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Frosted Branch Angiitis: A Case of Bilateral Frosted Branch Angiitis After Fever

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Abstract: Frosted branch angiitis (FBA) is a rare retinal vascular disease that usually affects healthy, young adolescents and tends to be bilateral. Currently, there are limited reports on FBA worldwide. In this paper, we will present a case of FBA after fever.

Keywords: Frosted branch angiitis; Fever; Case report

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1. Introduction

This paper discusses a case of frosted branch angiitis (FBA) after fever. FBA is a rare retinal vascular disease that usually affects healthy, young adolescents and tends to be bilateral [1]. It is characterized by the sheathing of retinal arteries and veins, which resembles frost-covered branches. Kleiner has summarized previous case reports and categorized FBA into three types [2]: (1) frosted branch-like appearance, caused by direct infiltration of tumor cells into vascular endothelial cells [3]; (2) frosted branch response or secondary FBA, which is caused by autoimmune dysfunction [4] or infection due to virus [5], bacteria [6], or parasite [7]; (3) acute idiopathic FBA, which is not accompanied by any eye disease or systemic disease. So far, there have been limited reports on FBA worldwide, and we will be presenting a case in this paper. This study adhered to the tenets of the Declaration of Helsinki, and informed consent was obtained from the patient.

2. Case study

A 23-year-old female presented with fever and slight headache for 8 days, along with face swelling for a day. She was admitted to the Department of Pulmonary and Critical Care Medicine of The First Affiliated Hospital of Xi'an Medical University on April 21, 2019. She had no complaints of other discomfort, and there was no history of any eye or systemic illnesses. She was given antibiotics and antiviral outside the hospital, and her temperature was still high, reaching 39°C. After admission, ganciclovir, ceftriaxone, and glycerin fructose were administered. Initial blood investigations revealed increased white blood cells (14.18×10^9/L), platelets (570×10^9/L) and antinuclear antibodies (1:100, +). Her liver and renal function, blood coagulation profile, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), IgG and IgM antibodies to Hantaan virus, procalcitonin, cardiac enzymes, troponin, hepatitis B virus (HBV), hepatitis C

virus (HCV), human immunodeficiency virus (HIV), rapid plasma regain (RPR), respiratory virus, gram-negative bacterial endotoxin, Epstein-Barr virus DNA, and mycoplasma pneumoniae titer were all normal. Head MRI, cerebrospinal fluid biochemical tests, and tuberculosis examination were all negative.

After 2 days, her fever resolved, but her vision was blurry. On examination, the patient's best-corrected visual acuity (BCVA) was 20/400 on the right eye and counting fingers on the left. Her intraocular pressure was normal. Keratic precipitates (KP) (+), Tyndall effect (+), anterior chamber cell (+), and vitreous cell (++) were all present in both eyes. Funduscopic examination revealed bilateral diffuse retinal periphlebitis and twisted vessels resembling FBA (**Figure 1A**). Fluorescein angiography (FA) showed slightly delayed retinal-artery-to-vein transit time of 14 seconds and slight leakage of dye from the optic disc (**Figure 1B**). Spectral domain optical coherence tomography (SD-OCT) of the macula revealed a large amount of cystoid macular edema and a serous foveal detachment in both eyes (**Figure 1C**). B-mode ultrasound also showed retinal and papillary edema (**Figure 1D**).

