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Peripapillary Granuloma with Optic Nerve Head Involvement Associated with Sarcoidosis

Dear Editor,

Sarcoidosis is a systemic granulomatous disease characterized by the presence of non-caseating granulomas in involved organs. Ocular involvement is common and has been reported to occur in approximately 20% to 50% of patients with sarcoidosis [1]. Ocular manifestations include anterior uveitis, retinal periphlebitis, multifocal choroiditis, papillitis, and orbital involvement. Optic nerve involvement in sarcoidosis is known to be relatively rare and occurs in only 0.5% to 7.0% of patients with ocular sarcoids [2]. We report a case of peripapillary granuloma with optic nerve head involvement associated with sarcoidosis, which was treated with an intravitreal dexamethasone implant. Written informed consent was obtained from the patient.

A 40-year-old man presented with gradually decreased vision in his right eye for the past 1 month. Best-corrected visual acuity of the right eye was 20 / 200. Eight months prior, the patient had been diagnosed with pulmonary tuberculosis (TB) based on abnormal findings in his chest X-ray during a routine medical check-up and TB polymerase chain reaction (PCR). However, his bilateral hilar lymphadenopathy had worsened despite 6 months of treatment with anti-TB medications (Fig. 1A), which necessitated further diagnostic evaluations. Fundus examination of the right eye showed a 4DD-sized, mass-like lesion involving the inferior optic nerve head surrounded by vitreous inflammatory cells (Fig. 1B). Optical coherence tomography revealed subretinal fluid and a peripapillary hyperreflective nodule protruding into the vitreous with a disorganized retinal architecture (Fig. 1C). Fluorescein angiography of the right eye showed a mass-like lesion surrounded by telangiectatic vessels with leakage in the early phase (Fig. 1D) and late leakage from the granuloma (Fig. 1E). In addition, indocyanine green angiography revealed hypofluorescence of the lesion in both the early (Fig. 1F) and late phases (Fig. 1G). During a follow-up evaluation two weeks later, the patient tested negative for TB PCR, and his serum angiotensin-converting enzyme levels were normal at 40.3 U/mL. However, bronchoscopic biopsy and bronchoalveolar lavage revealed a non-necrotizing granuloma, which was consistent with sarcoidosis. Oral prednisolone was initiated at 1 mg/kg/day followed by slow tapering, and a sustained-delivery implant of 0.7 mg dexamethasone (Ozurdex; Allergan, Irvine, CA, USA) was injected into the vitreous cavity. The size of the granuloma and subretinal fluid were significantly reduced (Fig. 1H, 1I), and the patient's vision was improved to 20 / 60; this level was maintained at the last follow-up visit 6 months later.

In this case, the patient had a peripapillary granuloma with optic nerve head involvement associated with sarcoidosis. The diagnosis of ocular sarcoidosis is challenging due to variability in its presentation. Sarcoid granuloma should always be considered in the differential diagnosis of patients with an optic disc or peripapillary nodule [3], and a systematic approach is required to rule out major differentials and establish a diagnosis of ocular sarcoidosis. In this case, the patient had been diagnosed with pulmonary TB through PCR and presented with bilateral hilar lymphadenopathy, which worsened despite treatment with TB medications. Therefore, further investigations were performed to rule out other possible infectious causes of the nodular mass, including tuberculoma due to multidrug-resistant TB. Bronchoscopic biopsy was performed, and the diagnosis was confirmed as sarcoidosis.

The patient was treated with an intravitreal dexamethasone implant along with oral steroids. Although systemic corticosteroids are the mainstay of therapy for sarcoidosis [4], no standardized treatment protocols exist. Previous studies have suggested that an intravitreal dexamethasone implant can improve visual acuity and macular thickness in various ocular conditions, including uveitis [5], and it may also play a role as an adjuvant therapy for sarcoidosis with posterior involvement. In this case, although the size of the granuloma and subretinal fluid decreased with systemic steroid treatment, outer retinal thinning from the prolonged existence of subretinal fluid was evident on optical coherence tomography. Therefore, we attempted to

