Case Report

VOGT KOYANAGI HARADA DISEASE: A CASE OF FAST RECURRENT

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ABSTRACT

A case report of a young lady diagnosed with Vogt-Koyanagi-Harada (VKH) disease with fast recurrence. An 18-years-old Maldivian lady presented with bilateral painless blurring of vision in both eyes (OU) for 1 week associated with redness. She also complained of severe headache and upper back pain. Her visual acuity (VA) was counting finger in the right eye (OD) and 1/60 in the left (OS). Anterior chamber revealed granulomatous reactions. Funduscopy revealed bilateral swollen optic disc, multiple punctate choroiditis and inferior exudative retinal detachment. Kernig sign was positive on neurological assessment. Optical computer tomography (OCT) of the macula showed multiple serous subretinal detachment OU. Fundus fluorescein angiography (FFA) OU revealed delayed choroidal filling, OU discs leakage, and multiple punctate hyperfluorescence with leakages. Infective screening was negative. There was marked visual gain to 6/12 after 3 days of intravenous Methylprednisolone., she was subsequently started on tapering dose of oral Prednisolone. Initiation of second line immunosuppressant was delayed due to incidental findings of anemia on blood investigation. The disease relapsed while she was on high dose of Prednisolone. Visual acuity was significantly reduced and OCT showed reappearance of subretinal fluid which required a second cycle of intravenous Methylprednisolone. This case highlights the importance of aggressive therapy in the case of VKH disease. Early commencement of other immunosuppressive therapies and the use of biologics need to be considered as a first-line treatment in severe cases.

CASE REPORT

An 18-year-old Maldivian lady with no known medical illness, presented with one-week history of bilateral progressive, generalized, painless blurring of vision associated with redness as well as headache and upper back pain. There was no previous history of ocular trauma or surgery. Visual acuity was counting finger in the right eye (OD) and 1/60 in the left eye (OS). Pupillary reactions were sluggish bilaterally. Anterior segment examination revealed bilateral granulomatous panuveitis, anterior chamber cells of 2+, the presence of keratic precipitates and busacca nodules. Funduscopic findings include bilateral optic disc swelling, macula oedema, as well as multiple punctate choroiditis associated with inferior exudative retinal detachment(Figure1a-b). Intraocular pressure was normal in both eyes. Neurological examination revealed a positive Kernig sign, however there was no other neurological signs found.

Optical computer tomography (OCT) of the macula showed multiple serous retinal detachment OU (figure 2a-b). Fundus fluorescein angiography (FFA) revealed delayed choroidal filling, discs leakage, and multiple punctate hyperfluorescence with leakages bilaterally (figure 3a-b). Blood investigation including erythrocyte sedimentation rate and infective screening were all normal except for a hypochromic microcytic anemia. Her Mantoux test was 0 mm. A diagnosis of bilateral acute Vogt-Koyanagi-Harada disease was made.

She was immediately started on intravenous methylprednisolone 250mg QID for three days, followed by oral prednisolone 60mg daily (1mg/kg/day). Topically she received Prednisolone acetate 1%, Mydriacyl 1%. Best corrected visual acuity (BCVA) markedly improved to 6/12 OD and 6/18 OS upon completion of intravenous methylprednisolone. Initiation of second-line immunosuppressant was deferred due to the anemia which warranted further investigation. While on the high dose of the oral prednisolone, her vision deteriorated due to recurrent multifocal choroiditis and subretinal fluid which was seen on OCT. She was then readmitted for another cycle of intravenous methylprednisolone. Unfortunately, the patient was loss to follow-up thereafter.

DISCUSSION

Vogt–Koyanagi–Harada (VKH) disease is a multisystem granulomatous inflammatory autoimmune disorder with ophthalmic, dermatologic, neurologic and auditory manifestations. The ocular