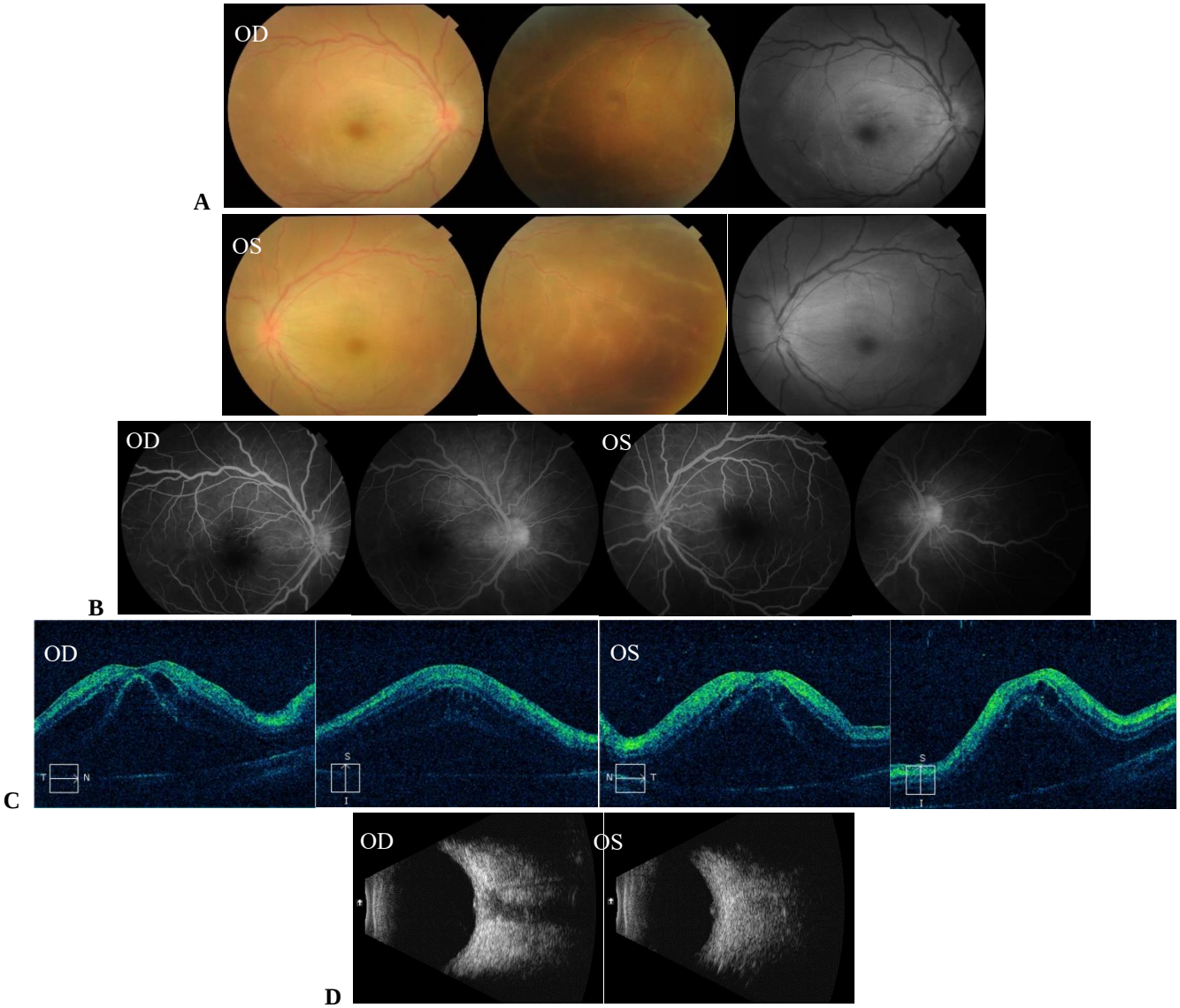

Figure 1. Eye examination on the day the patient was transferred to the Department of Ophthalmology on April 21, 2019; **A**: Fundus photograph of the right eye (OD) and the left eye (OS), showing diffuse retinal periphlebitis and twisted vessels; **B**: Late-phase fluorescein angiogram of both eyes showing slight leakage of dye from the optic papilla; **C**: Spectral domain ocular coherence tomography (SD-OCT) through the fovea of both eyes, showing cystoid macular edema and serous foveal detachment; **D**: B-mode ultrasound of both eyes, showing retinal and papillary edema

The diagnosis of FBA was made, and high-dose oral prednisone was started at 1.5 mg/kg/day. Prednisone was firstly reduced by 10 mg every 14 days, and when it reached 30 mg, the dose was further reduced by 10 mg per month. After treatment, the symptoms and signs gradually improved (**Figure 2**).

