scleritis is achieved through observation of sun set glow fundus and neurologic or dermatologic signs not seen in posterior scleritis. Posterior scleritis is also usually unilateral, and T-sign is a unique characteristic of this disease. Recently, there have been some reports, especially in Japan, of posterior scleritis appearing concurrently with VKH disease in patients; Kouda et al. [4] reported that posterior scleritis was an early manifestation of VKH.

In our case, neurologic symptoms such as headache and tinnitus appeared as clinical features of VKH, while T-sign and lid swelling were signs of posterior scleritis. Anterior chamber reaction and serous retinal detachment were also observed in both VKH and posterior scleritis. Thus, we conclude that our case involved unilateral VKH disease with posterior scleritis.

Su Young Moon, Won Tae Yoon, Sung Pyo Park
Department of Ophthalmology, Kangdong Sacred Heart Hospital, Hallym University College of Medicine, Seoul, Korea
E-mail (Sung Pyo Park): sungpyo@hanafos.com

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Intravitreal Bevacizumab for the Treatment of Optic Disc Edema in a Patient with POEMS Syndrome

Dear Editor,

Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome is a rare multisystem disorder of obscure etiology that is associated with plasma cell dyscrasia [1]. Optic disc edema (ODE) has been reported to be a common ocular manifestation of POEMS syndrome [2]. Vascular endothelial growth factor (VEGF) has been regarded to have an important role in the pathophysiology of POEMS syndrome. We present a case of a young female patient presenting with bilateral optic nerve head edema after being diagnosed with POEMS syndrome, who was successfully treated with repeated intravitreal bevacizumab injections. The study protocol was approved by the Institutional Review Board of Yonsei University Severance Hospital and followed the tenets of the Declaration of Helsinki.

A 34-year-old female patient presented with progressive narrowing of her visual field (VF). She had been diagnosed with POEMS syndrome 3 months previously at the department of hemato-oncology and had received palliative radiotherapy of 5,000 cGy to her right iliac crest, in which a plasmacytoma was located. She was also prescribed thalidomide 100 mg/day orally. According to her medical records, she had bilateral ODE at the time of diagnosis. At that time, lumbar puncture had revealed a slightly increased intracranial pressure of 16.18 mmHg. Her best-corrected visual acuity and intraocular pressure were 20 / 25 and 18 mmHg, respectively. There were no abnormal findings in either eye, with the exception of severe bilateral ODE.

Automated perimetry showed bilateral enlarged blind spots and VF constriction. A fluorescein angiogram revealed early, well delineated hyperfluorescence in both optic discs, compatible with ODE (Fig. 1A-1H). Indocyanine green angiography revealed no abnormalities of choroidal perfusion.

