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Letter to the Editor

Vogt-Koyanagi-Harada disease – A diagnostic pitfall for neurologists

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Dear Sirs,

A 34-year-old woman presented with headache, nausea and transient visual obscuration in both eyes for two weeks. At admission, the blood pressure was 125/75 mmHg and body weight was 55 kg (BMI of 20.6 kg/m^2). The patient described throbbing headache in association with photophobia and phonophobia, which was severe enough to awaken her during night. The patient had no prior history of ocular trauma or surgery. The patient denied tinnitus, ear fullness, or hearing loss. CT scan of the brain elsewhere was normal.

Her best-corrected visual acuity was 20/32 in the right, and 20/40 in the left eye. She did not show relative afferent pupillary defect (RAPD) or light-near dissociation (LND) in either eye. A prominent optic disc swelling was observed in both eyes (Frisen scale 3) with direct funduscopy. The patient also showed Kernig's and Brudzinski's signs. Visual fields were normal on the standard automated perimetry.

Under a suspicion of papilledema, she underwent emergent brain imaging and neuro-ophthalmologic evaluation. However, brain MRI and MR venography were unrevealing. Pure tone audiometry was also normal. The CSF opening pressure was 6.5 cmH_2O. CSF analyses showed pleocytosis of 110/mm^3 (99% of lymphocytes), protein of 24.6 mg/dL, and glucose of 94 mg/dL. Fundus photography revealed optic disc edema and multiple serous retinal detachment in both eyes (Fig. 1A). Optical coherence tomography (OCT) demonstrated multilobar serous peripapillary retinal detachments, subretinal fluid collections, and a membranous structure separating the cystoid space from subretinal fluid (Fig. 1B), which are common features of Vogt-Koyanagi-Harada disease (VKH). Fluorescein angiography revealed numerous points of leakage in the retinal pigment epithelium (Fig. 1C). Serologic tests for viral and autoimmune diseases were all negative including venereal disease research laboratory, herpes zoster, cytomegalovirus, enterovirus, rheumatoid factor, lupus anticoagulant, angiotensin-converting enzyme, HLA—B27, HLA—B51, anti-neutrophil cytoplasmic, anti-nuclear, paraneoplastic, anti-myelin oligodendrocyte glycoprotein and anti-aquaporin 4 antibodies. She was subjected to 1 g per day of intravenous methylprednisolone for five consecutive days and 2 mg per day of tacrolimus for nine months. Three months later, the visual acuity became 20/20 in both eyes as well as normalization of the optic disc swelling and subretinal fluid collection (Fig. 1D and 1E). In the meanwhile, there was no vitiligo, alopecia, and poliosis during the follow-up.

VKH refers to an inflammatory disorder that typically presents with bilateral choroiditis and exudative retinal detachment [1]. The pathogenesis of VKH is thought to be related to an aberrant T-cell mediated immune response directed against a self-antigen found on the melanocytes [2]. The diagnosis is usually made by ophthalmologists since the granulomatous uveitis and serious retinal detachments are the most common findings [2]. When encountered during the earliest stage, however, VKH may avoid detection since it mostly manifests with systemic symptoms such as fever, headache, or meningismus [2]. Besides, likewise in our patient, the subretinal fluid collection and choroiditis, which are the characteristic findings of VKH, can be easily missed with direct funduscopy [3].

VKH is known to be associated with disc edema in 4–28% during the early phase [4,5]. Given the relative preservation of visual acuity and field, as well as the absence of RAPD or LND, the disc edema was ascribed to papilledema in our patient. Although papilledema can be distinguished from optic neuropathy by bilateral involvement and relative preservation of visual function, the distinction often can be obscure [6]. Instead, optic disc edema in VKH is ascribed to severe choroidal inflammation causing axonal flow stasis and secondary axonal swelling of the optic nerve [5].

As brain imaging did not reveal any intracranial mass responsible for increased intracranial pressure (ICP), our patient had an initial presumptive diagnosis of idiopathic intracranial hypertension (IIH). However, CSF pressure was normal, and there was no other radiologic evidence of elevated ICP [7]. Besides, since more than 90% of IIH typically affects overweight women [8], normal BMI also stands against the diagnosis of IIH. Likewise, various infectious, inflammatory or metastatic diseases can also give rise to bilateral granulomatous uveitis that includes sympathetic ophthalmia, sarcoidosis, systemic lupus erythematosus, giant cell arteritis, lymphoma, or syphilis, which all came out to be negative in our patient though extensive serologic and neuro-ophthalmologic evaluations.

Our patient emphasizes a scrutinized evaluation of the retina during the fundoscopy in patients presenting with headache and disc edema, since it allows detection of VKH, which can be a diagnostic pitfall for neurologist.

