

Case Report

A Case of Large Sarcoid Choroidal Granuloma Treated with Steroid Pulse Therapy

Takatoshi Kobayashi^a Nanae Takai^b Takaki Sato^a
Rei Tada^c Eri Maruyama^a Hiromi Shouda^a Takako Okamoto^a
Kohichi Maruyama^d Tsunehiko Ikeda^a

^aDepartment of Ophthalmology, Osaka Medical College, Takatsuki, Japan; ^bTakai Eye Clinic, Kawachinagano, Japan; ^cTada Eye Clinic, Ikeda, Japan; ^dMaruyama Eye Clinic, Takatsuki, Japan

Keywords

Sarcoidosis · Choroidal granuloma · Steroid pulse therapy

Abstract

Purpose: To report a case of large sarcoid choroidal granuloma that was successfully treated with steroid pulse therapy. **Case Report:** A 38-year-old man presented with the primary complaint of decreased visual acuity (VA) in his left eye. Upon examination, a large white protruding lesion of 10 × 8 papilla diameter in size was observed in the macular region, and slightly temporal to it, in the patient's left eye. Whole-body contrast-enhanced computed tomography performed for differential diagnosis detected numerous enlarged lymph nodes throughout the body, including the bilateral hilar regions. Sarcoidosis was diagnosed by biopsy of the right cervical lymph nodes showing noncaseating epithelioid cell granuloma. The fundus lesion was found to be a choroidal granuloma caused by sarcoidosis, and steroid pulse therapy was started. The granuloma was considerably decreased, and the VA in the left eye improved to 0.7 after 2 months. **Conclusion:** Steroid pulse therapy was found to be effective as an initial treatment for a large sarcoid choroidal granuloma.

© 2020 The Author(s)
Published by S. Karger AG, Basel

Introduction

Sarcoidosis is an idiopathic systemic disease characterized by a noncaseating epithelioid cell granuloma lesion, and it is a major causative disease of uveitis [1]. Choroidal granuloma is a relatively rare lesion that occurs in approximately 5% of cases with uveitis lesions caused by sarcoidosis [2], whereas a large choroidal granuloma may mimic a choroidal tumor in the fundus, thus making diagnosis and treatment difficult [3]. In this study, we report a patient with a large sarcoid choroidal granuloma who was successfully treated with steroid pulse therapy.

Case Report

A 41-year-old male presented at his local eye clinic with the primary complaint of decreased visual acuity (VA) in his left eye. Upon examination, a subretinal tumor mass was observed in his left eye, and he was subsequently referred to the Department of Ophthalmology, Osaka Medical College, Takatsuki, Osaka, Japan for more detailed examination and treatment. His medical and family history was unremarkable.

Ophthalmologic Findings

His VA was RV = (1.2 × S-7.5D) and LV = (0.04 × S-8.0D), and his intraocular pressure was 17 mm Hg in both eyes. No abnormalities were noted in the right eye, but a few inflammatory cells were observed in the vitreous and anterior chamber of the left eye. Fundus examination showed no particular abnormalities in the right eye (Fig. 1a), but a large white protruding lesion of 10 papilla diameter (PD) (lateral diameter) × 8 PD (longitudinal diameter) in the macular region, and slightly temporal to it, in the left eye (Fig. 1b). B-scan ultrasonography (UD-8000; Tomey® Corp., Nagoya, Japan) was performed and revealed that the lesion in the left eye appeared to have high internal reflectivity (Fig. 2). Examination by optical coherence tomography (OCT) (SPECTRALIS®; Heidelberg Engineering GmbH, Heidelberg, Germany) revealed no abnormalities in the right eye. In the left eye, a protruding lesion was observed with a homogenous shadow on the choroid and under the retina, which was complicated by exudative retinal detachment around it (Fig. 3a, b). Fluorescein angiography (TRC-50DX Type IA; Topcon Corp., Tokyo, Japan) showed slight fluorescein leakage in the peripheral region of the right eye (Fig. 4a). In the left eye, punctate fluorescein leakage corresponding to the tumor-like mass was noted at the early phase (Fig. 4b, c), and tissue staining and fluorescein pooling were observed from the middle to late phase (Fig. 4d, e). Indocyanine green angiography (TRC-50DX Type IA) revealed low fluorescence and filling delay in the area corresponding to the tumor mass (Fig. 5a, b).

