolism in Cushing disease: a case report

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Abstract

We report on a case of bilateral exudative retinal detachment secondary to fibrinous central serous chorioretinopathy (CSC) caused by endogenous hypercortisolism in Cushing disease in a 37-year-old man. The treatment-resistant CSC along with Cushingoid features raised the suspicion of underlying systemic conditions. Further examination confirmed an adrenocorticotropic hormone–secreting pituitary adenoma (Cushing disease). The patient underwent transsphenoidal adenoma resection. Cortisol levels normalized within 1 week, and the retinal detachment resolved without recurrence. To prevent irreversible vision loss, clinicians should consider the possibility of systemic causes, particularly in bilateral CSC refractory to treatment. Multifocal leakage points on angiography are suggestive of underlying systemic etiology.

Keywords: Central serous chorioretinopathy; Cushing syndrome; Pituitary ACTH hypersecretion

Introduction

Cushing syndrome is a rare endocrine disorder characterized by chronic exposure to excess glucocorticoids secondary to either endogenous overproduction or exogenous administration. The endogenous form, with an estimated incidence of 1.2 to 2.4 per million people, is most commonly caused by an adrenocorticotropic hormone (ACTH)–secreting pituitary adenoma (Cushing disease) or autonomous cortisol secretion from adrenal tumors. Clinical manifestations include central obesity, moon face, hypertension, glucose intolerance, muscle weakness, and neuropsychiatric disturbances.

Central serous chorioretinopathy (CSC) is a vision-threatening condition characterized by serous retinal detachment. It is strongly associated with glucocorticoid excess. Exogenous steroid use is a well-known risk factor. Endogenous hypercortisolism is also a potential cause, particularly in atypical or refractory CSC cases. Up to 7.7% of patients with Cushing syndrome may develop CSC,[1] which may precede the diagnosis of Cushing syndrome. Visual loss in chronic CSC may be irreversible; early recognition of hypercortisolism with appropriate treatment is crucial. We report on a case of treatment-resistant CSC secondary to endogenous hypercortisolism in Cushing disease.

Case presentation

In July 2022, a 37-year-old man, a non-smoker, presented with a 2-to-3-month history of bilateral blurry vision. His visual acuity was 20/60 in the right eye and 20/100 in the left eye. Anterior segment examination showed normal intraocular pressure. Fundal examination revealed bilateral inferior bullous exudative retinal detachment and subretinal fibrotic changes in the macula, with no signs of intraocular inflammation or optic nerve anomalies such as optic disc pits (**Figure 1**). Optical coherence tomography confirmed the presence of subretinal fluid, subretinal fibrin, serous pigment epithelial detachments, and a thickened choroid with

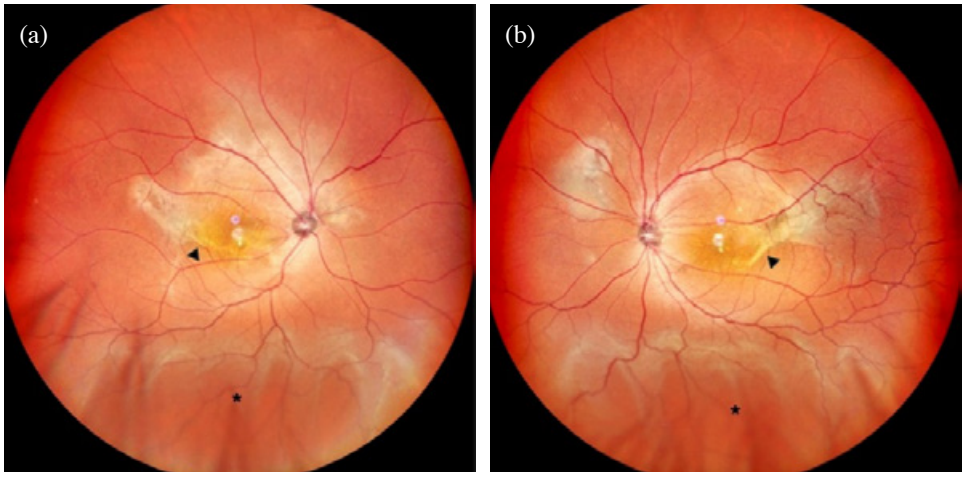

Figure 1. Colour fundus photographs of the (a) right eye and (b) left eye showing bilateral inferior bullous exudative retinal detachment (asterisks) and subretinal fibrin in the macular region (arrowheads).