Abbreviations: RAPD, relative afferent pupillary defect; VKH, Vogt-Koyanagi-Harada disease; LND, Light-near dissociation; IIH, idiopathic intracranial hypertension.

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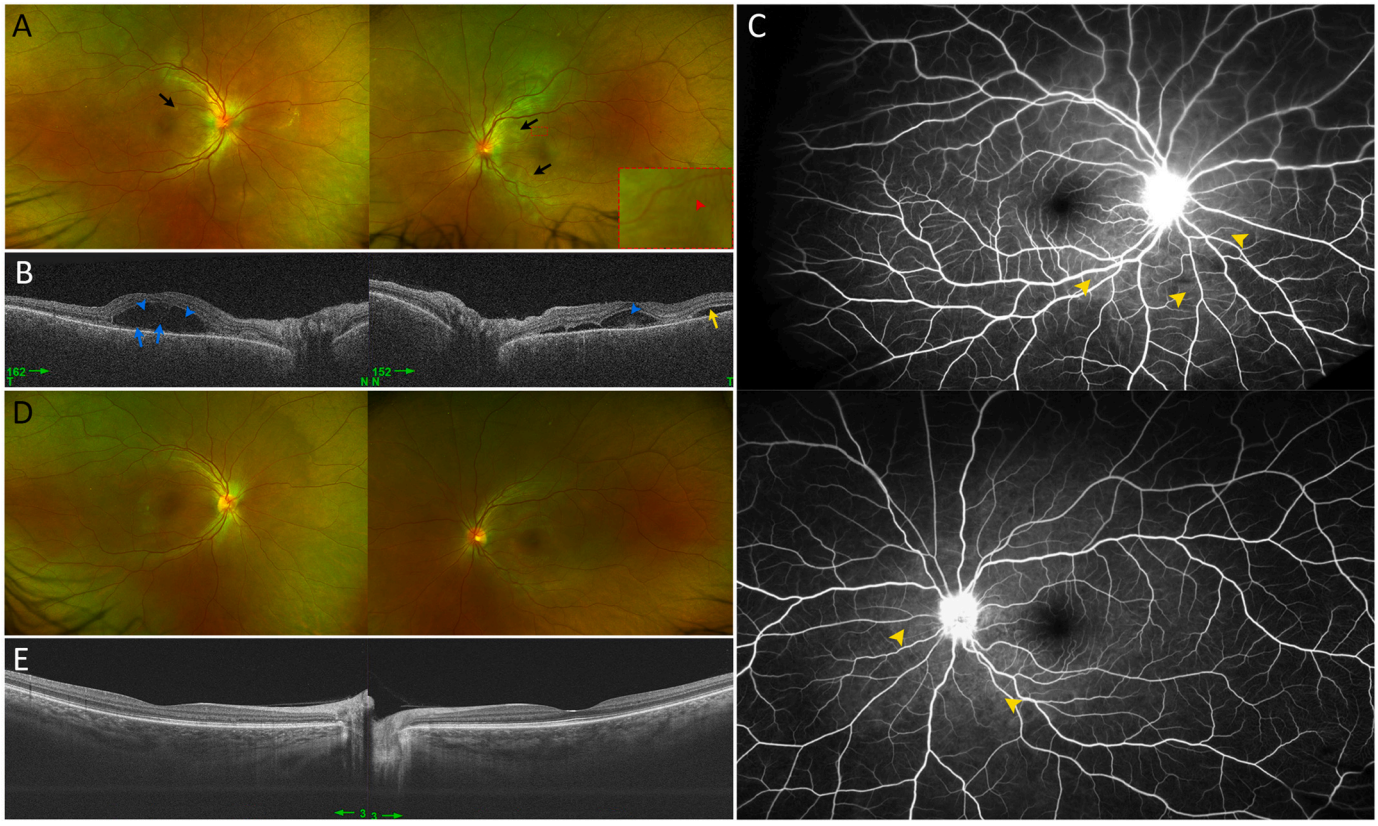

Fig. 1. A. Fundus photography shows optic disc edema and multifocal hypopigmented choroditis patches (black arrows) with choroidal folds (red arrow in the red inset) along the inferior arcade in both eyes. B. Optical coherence tomography (OCT) discloses multilobar serous peripapillary retinal detachments, subretinal fluid collection (yellow arrow) and a membranous structure (blue arrows) separating the cystoid space (blue arrowheads) from subretinal fluid C. Fluorescein angiography shows disc hyperemia and hyperfluorescent leakage at the retinal pigment epithelium level during the arteriovenous phase (yellow arrowheads). D-E. The optic disc swelling and subretinal fluid collection disappear on fundus photography and OCT three months later. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

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