General Examination Findings

Whole-body contrast-enhanced computed tomography (CT) was performed, since the choroidal tumor was also suspected on the basis of the above findings. The CT results showed that there were enlarged lymph nodes throughout the body, including both sides of the neck, the supraclavicular fossa, both sides of the mediastinum, and the pulmonary hilum. Thus, the patient was subsequently referred to the Department of Respiratory Medicine at Osaka Medical College Hospital, and since sarcoidosis was suspected, a biopsy of the right cervical lymph nodes was performed at the Department of Plastic Surgery. The results showed a noncaseating epithelioid cell granuloma. Blood test results were as follows: the angiotensin-converting

enzyme finding was 15.2 U/L (reference range 8.3–21.4 U/L), the soluble interleukin-2 receptor finding was 987 U/mL (reference range 145–519 U/mL), the toxoplasma IgM and IgG test findings were within the normal reference range, and the enzyme-linked immune absorbent spot assay finding (tuberculosis) was negative.

Course of Treatment

Choroidal granuloma due to sarcoidosis was diagnosed on the basis of both the pathological examination results and the ophthalmologic findings. Steroid pulse therapy (methylprednisolone 1,000 mg for 3 days) was performed due to a rather large lesion compared with that of past reports. After one cycle of steroid pulse therapy, the prednisolone was tapered to 60 mg/day. Triamcinolone was once injected into the sub-Tenon capsule, but it was withdrawn due to an increase in intraocular pressure. After the initiation of steroid therapy, the choroidal granuloma gradually decreased in size, and the patient's VA gradually improved at 3 weeks and beyond after the start of treatment (Fig. 5a, b). At 2 months after the start of treatment, the choroidal granuloma was markedly decreased, the exudative retinal detachment had disappeared, and the patient's corrected VA had improved to 0.7 (Fig. 6a, b). At 1 year after the start of treatment, the choroidal granuloma had further decreased, although atrophy of the retinal pigment epithelium was detected, and the patient's corrected VA was maintained at 0.8. There was no increase in intraocular pressure during the 1-year period of steroid pulse therapy.

Discussion

The prevalence of choroidal granuloma in patients with sarcoidosis is reportedly 5%, and it commonly occurs unilaterally in the macular region or around the macula. Reportedly, there is no sex difference and it most commonly occurs in individuals 60 years of age or younger [2]. Unlike usual ocular sarcoidosis, this pathology commonly shows mild inflammation in the anterior eye or the vitreous body and often presents diagnostic difficulties. The lesion is well-defined and pale yellow, and the size ranges between 1 and 10 PD [3–5]. Fluorescein angiography examination shows low fluorescence or punctate hyperfluorescence corresponding to the lesion at the early phase and hyperfluorescence from the middle to late phase. It has been reported that indocyanine green angiography examination can show low fluorescence until the late phase [6], and that OCT findings show a homogenous shadow under the retina [7–10]. Fundus findings of this disorder mimic those of a choroidal tumor in many respects [3], and thus, a general examination including head CT/magnetic resonance imaging is important. There are many reports of sarcoidosis being diagnosed histologically, especially in order to differentiate it from a metastatic choroidal tumor or a choroidal malignant melanoma [4–6, 11]. In our present case, whole-body contrast-enhanced CT revealed enlarged lymph nodes throughout the patient's body, including both sides of the neck, the supraclavicular fossa, both sides of the mediastinum, and the pulmonary hilum. Thus, we suspected sarcoidosis, and a biopsy of the right cervical lymph nodes was performed. The results showed a non-caseating epithelioid cell granuloma, thus leading to the definitive diagnosis of sarcoidosis. All other disorders that need to be differentiated, including tuberculous uveitis, ocular toxoplasmosis, and ocular toxocariasis, were found to be serologically negative, except for ocular toxocariasis.

There have been many reports describing the use of steroid pulse therapy for sarcoidosis, including cardiac sarcoidosis [12, 13], neurosarcoidosis [14], and pulmonary sarcoidosis [15,

16]. With respect to ocular sarcoidosis, a few previous reports described the use of steroid pulse therapy for panuveitis [17] or optic neuropathy [18], yet to the best of our knowledge, there are no previously published reports on choroidal granuloma. However, Knickelbein et al. [19] reported a refractory patient with a large choroidal granuloma, panuveitis, and optic disc edema being treated with high-dose intravenous steroid therapy. A choroidal granuloma associated with sarcoidosis often responds well to oral steroid therapy, with the therapy often being started with a dose equivalent to prednisolone 20–80 mg, and then gradually tapered and discontinued in 3–10 months [6, 20, 21]. Some previous studies reported that injection of steroid into the Tenon capsule [22] or the vitreous body [23, 24] was effective.