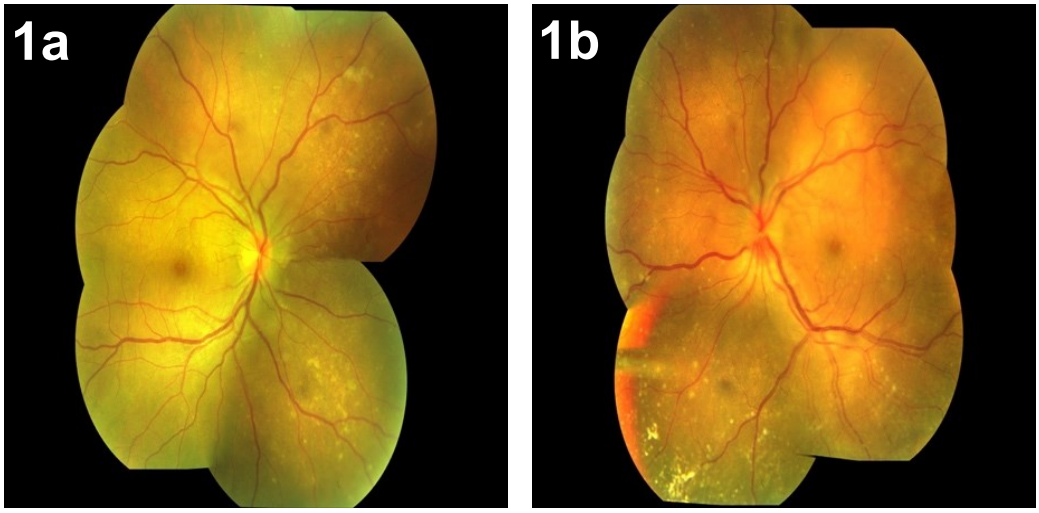

Figure 1: Fundus photography OD (1a) and OS(1b) show optic disc swelling with multiple punctate choroiditis.

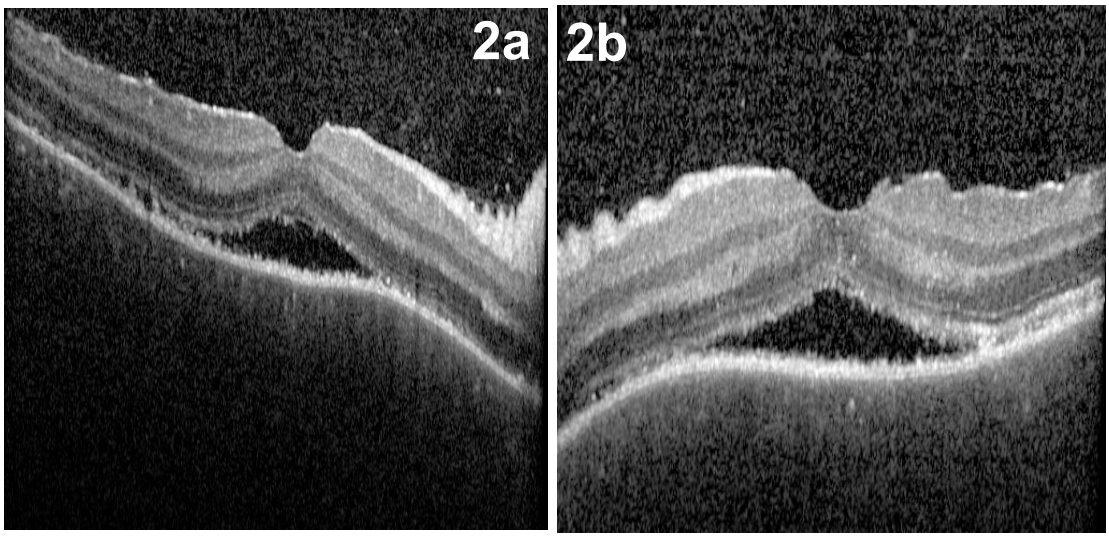

Figure 2: OCT of macula: OD (2a) and OS (2b) show the presence of subretinal fluid.