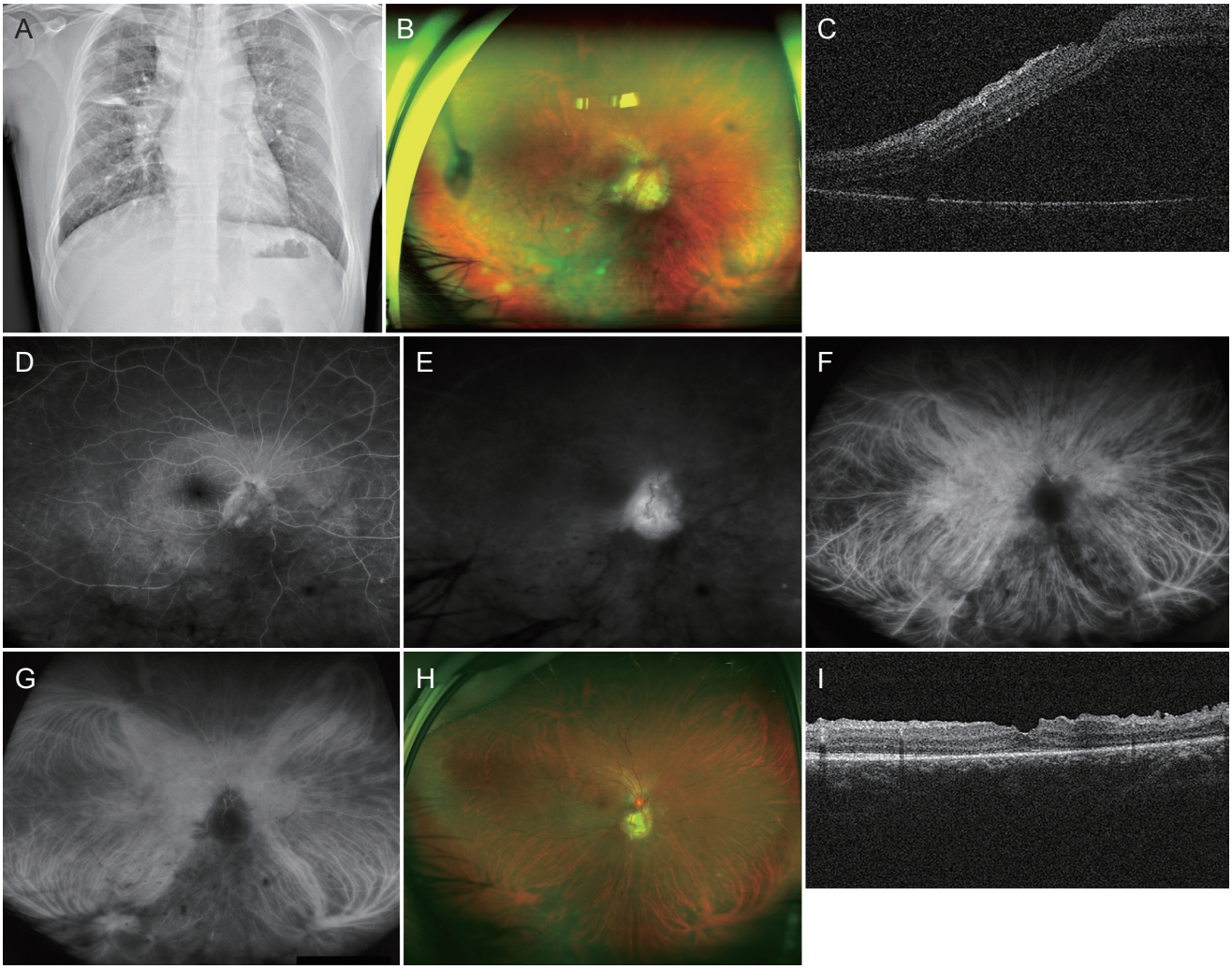

Fig. 1. A 40-year-old man with a history of tuberculosis presented for a medical evaluation after complaining of gradually decreased vision in his right eye for the past month. (A) Chest X-ray demonstrated bilateral hilar lymphadenopathy. (B) Fundus photography of the right eye at the initial visit revealed a nodular mass involving the optic nerve head surrounded by vitreous hemorrhaging and inflammatory cells. (C) Optical coherence tomography indicated subretinal fluid involving the macula with photoreceptor cell layer thinning. Fluorescein angiography showed hypofluorescence of the granuloma surrounded by tortuous vessels with (D) leakage in the early phase and (E) late leakage from the granuloma. Indocyanine green angiography revealed hypofluorescence of the lesion in both the (F) early and (G) late phases. The patient was diagnosed with sarcoidosis and treated with oral prednisolone and an intravitreal dexamethasone implant. (H) Follow-up fundus photography and (I) optical coherence tomography 6 months later demonstrated shrinkage of the granuloma and resolution of the subretinal fluid. Written informed consent was obtained from the patient.

facilitate rapid absorption of the fluid using an intravitreal dexamethasone implant. We could not conclude any additional or synergistic effects of intravitreal dexamethasone use with systemic steroids in this case, so further studies are necessary.

In conclusion, ophthalmologists should be aware that sarcoidosis may cause peripapillary granuloma with optic nerve involvement and should perform systemic examinations to establish the diagnosis. An intravitreal injection of a dexamethasone implant in conjunction with a systemic steroid may be a reasonable treatment option.

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Conflict of Interest

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

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