In our present case, we selected steroid pulse therapy after obtaining the patient's consent in order to recover his VA as soon as possible, because (1) the patient had a quite thick, large choroidal granuloma of 10 × 8 PD, (2) the lesion reached a part of the macular region with highly reduced corrected VA to 0.04, and (3) the lesion was associated with exudative retinal detachment. As a result, the OCT findings showed that the choroidal granuloma began to shrink in size, and the patient's VA improved after 3 weeks of treatment. Two months later, the thickness of the choroidal granuloma had considerably decreased, the exudative retinal detachment had completely disappeared, and the corrected VA had markedly improved from 0.04 to 0.7.

In conclusion, our findings reveal that in case of a large choroidal granuloma associated with sarcoidosis, like in our present case, steroid pulse therapy may be a viable option as the initial treatment.

Acknowledgement

The authors wish to thank John Bush for editing the manuscript.

Statement of Ethics

The protocols of this study were approved by the Ethics Committee of Osaka Medical College, Takatsuki, Osaka, Japan. In accordance with the tenets set forth in the Declaration of Helsinki, prior written informed consent was obtained from the patient.

Disclosure Statement

There are no conflicts of interest to report for any of the authors.

Funding Sources

Not applicable.

Author Contributions

T. Kobayashi, N. Takai, T. Sato, E. Maruyama, H. Shouda, T. Okamoto, and K. Maruyama: equal contribution to patient management, conception of the paper, data analysis and interpretation, manuscript drafting, and literature search. R. Tada: co-writing of the manuscript and literature search. T. Ikeda: design of the paper, co-writing of the manuscript, literature search, manuscript editing and revision, and final approval. All authors read and approved the final manuscript.

References

- 1 Ma SP, Rogers SL, Hall AJ, Hodgson L, Brennan J, Stawell RJ, et al. Sarcoidosis-related Uveitis: clinical presentation, disease course, and rates of systemic disease progression after uveitis diagnosis. *Am J Ophthalmol*. 2019 Feb;198:30–6.
- 2 Obenauf CD, Shaw HE, Sydnor CF, Klintworth GK. Sarcoidosis and its ophthalmic manifestations. *Am J Ophthalmol*. 1978 Nov;86(5):648–55.
- 3 Campo RV, Aaberg TM. Choroidal granuloma in sarcoidosis. *Am J Ophthalmol*. 1984 Apr;97(4):419–27.
- 4 Letocha CE, Shields JA, Goldberg RE. Retinal changes in sarcoidosis. *Can J Ophthalmol*. 1975 Apr;10(2):184–92.
- 5 Olk RJ, Lipmann MJ, Cundiff HC, Daniels J. Solitary choroidal mass as the presenting sign in systemic sarcoidosis. *Br J Ophthalmol*. 1983 Dec;67(12):826–9.
- 6 Desai UR, Tawansy KA, Joondeph BC, Schiffman RM. Choroidal granulomas in systemic sarcoidosis. *Retina*. 2001;21(1):40–7.
- 7 Ishihara M, Shibuya E, Tanaka S, Mizuki N. Diagnostic and therapeutic evaluation of multiple choroidal granulomas in a patient with confirmed sarcoidosis using enhanced depth imaging optical coherence tomography. *Int Ophthalmol*. 2018 Dec;38(6):2603–8.
- 8 Turkoglu EB, Lally SE, Shields CL. Choroidal sarcoid granuloma simulating prostate carcinoma metastasis. *Retin Cases Brief Rep*. 2017 Winter;11 Suppl 1:S226–8.
- 9 Invernizzi A, Mapelli C, Viola F, Cigada M, Cimino L, Ratiglia R, et al. Choroidal granulomas visualized by enhanced depth imaging optical coherence tomography. *Retina*. 2015 Mar;35(3):525–31.
- 10 Rostaqui O, Querques G, Haymann P, Fardeau C, Coscas G, Souied EH. Visualization of sarcoid choroidal granuloma by enhanced depth imaging optical coherence tomography. *Ocul Immunol Inflamm*. 2014 Jun;22(3):239–41.
- 11 Tingey DP, Gonder JR. Ocular sarcoidosis presenting as a solitary choroidal mass. *Can J Ophthalmol*. 1992 Feb;27(1):25–9.
- 12 Okabe T, Yakushiji T, Hiroe M, Oyama Y, Igawa W, Ono M, et al. Steroid pulse therapy was effective for cardiac sarcoidosis with ventricular tachycardia and systolic dysfunction. *ESC Heart Fail*. 2016 Dec;3(4):288–92.
- 13 Watanabe H, Eguchi K, Saitou T, Shimpo M, Hiroe M, Kario K. Cardiac sarcoidosis, the complete atrioventricular block of which was completely recovered by intravenous steroid pulse therapy. *J Cardiol Cases*. 2015 Oct;13(1):21–4.
- 14 Saito H, Yamaguchi T, Adachi Y, Yamashita T, Wakai Y, Saito K, et al. Neurological symptoms of sarcoidosis-induced small fiber neuropathy effectively relieved with high-dose steroid pulse therapy. *Intern Med*. 2015;54(10):1281–6.
- 15 Thulasidoss K, Asokan L, Chandra P, Rejliwal P. The clinical conundrum of diagnosing and treating systemic sarcoidosis in a high TB burden area. *BMJ Case Rep*. 2017 May;2017:bcr-2016-218741.
- 16 Knopf A, Lahmer T, Chaker A, Stark T, Hofauer B, Pickhard A, et al. Head and neck sarcoidosis, from wait and see to tumor necrosis factor alpha therapy: a pilot study. *Head Neck*. 2013 May;35(5):715–9.
- 17 Yoshida-Hata N, Yashiro S, Arai N, Takeuchi S. Brainstem infarction and panuveitis due to sarcoidosis successfully treated with steroid pulse therapy. *Case Rep Med*. 2012;2012:356743.
- 18 Kiuchi K, Miyashiro M, Kitagawa C, Wada S. Linezolid-associated optic neuropathy in a patient with ocular sarcoidosis. *Jpn J Ophthalmol*. 2009 Jul;53(4):420–4.
- 19 Knickebein JE, Wei M, Nussenblatt RB, Sen HN. Retinal pigment epithelium tear after immunosuppressive treatment for sarcoidosis-related choroidal granuloma. *Ocul Immunol Inflamm*. 2017 Dec;25(6):820–4.
- 20 Verma A, Biswas J. Choroidal granuloma as an initial manifestation of systemic sarcoidosis. *Int Ophthalmol*. 2010 Oct;30(5):603–6.
- 21 Marcus DF, Bovino JA, Burton TC. Sarcoid granuloma of the choroid. *Ophthalmology*. 1982 Dec;89(12):1326–30.

