Clinical Features

VKH disease is a bilateral granulomatous panuveitis often associated with exudative retinal detachment and extraocular manifestations, such as meningismus, vitiligo, poliosis, alopecia, and dysacusis. To diagnose this disease, any history of trauma or ocular surgery must be excluded. VKH disease could be staged as the prodromal, acute uveitic, chronic uveitic, and chronic recurrent stages. In the prodromal stage, a non-specific viral-like illness could appear and cerebrospinal fluid analysis usually reveals pleocytosis. The vision is blurred at the acute uveitic stage and uveitis is characterized as thickening of the posterior choroid with multiple serous retinal detachments, optic disc hyperemia, and edema. In the chronic stage, cutaneous manifestations such as vitiligo and poliosis develop and choroidal depigmentation, known as sunset glow fundus, occurs. Extensive chorioretinal atrophy and glaucoma, subretinal neovascular membrane could appear in the chronic recurrent stage. The diagnosis of VKH is a clinical one in most cases, but in some atypical cases, FA is essential for the diagnosis. ICGA is also useful to evaluate choroid inflammatory changes. The treatment of VKH is early use of high-dose systemic corticosteroids followed by a gradual tapering over 3–6 months.

CSC is a chorioretinal disorder characterized by serous retinal detachment usually located in the posterior pole. The pathogenesis of CSC is still unclear. However, it has been thought to have a multifactorial etiology and be associated with widespread choroidal circulation abnormalities. Particularly, it the atypical form of CSC is known to cause multifocal neurosensory retinal detachment and be closely correlated with steroid use. In the clinical field, atypical CSC could be misdiagnosed as choroiditis or VKH disease, both of which conditions require steroid treatment. Therefore, proper differential diagnosis is critical, because atypical CSC could be aggravated by steroid treatment.

Case Presentations

Case 1

A 52-year-old male visited our clinic complaining of blurred vision in both eyes for 2 days. He also complained of headache and neck pain. Vision was 0.63 in each eye. No sign of inflammation could be found in the anterior chamber and vitreous. Fundus examination showed optic disc edema in both eyes, but otherwise a normal retina (Fig. 12-5-1). The results of visual field and color vision tests were normal. Brain magnetic resonance imaging (MRI) showed no abnormality along the optic nerve pathway or in the brain parenchyma.
Two weeks later, vision had decreased to 0.3 in each eye. Slitlamp examination showed cells and fibrinoid materials in the anterior chamber. Fundus examination showed exacerbated optic disc edema and subretinal fluid (Fig. 12-5-2). OCT revealed multiple dome-shaped serous retinal detachments with choroidal folds (Fig. 12-5-3). There were multiple pinpoint leakages and fluorescein pooling on FA (Fig. 12-5-4).

Fig. 12-5-1 Fundus images at the initial visit showing disc edema and a virtually normal retina in both eyes.

Fig. 12-5-2 2 weeks after the initial presentation, fundus examination revealed exacerbated optic disc edema and subretinal fluid in both eyes.

Fig. 12-5-3 OCT of the right (A) and left (B) eyes shows multiple dome-shaped serous retinal detachments with choroidal folds.
He was then diagnosed as VKH disease. Spinal fluid examination showed lymphocyte dominant pleocytosis, which supported the diagnosis of VKH disease. After intensive therapy with intravenous and oral steroid, the subretinal fluid and disc edema were resolved.

**Case 2**

A 51-year-old female presented with sudden onset of visual disturbance in the left eye for 2 days. She had no specific systemic illness. Visual acuity was 0.6 in the right eye and 0.4 in the left eye. Fundus examination demonstrated bilateral mild disc hyperemia with serous macular detachment in the left eye (Fig. 12-5-5A,B). FA revealed diffuse leakage in the early and late phases. Several pinpoint hyperfluorescences and disc leakages were also seen (Fig. 12-5-5C,D).

VKH disease was suspected and she underwent intravenous steroid pulse therapy. As there were no changes in the amount of subretinal fluid during pulse therapy, intravenous steroid was switched to oral prednisolone, which was tapered over the following weeks.
At 2 weeks, the amount of subretinal fluid increased in the left eye and new serous retinal detachment was found in the right eye (Fig. 12-5-6A). FA findings did not confirm VKH disease (Fig. 12-5-6B). OCT displayed serous macular detachment in both eyes (Fig. 12-5-6C). The clinical diagnosis was revised to atypical CSC, and steroid medication was stopped promptly to prevent further progression.