Although her central vision was largely unaffected, the

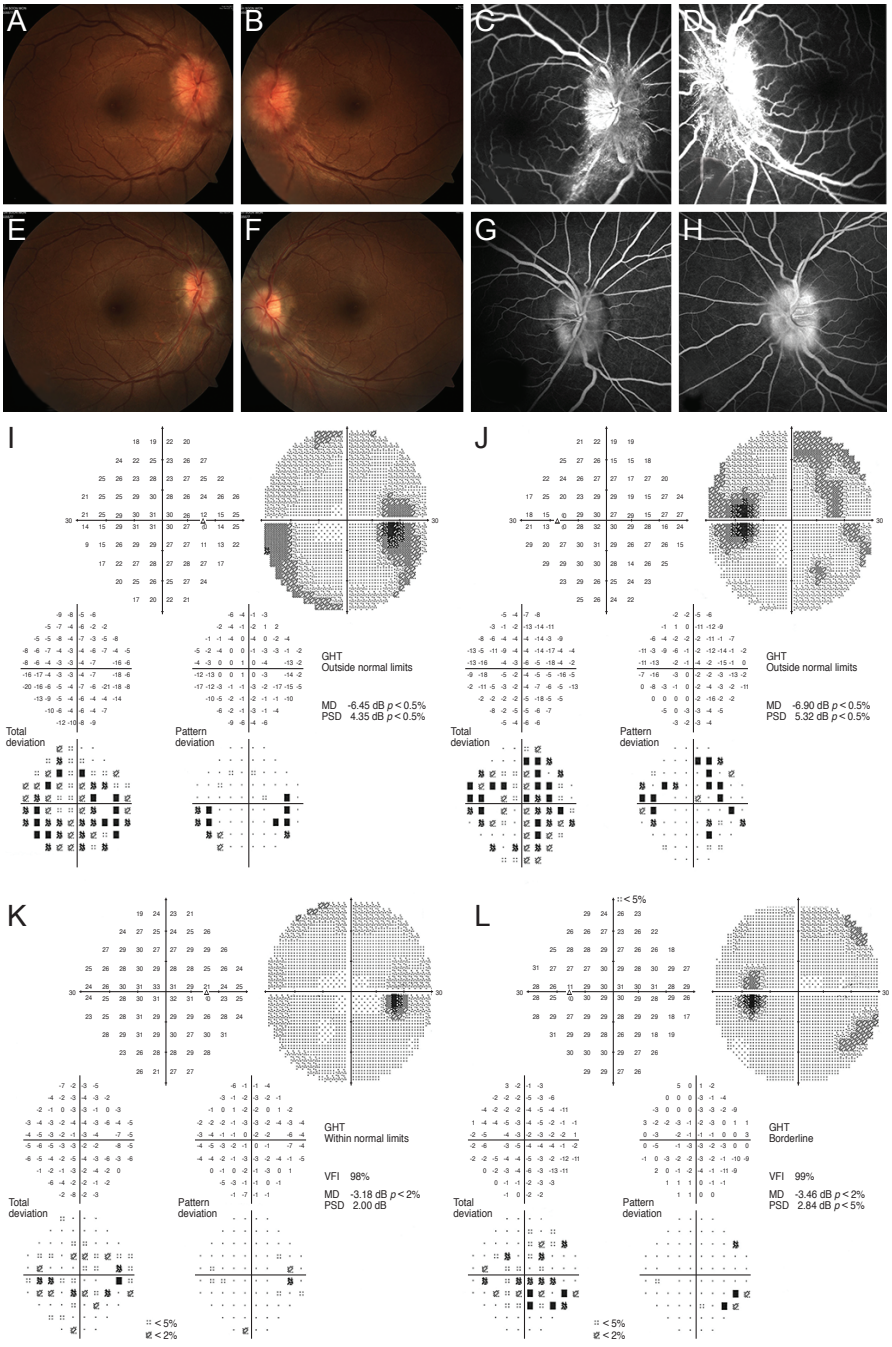

Fig. 1. Fundus photographs, fluorescein angiograms, and Humphrey automated perimetry (HAP) before and 3 months after the last intravitral bevacizumab Injection. (A,B) Bilateral fundus photographs showing optic nerve head edema (ODE). The disc is congested and hyperemic with elevation of the retinal surface. Signs of secondary gliosis due to long term ODE are evident (fundus camera; Canon, Tokyo, Japan) (A, right eye; B, left eye). (C,D) Bilateral fluorescein angiograms and indocyanine green angiograms showing early irregular hyperfluorescence emanating from the disc and spreading to the proximal vascular arcades. Long standing fibrotic change, probably due to secondary gliosis, is also present. Indocyanine green angiograms A shows mild blocked-fluorescence at the disc with no abnormal choroidal perfusion (0:58.23, 30 degrees; Heidelberg Retinal Angiogram 2, Heidelberg Engineering, Heidelberg, Germany) (C, right eye; D, left eye). (E,F) Bilateral fundus photographs 3 months after the last intravitreal bevacizumab injections. Resolution of the prior ODE is evident (E, right eye; F, left eye). (G,H) Bilateral fluorescein angiograms of the same patient 3 months after the last injection of intravitreal bevacizumab. The disc is only mildly stained in the late phase, with no evidence of active leakage (20:04.09) (G, right eye; H, left eye). (I,J) Pre-injection HAP showing enlarged inferior blind spots and superior arcuate visual field defects developing in the right (I) and left (J) eyes, respectively. (K,L) HAP three months after the last intravitreal bevacizumab injection showing resolution of the previous scotomata with a small nasal step remaining in the left eye (K, right eye; L, left eye).

patient complained of progressively enlarging VF defects, which was confirmed by automated perimetry. The worsening VF defects coupled with the presence of ODE following the completion of radiotherapy warranted further intervention. Bilateral intravitreal bevacizumab injections were performed after informed consent had been obtained. Quantification of the intravitreal VEGF at the time of injection was within the established normal range (<30 pg/mL); however, the systemic VEGF level was elevated at 166.0 pg/mL (normal, 0 to 38.3 pg/mL).

One week after the intravitreal injection of bevacizumab, repeat fluorescein angiogram and fundus photography revealed bilateral regression of the ODE accompanied by improvement in the VF. Approximately 50 days later, the patient complained of recurring VF defects, and increasing ODE was observed in both eyes. Repeat bilateral injections were administered with improvement in both the symptoms and ODE (Fig. 1I-1L). At follow-up 11 months after the injections, the patient had 20 / 20 vision with complete regression of the ODE, likely due to systemic improvement.

The presence of ODE in POEMS syndrome has been well documented [2]. However, its etiology remains a matter of controversy. Increased systemic VEGF [3], increased intracranial pressure [4], nerve infiltration [5], and vascular hyperpermeability [4] have been suggested as possible causes.

The patient presented here showed an initial improvement in symptomatic ODE after intravitreal bevacizumab injection; however, the ODE redeveloped approximately 50 days later. This timeline coincides with the known duration of anti-VEGF antibody. The successful management of ODE with anti-VEGF therapy before systemic improvement seems to imply that VEGF is an important component in the pathophysiology of ODE in POEMS syndrome. The lack of increase in VEGF titer from vitreous samples in our case coincides with data from previous report [3]. This suggests that ocular manifestations, such as ODE, are related to elevated level of systemic VEGF rather than intraocular VEGF. Systemic VEGF might alter optic nerve head vascular permeability via the choroidal blood flow, which is relatively free to communicate with the systemic circulation.

We report a case of POEMS syndrome that was treated with intravitreal bevacizumab injection. Our findings suggest that the cause of ODE in POEMS syndrome might be associated with VEGF. A relationship between the cause of ODE in POEMS syndrome and systemic VEGF should be further investigated.

Do Wook Kim
Department of Ophthalmology, Institute of Vision Research, Severance Hospital, Yonsei University College of Medicine, Seoul, Korea

Sung Yong Kang
Eyereum Ophthalmic Clinic, Seoul, Korea

Hyoung Won Bae, Samin Hong, Gong Je Seong, Chan Yun Kim
Department of Ophthalmology, Institute of Vision Research, Severance Hospital, Yonsei University College of Medicine, Seoul, Korea
E-mail (Chan Yun Kim): kcyeye@yuhs.ac

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