- 22 Kumar V, Gupta T, Jain S, Chanana B. Posterior sub-Tenon's triamcinolone in choroidal granuloma due to probable ocular sarcoidosis. *Oman J Ophthalmol*. 2013 May;6(2):127–8.
- 23 Khatib T, Hughes PW, Burton BJ. Unilateral choroidal granulomas complicated by choroidal neovascular membrane treated successfully with intravitreal triamcinolone in a patient with sarcoidosis. *BMJ Case Rep*. 2014 Aug;2014:bcr2014204794.
- 24 Chan WM, Lim E, Liu DT, Law RW, Lam DS. Intravitreal triamcinolone acetonide for choroidal granuloma in sarcoidosis. *Am J Ophthalmol*. 2005 Jun;139(6):1116–8.

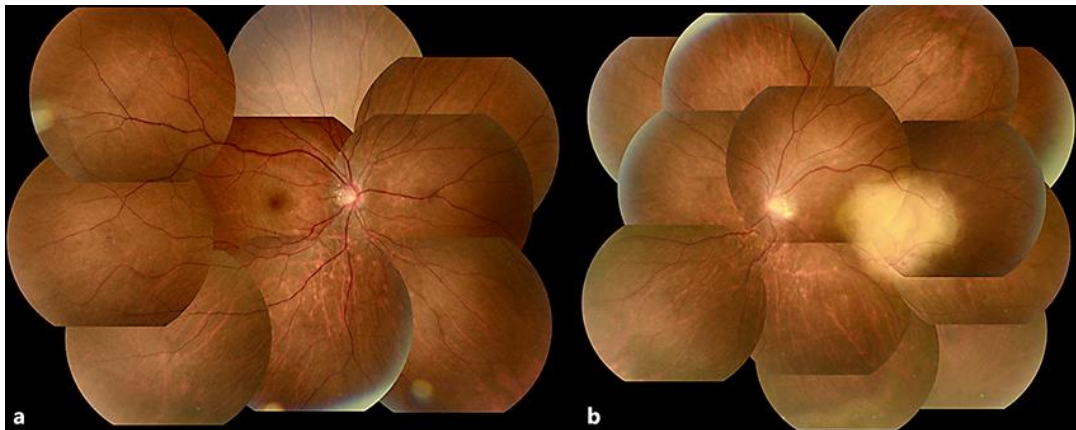


Fig. 1. Fundus photographs obtained at the initial examination (**a** right eye, **b** left eye). The fundus examination findings showed no particular abnormalities in the right eye, but a large white protruding lesion of 10 PD (lateral diameter) × 8 PD (longitudinal diameter) in the macular region, and slightly temporal to it, in the left eye. PD, papilla diameter.