However, serous detachment progressed to bullous serous retinal detachment at 5 weeks (Fig. 15-5-7A). FA demonstrated multiple pinpoint leakages and late pooling consistent with VKH disease (Fig. 15-5-7B), and OCT revealed bullous macular detachment in both eyes (Fig. 12-5-7C). Steroid pulse therapy was prescribed again, and the subretinal fluid was resolved rapidly with visual improvement (Fig. 12-5-8). Visual acuity was recovered to 0.5 in the right eye and 1.0 in the left eye after treatment.

Fig. 12-5-5 Fundus image (A) and OCT (B) at presentation showing serous retinal detachment in the macula of the left eye. Diffuse leakage is noted in the early and late phases of FA (C and D). Several pinpoint hyperfluorescences and disc leakages are also seen.
Fig. 12-5-6  (A) 2 weeks after onset, there was new serous macular detachment observed in the right eye without improvement of the left eye. (B) FA demonstrates diffuse hyperfluorescence in the macula and disc (yellow arrows) of both eyes. Several pinpoint leakages are seen only in the right eye. (C) OCT displays serous macular detachment with choroidal folds (white arrows) in both eyes.
Misdiagnosed Vogt-Koyanagi-Harada (VKH) disease and atypical central serous chorioretinopathy (CSC)

Fig. 12-5-7 (A) At 5 weeks, serous macular detachment was aggravated in both eyes. (B) FA shows pooling of dye from multiple leaking points in the late phase. (C) OCT discloses bullous serous macular detachment with choroidal folds.

Fig. 12-5-8 Fundus images (A) and OCT (B) demonstrating complete resolution of serous macular detachment after systemic steroid therapy.
Case 3

A 48–year-old male visited the clinic because of a sudden onset of visual disturbance in the left eye for 20 days. He had no medical history of illness. Before being referred to a retinal consultant, he had visited a general ophthalmology clinic and been prescribed oral steroid under the impression of VKH disease. Visual acuity was 0.8 in the right eye and 0.6 in the left eye. The anterior chamber and the vitreous were normal. Fundus examination revealed multiple pigment epithelial detachments (PEDs) in both eyes and serous macular detachment in the left eye (Fig. 12-5-9). FA confirmed PEDs around the macula in both eyes and also revealed leakage with a smokestack appearance in the left eye (Fig. 12-5-10). OCT showed multiple PEDs in both eyes and serous retinal detachment on the macula in the left eye (Fig. 12-5-11).

Under the impression of atypical CSC, oral steroid was discontinued and he received two intravitreal bevacizumab injections (1.25mg) at a 6-week interval. After bevacizumab treatment, serous macular detachment was resolved and vision improved to 0.8 in the left eye.

![Image](Fig. 12-5-9) Fundus images showing some decompensated RPE and multiple PEDs (black arrows) in both eyes. Serous retinal detachment (white arrows) at the macula was also noted in the left eye (B).

![Image](Fig. 12-5-10) FA of the right (A) and the left eyes (B and C) demonstrating multiple PEDs around the macula. A smokestack appearance of leakage is noted at the temporal side of the fovea in the left eye.
Summary and Key points

Patients with VKH disease usually have bilateral granulomatous panuveitis associated with signs of meningeal irritation. Optic disc hyperemia and edema can often be seen in VKH, and optic nerve head changes with little or no exudative retinal detachment in the early stage of VKH could be mistaken for optic neuropathy such as bilateral optic neuritis. Evaluation for optic nerve function, which would be normal in VKH, signs for meningismus, and associated intraocular inflammation may be clues for the diagnosis of VKH.

Atypical forms of CSC must be distinguished from VKH disease, in order to avoid the inappropriate use of systemic corticosteroids that can cause worsening of CSC. In the early phase of VKH disease with severe inflammatory reaction, rapidly deteriorating clinical features may not respond well to steroid therapy. This may lead to a misdiagnosis as atypical CSC. Recent studies suggested that several OCT findings, including choroidal folds and subretinal septa, could be simple and effective differential clues to help a diagnosis of VKH disease. Fluorescein staining or leakage of the optic disc in FA also can be observed in VKH disease but not in CSC. Although these findings are not confirmative, they are helpful in differentiating VKH disease from other confounding conditions.

**Differential Diagnoses**

- Choroiditis
- Sympathetic ophthalmia
- Posterior scleritis
- Uveal effusion syndrome
- Hypertensive retinopathy

Case 1 presented by Jung Hyun Park
Case 2 presented by Ji Eun Lee
Case 3 presented by Ji Hun Song

**Fig. 12-5-11** (A) OCT of the left eye shows the presence of subretinal fluid and PEDs at presentation. (B) After bevacizumab treatment, the subretinal fluid was markedly decreased with vision improvement.
References