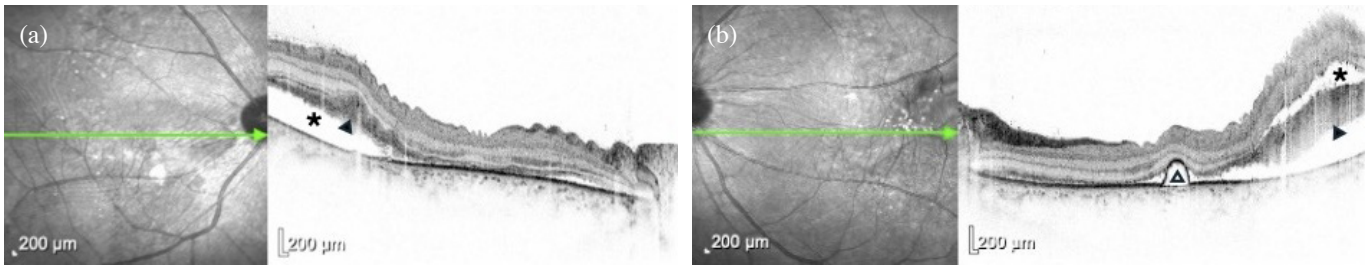

Figure 2. Optical coherence tomography of the (a) right eye and (b) left eye showing subretinal fluid (asterisks), subretinal hyperreflective fibrinous materials (black arrowheads), and serous pigment epithelial detachments (white arrowhead).

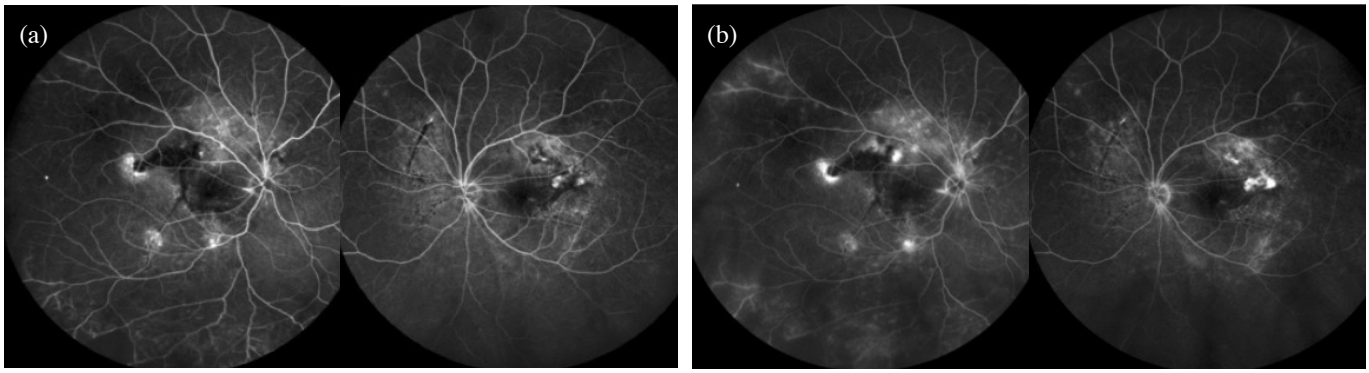

Figure 3. (a) Early- and (b) mid-phase fundus fluorescein angiography of the right and left eyes showing multiple focal leakage points in the retina.

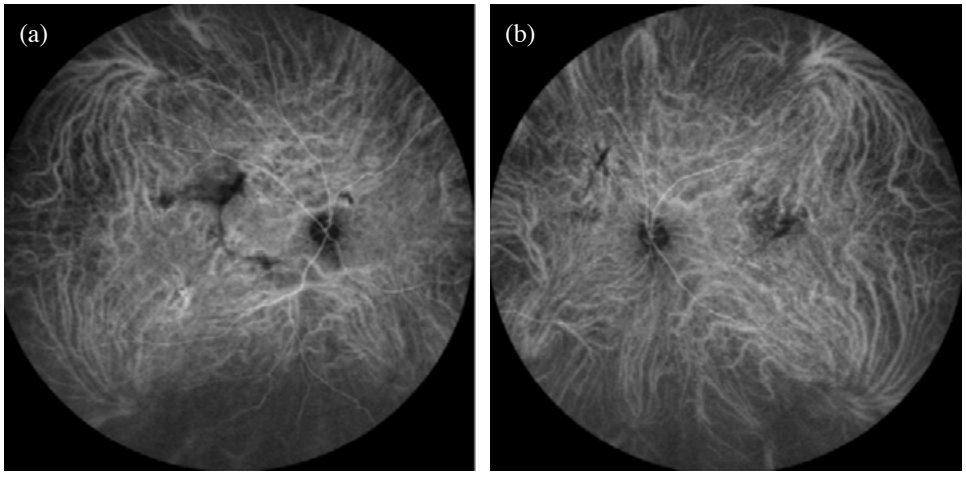

Figure 4. Mid-phase indocyanine green angiography of the (a) right eye and (b) left eye showing choroidal hyperpermeability with dilated choroidal vessels and hypocyanescence secondary to blockage by the subretinal fibrin.