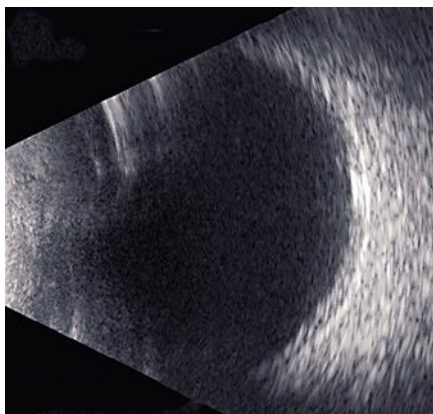


Fig. 2. B-scan ultrasonography image of the patient's left eye obtained at the initial examination. The B-scan ultrasonography image revealed that the lesion in the left eye appeared to have high internal reflectivity.

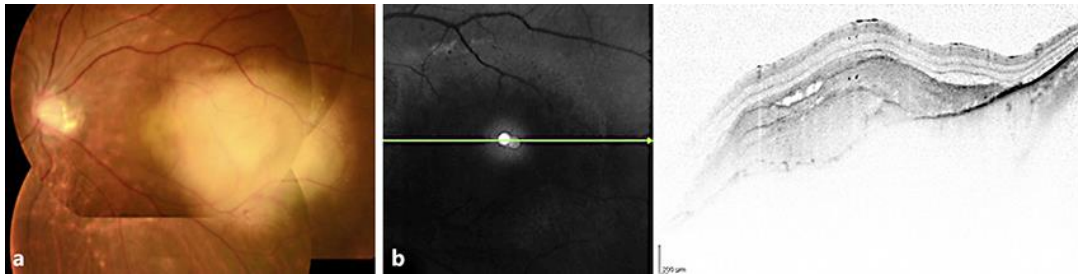


Fig. 3. Fundus photograph (a) and OCT image (b) of the patient's left eye. A protruding lesion was observed with a homogenous shadow on the choroid and under the retina, which was complicated by surrounding exudative retinal detachment. OCT, optical coherence tomography.