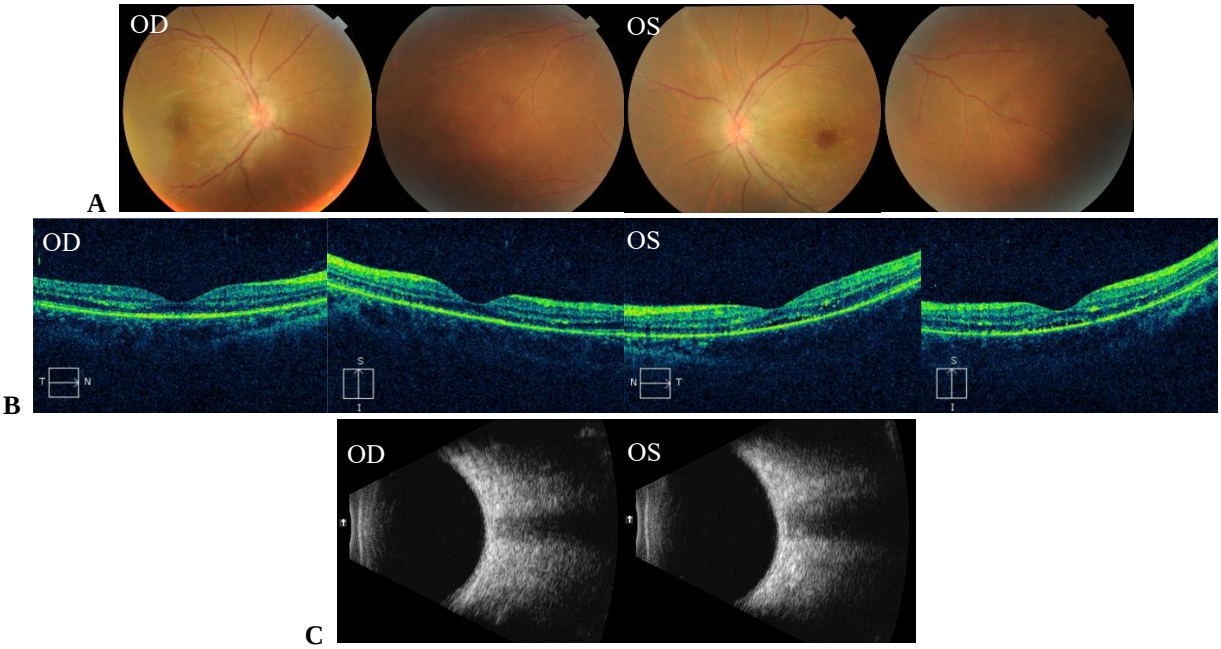

Figure 2. Eye examination after a week of treatment on April 30, 2019; **A**: Fundus photograph of both eyes, showing diminished retinal periphlebitis; **B**: SD-OCT through the fovea of both eyes showing fading macular edema in the right eye and slight foveal detachment in the left eye; **C**: B-mode ultrasound of both eyes, showing resolved retinal and papillary edema

On the day of discharge (May 9, 2019), the patient's BCVA was 20/25 on the right eye and 20/100 on the left. Vitreous cell (+) and markedly diminished bilateral perivascular sheathing were noted. The bilateral macular edema subsided, leaving the deposition of yellowish-white dotted exudates (**Figure 3**).