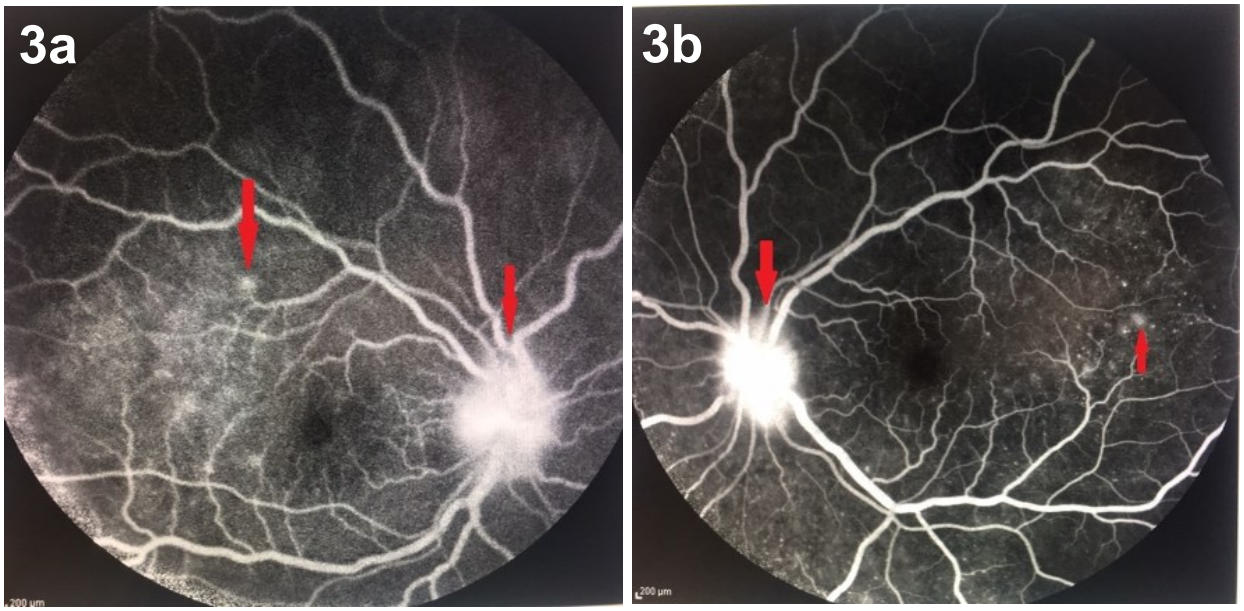

Figure 3: Fundus Fluorescein Angiography: OD (3a) and OS (3b) show disc leakages and multiple hyperfluorescence with leakages.

manifestations include severe panuveitis with iridocyclitis, diffuse choroidal swelling, serous retinal detachment and optic disc hyperaemia. Patients can present with bilateral granulomatous panuveitis with or without extraocular manifestations [1]. Incidence of VKH disease varies depending on the geographic location and ethnicity. Darkly pigmented races are primarily affected such as Hispanics, Asians, and Asian Indians. It mostly affected female and patients in their fifth decades of life [2].

The diagnostic criteria of VKH disease based on the recommendation by The American Uveitis Society in 1978 includes (1) the absence of any history of ocular trauma or surgery; and (2) the presence of at least three of the following four signs: (a) bilateral chronic iridocyclitis; (b) posterior uveitis, including exudative retinal detachment, forme fruste of exudative retinal detachment, disc hyperaemia or oedema and “sunset-glow” fundus; (c) neurologic signs of tinnitus, neck stiffness, cranial nerve, or central nervous system disorders, or cerebrospinal fluid pleocytosis; and (d) cutaneous findings of alopecia, poliosis, or vitiligo. However, other disease entities such as sympathetic ophthalmia, ocular syphilis, posterior scleritis and acute posterior multi-focal placoid pigment epitheliopathy may have similar presentation with VKH disease and needed to be ruled out first [1].

The aim of treatment in the acute stage is to halt the disease process from entering the chronic recurrent phase. Standard treatment includes oral prednisolone (1 – 1.5 mg/kg) or initial intravenous methylprednisolone, preferably within 2 weeks from the beginning of symptoms, followed by slow tapering of oral corticosteroids. Treatment should be no less than 6 months and the dose should be titrated according to the severity of the inflammation [2].

Immunomodulators such as azathioprine and methotrexate in combination with corticosteroids as first-line therapy have been reported to significantly reduce the recurrence rates, late complications and improvement in visual outcome in acute disease [2]. In our case, the initiation of the immunomodulator was delayed due to the fear of bone marrow suppression effects causing deterioration of anemia which was incidentally found during investigation. Hematological toxicity is a serious and potentially life threatening effect which presents as myelosuppression and pancytopenia, which can occur even on low dose immunomodulatory treatment [3].

Biologics are recently studied medication and have been used widely for the treatment of non-infectious uveitis and other ocular inflammatory diseases. It has been used as an alternative or as an adjunct treatment in patients with inadequate response or intolerance towards conventional Immunotherapy. Biologics agents such as Infliximab and Adalimumab (anti – TNF) have been reported to be an effective treatment of VKH disease [2]. It was reported to significantly increase the hemoglobin value based on studies in a patient with rheumatoid arthritis, psoriatic arthritis and ankylosing spondylitis [2,4,5].

This case highlights the importance of aggressive therapy in Vogt-Koyanagi-Harada disease. In cases where immunomodulators cannot be commenced, early consideration of biologics such as infliximab and adalimumab should be sought to prevent the progression of this potentially blinding condition.

DECLARATION OF CONFLICT OF INTEREST

The authors report no conflicts of interest.

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