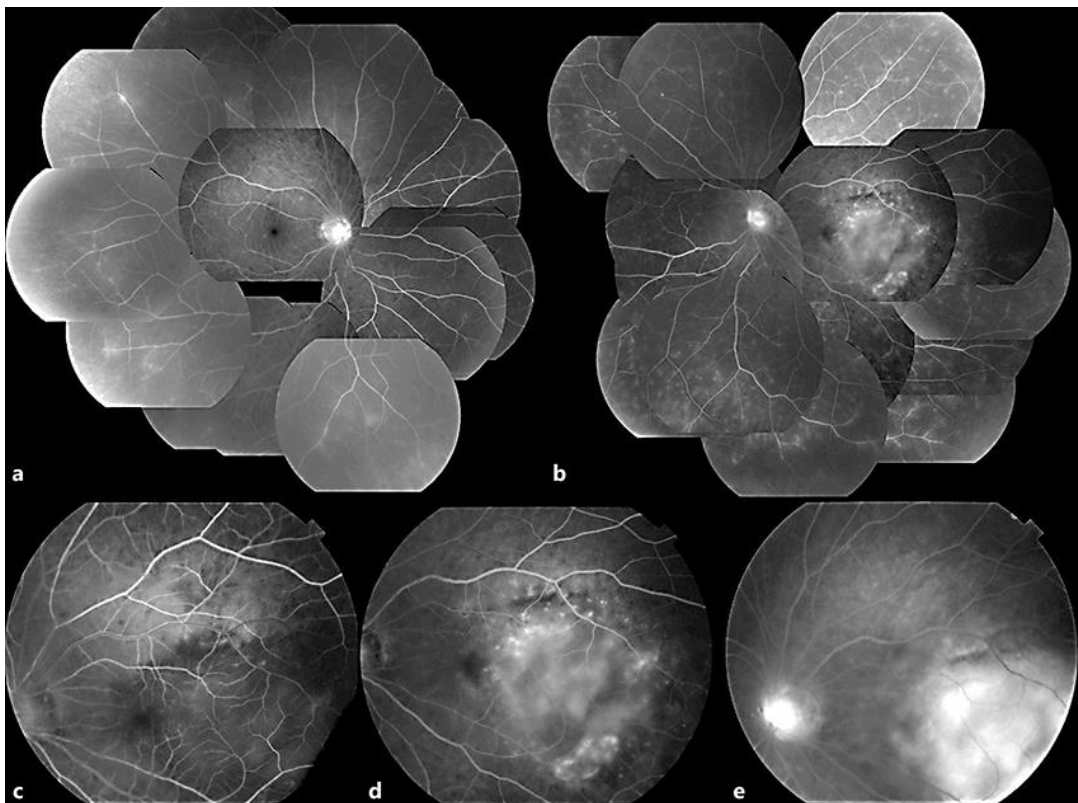


Fig. 4. Fluorescein angiography images (a right eye, b–e left eye). The fluorescein angiography findings showed slight fluorescein leakage in the peripheral region of the right eye (a). In the left eye, punctate fluorescein leakage corresponding to the tumor-like mass was noted at the early phase (b, c), and tissue staining and fluorescein pooling were observed from the middle to late phase (d, e).

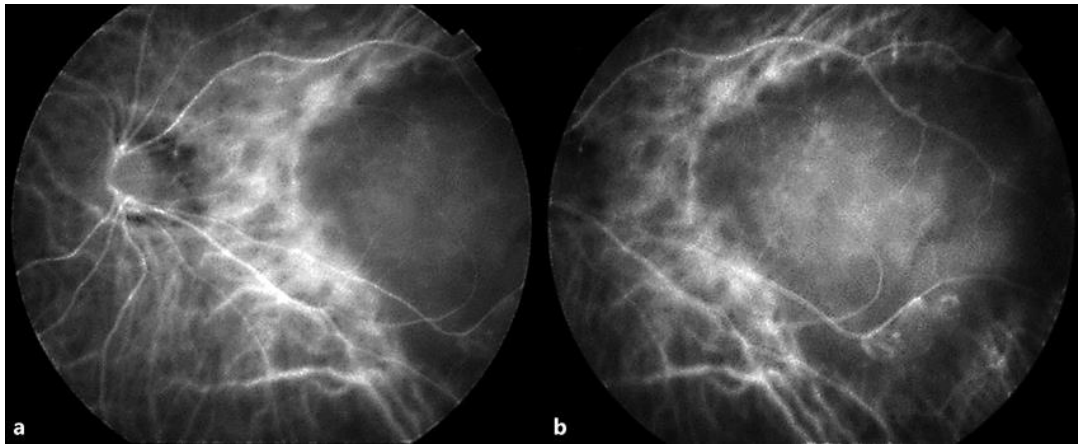


Fig. 5. Indocyanine green angiography image of the patient's left eye. Indocyanine green angiography revealed low fluorescence (a) and filling delay (b) in the area corresponding to the tumor mass.

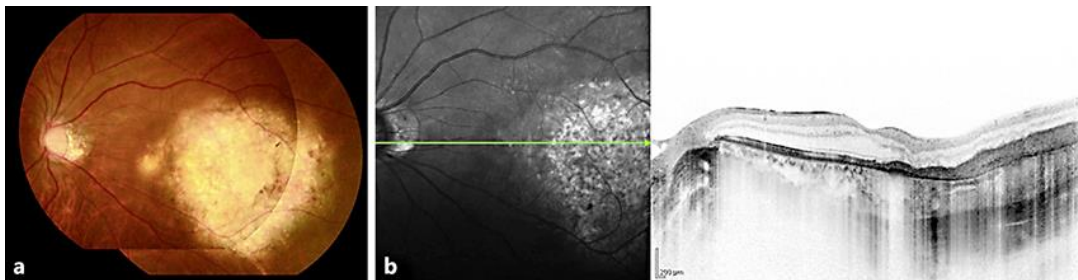


Fig. 6. Fundus photograph (a) and OCT image (b) of the patient's left eye 2 months after the start of steroid pulse therapy showing that the choroidal granuloma had markedly decreased. In addition, the patient's corrected VA was found to have improved to 0.7. OCT, optical coherence tomography; VA, visual acuity.