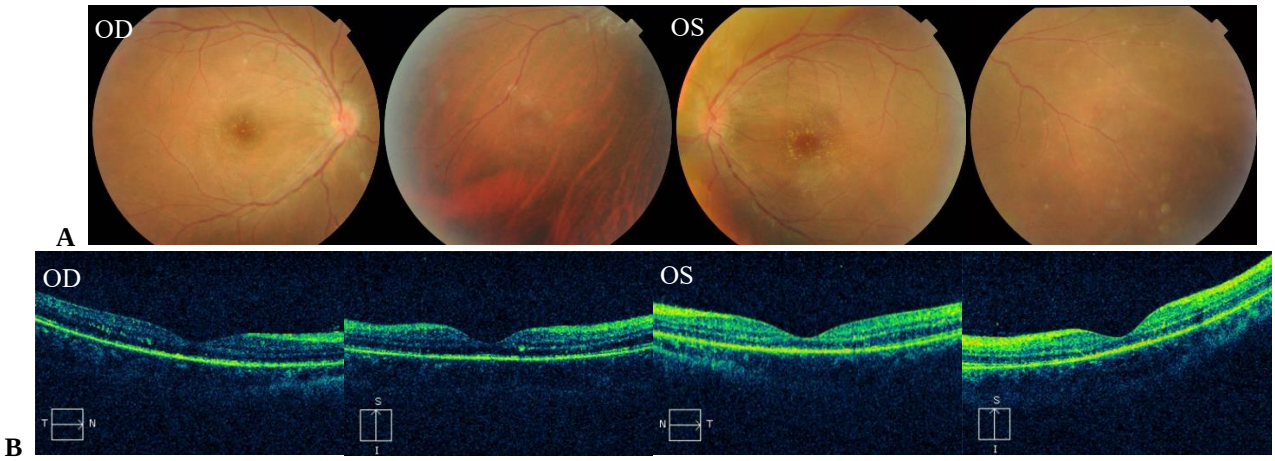

Figure 3. Eye examination after 2 weeks of treatment on May 9, 2019; **A**: Fundus photograph of both eyes, showing astral form exudates in both maculae; **B**: SD-OCT through the fovea, showing fading macular edema in both eyes

During her follow-up after nearly 3 months, her BCVA was maintained at 20/25 for both eyes, with only a few vitreous cells seen in addition to reduced macular exudative lesions in both eyes (**Figure 4** and **Figure 5**).

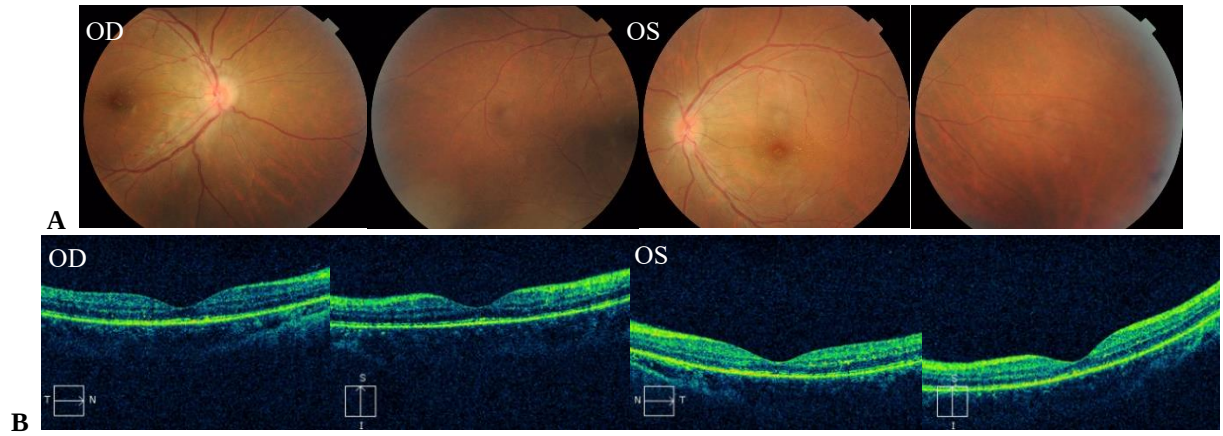

Figure 4. Eye examination after treating for 6 weeks on June 3, 2019; **A**: Fundus photograph of both eyes, showing reduced macular exudates in both eyes; **B**: SD-OCT through the fovea of both eyes, showing normal structures