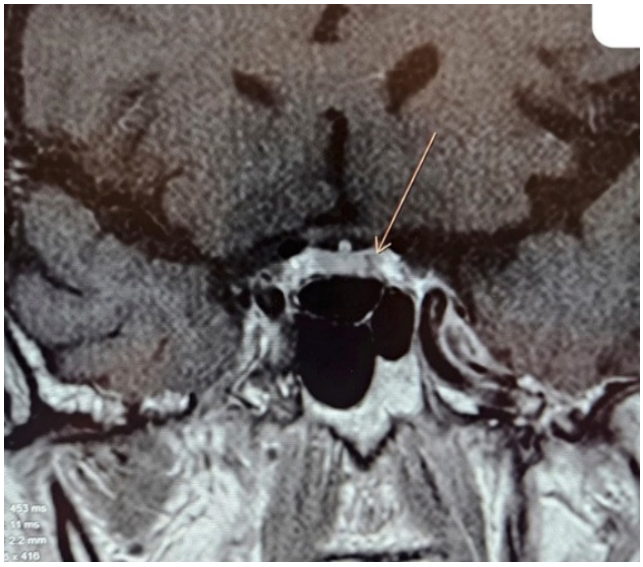

Figure 5. Coronal contrast-enhanced T1-weighted magnetic resonance imaging of the brain showing a non-discrete hypointense area in the pituitary gland suspicious of a microadenoma (arrow).

pachyvessels (**Figure 2**). Fundus fluorescein angiography demonstrated multiple inkblot leakage points (**Figure 3**), and indocyanine green angiography demonstrated choroidal hyperpermeability with dilated choroidal vessels and hypocyanescence secondary to blockage by the subretinal fibrin (**Figure 4**). The working diagnosis was bilateral exudative retinal detachment secondary to CSC.

Both eyes were treated with half-dose photodynamic therapy and sessions of micropulse laser over a course of 8 months. The subretinal fluid in the left eye resolved, but serous macular detachment in the right eye persisted. Further examination revealed Cushingoid features including moon face, hirsutism, central obesity, and proximal muscle weakness. Overnight dexamethasone suppression test confirmed endogenous hypercortisolism. Magnetic resonance imaging of the brain identified a non-discrete hypointense area suspicious of a pituitary microadenoma (**Figure 5**). A corticotrophin-releasing hormone stimulation test confirmed the endogenous hypercortisolism of pituitary origin.

The patient underwent endoscopic transsphenoidal resection of a pituitary microadenoma. Immunostaining confirmed an ACTH-secreting microadenoma. The early morning blood cortisol level normalized within 1 week, and the serous macular detachment resolved 2 months later, with bilateral atrophic maculae (**Figure 6**). At 1-year follow-up, there were no signs of CSC recurrence. The final visual acuity was 0.3 in the right eye and 0.2 in the left eye.

Discussion

The etiology of CSC is likely multifactorial. Glucocorticoid exposure is a risk factor and can alter choriocapillaris permeability and fragility, resulting in choroidal circulation decompensation and subretinal fluid accumulation.[2] Both exogenous and endogenous glucocorticoids are predisposing factors for CSC. Endogenous hypercortisolism is a rare condition with an estimated incidence ranging from 1.2 to 2.4 per million people.[3] It can be caused by an ACTH-dependent mass, which is most commonly an ACTH-secreting pituitary adenoma. It can also be caused by ACTH-independent adrenal neoplasms. In patients with endogenous hypercortisolism, CSC is a rare ophthalmic complication.[4] In our patient, the atypical refractory CSC prompted additional examination that eventually led to the diagnosis of Cushing disease.

To prevent irreversible vision loss secondary to macular neovascularization and outer retinal degeneration, clinicians should consider the possibility of systemic causes, particularly in bilateral CSC refractory to treatment. Multifocal leakage points on angiography are suggestive of underlying systemic etiology.

Contributors

All authors designed the study, acquired the data, analyzed the data, drafted the manuscript, and critically revised the manuscript for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

As an editor of the journal, SKHS was not involved in the peer review process. Other authors have disclosed no conflicts of interest.

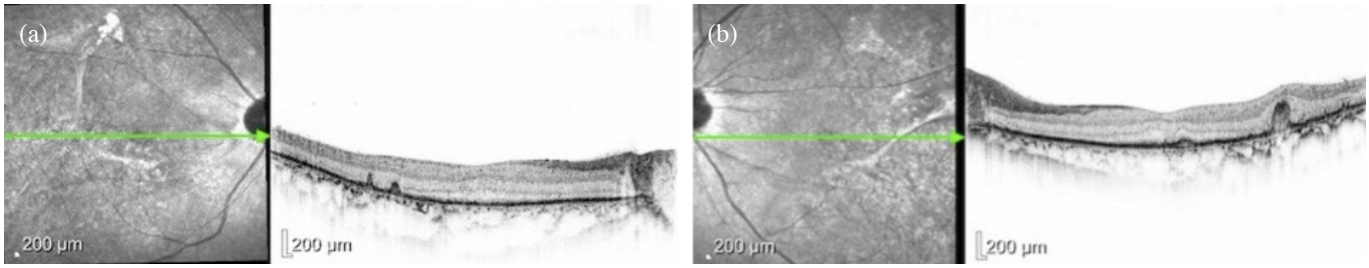

Figure 6. Optical coherence tomography of the (a) right eye and (b) left eye showing resolution of subretinal fluid after endoscopic transsphenoidal resection of the pituitary microadenoma.

Funding/support

This study received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Data availability

All data analyzed in this study are available from the corresponding author upon reasonable request.

Ethics approval

The patient was treated in accordance with the tenets of the Declaration of Helsinki. The patient provided written informed consent for all treatment and procedures and for publication.

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