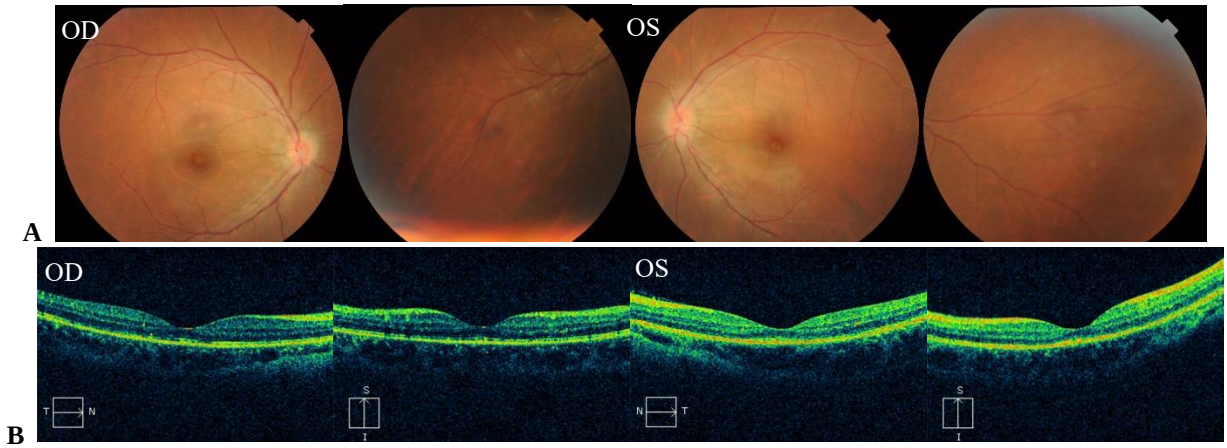

Figure 5. Eye examination after 10 weeks of treatment on July 1, 2019; **A**: Fundus photograph of both eyes, showing a few macular exudates in both eyes; **B**: SD-OCT through the fovea, showing binocular normal structures

The distinctive features of FBA include fast onset, rapid development, early uveitis and vitritis, sheath of retinal vessels presented as segmental sausage-shaped lesions, as well as apparent frosted branch-like changes to retinal vessels, especially in the peripherals on FA, without any vascular occlusion [8]. The patient's clinical manifestations were consistent with the features of FBA, and most parts of her retina were involved, thus resulting in poor vision in the initial stage. After active treatment for almost 3 months, her vision significantly improved, and fundus examination showed that the frosted branch-like changes in both retinas had resolved, with only a few yellowish-white punctate in the macula. The patient presented with persistent fever, positive for antinuclear antibody, and increased white blood cells, suggesting that the antigens of certain viruses or bacteria had triggered an inflammatory response, although detailed laboratory tests found no evidence of pathogens. Besides, the positive effect from a large amount of glucocorticoids also supports the diagnosis of secondary FBA.

Often, retinal edema is the main manifestation at the initial onset. As a result, the frosted branches obscured by retinal edema may not be visible. Therefore, making an accurate initial diagnosis is not very easy for an inexperienced doctor at a glance, thus requiring detailed eye examinations. Furthermore, FBA needs to be distinguished from acute retinal necrosis, intermediate uveitis, cytomegalovirus or herpes simplex retinitis, and Eales disease. Fundus photography, FA, OCT, and B-ultrasound examinations were carried out in a timely manner for this patient, and treatment with systemic steroids was promptly initiated, aiming at inflammation control and risk reduction of blindness. Although the relapse is rare, close follow-

up is necessary to observe the formation of macular scar or retinal neovascularization, which may occur in some cases.

Disclosure statement

The authors declare no conflict of interest.

References

[1] Walker S, Iguchi A, Jones NP, 2004, Frosted Branch Angiitis: A Review. Eye (Lond), 18(5): 527-533.

[2] Kleiner RC, 1997, Frosted Branch Angiitis: Clinical Syndrome or Clinical Sign?. Retina, 17(5): 370-371.

[3] Alhaj Moustafa M, Crowell EL, Elmahdy S, et al., 2018, Paraneoplastic Frosted Branch Angiitis as First Sign of Relapsed Hodgkin Lymphoma. Clin Case Rep, 6(10): 1978-1981.

[4] Ferreira BFA, Rodriguez EEC, Prado LLD, et al., 2017, Frosted Branch Angiitis and Cerebral Venous Sinus Thrombosis As An Initial Onset of Neuro-Behcet's Disease: A Case Report and Review of the Literature. J Med Case Rep, 11(1): 104.

[5] Mao F, Wu J, Sun H, et al., 2016, Frosted Branch Angiitis in an AIDS Patient with Cytomegalovirus Retinitis. Int J Infect Dis, 52: 9-11.

[6] Agarwal M, Shrivastav A, Waris A, 2018, Tubercular Retinal Vasculitis Mimicking Frosted Branch Angiitis: A Case Report. Ophthalmic Inflamm Infect, 8(1): 3.

[7] Suzuki T, Onouchi H, Nakagawa Y, et al., 2010, A Case of Recurrence of Congenital Ocular Toxoplasmosis with Frosted Branch Angiitis (Ocular toxoplasmosis with Frosted Branch Angiitis). Tokai J Exp Clin Med, 35(4): 122-125.

[8] He L, Zheng XL, 2011, A Case of Binocular Frosted Branch Angiitis. Int J Ophthalmol, 11(11): 2060-2061.