A Case of Vogt- Koyanagi - Harada Disease in a 12 years old boy at a Tertiary Eye Hospital – A Rare Case Report

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Abstract

Vogt–Koyanagi–Harada (VKH) disease is a T-cell-mediated autoimmune disorder characterized by bilateral granulomatous panuveitis with various systemic manifestations. VKH disease rarely occurs in the pediatric population, the clinical course tends to be aggressive. We report a case of Vogt–Koyanagi–Harada (VKH) disease in a 12-year-old male child who presented with sudden severe loss of vision 2/60 in both the eyes. The anterior segment examination showed cells, flare (3+), fine keratic precipitates, posterior synechia and fundus examination showing hyperemic disc with blurred margin and exudative retinal detachment. Color fundus photography, Fundus fluorescein angiography, OCT macula and B-Scan further confirmed the diagnosis. The patient was treated with oral prednisolone for 1.5 months with tapering dose. Visual acuity improved to 6/12 both eyes. To the best of our knowledge paediatric VKH is rare and more aggressive but this patient responds very well with oral corticosteroid alone.

Keywords : *Vogt–Koyanagi–Harada disease, Corticosteroid, Paediatric.*

Introduction

Vogt-Koyanagi-Harada (VKH) disease is a chronic, bilateral, granulomatous panuveitis associated with cutaneous, neurologic, and auditory manifestations.[1] The exact cause remains unknown, but evidence suggests that it involves a T-lymphocyte-mediated autoimmune process directed against antigens associated with melanocytes. Several studies demonstrated that tyrosinase family proteins are the antigens associated with VKH syndrome disease.[2,3] The onset age of VKH disease tends to be approximately from 20 to 50 years. Therefore, VKH disease in children is very rare[4]. We experienced a twelve-year-old boy affirmatively diagnosed with VKH disease though aggressive but who was successfully treated with oral steroid.

Case Report

A 12 years old male boy presented with sudden severe loss of vision in both eyes for about 15 days which was painless. No history of headache, hearing problem and skin changes. After that he went to local ophthalmologist and referred to our hospital for evaluation and management.

On examination he was found to have visual acuity 2/60 in both the eyes, not improved with pinhole. Anterior segment examination revealed Cells(+++), posterior synechia, vitritis and Fundus examination showed optic disc hyperemia and exudative retinal detachment in both the eyes. He was diagnosed as paediatric probable VKH in acute uveitic phase and advised for B-scan, FFA and OCT macula. B-scan revealed mild vitreous opacity, Exudative retinal detachment and increased chorioretinal thickness (Fig-1), FFA showed Hyperfluorescein area, capillary non-perfusion in the peripheral retina(Fig-2) and OCT macula revealed neuro-sensory detachment(Fig-3) in both eyes .

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Patient was treated with high dose of oral prednisolone and tapered over one and half months. Patient responded well with good visual recovery 6/12 in both eyes.

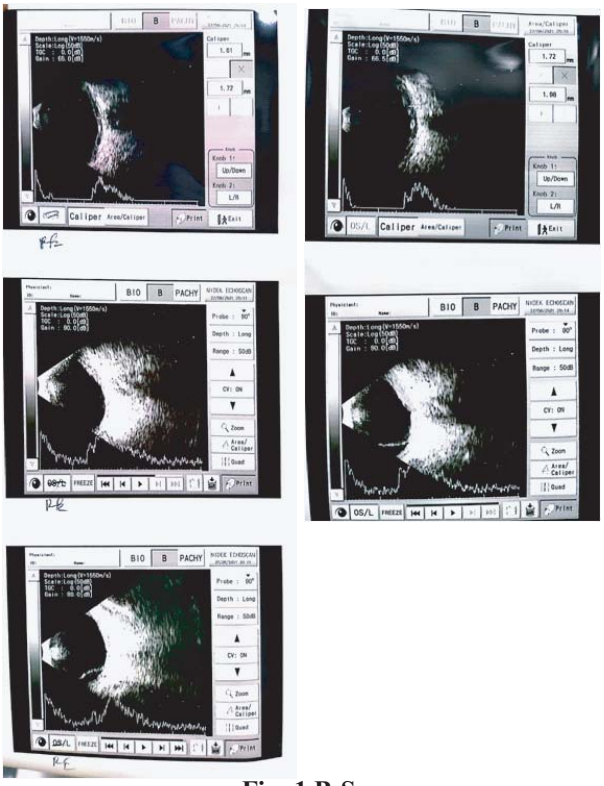

Fig: 1 B-Scan

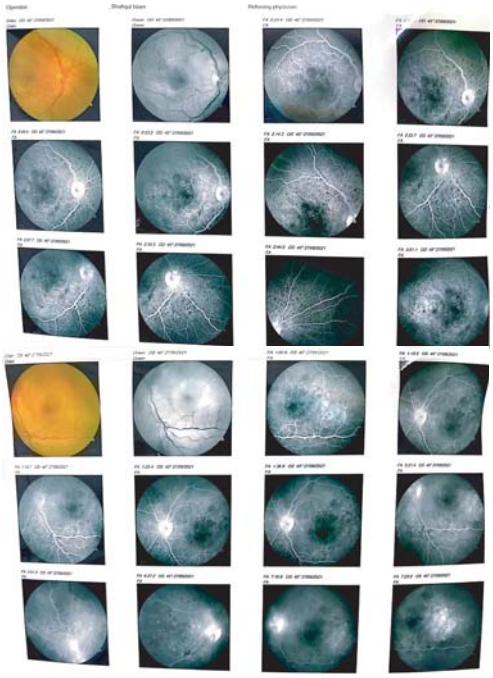

Fig: 2 FFA

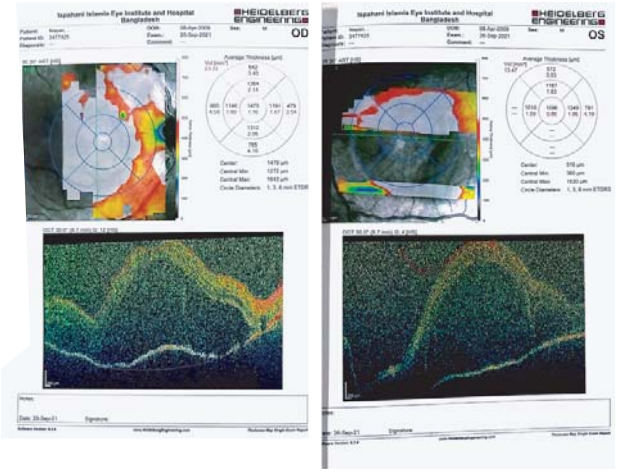

Fig: 3 OCT macula

Discussion

VKH is a disease uncommonly affects the pediatric age group. Only 13%–15% of VKH patients were found to be children.[5,6] To the best of our knowledge, the youngest patient is three-year-old boy reported by Takada et al.[7]. He was diagnosed with VKH disease by ocular symptoms (serous retinal detachments and papillitis and inflammation of anterior chamber), aseptic meningitis, and OCT images, although fluorescent fundus angiography was not performed. A retrospective case review of VKH disease reported that the ratio of final BCVA of 20/200 or worse in the pediatric group (61%) was significantly higher than that in the adult group (26%) because of aggressive inflammation and subsequent severe ocular complications[5].

In our case ocular inflammation was so severe that high-dose of oral corticosteroid therapy were required to resolve the serous retinal detachment. No neurological and auditory manifestations was there. Though aggressive inflammation our patient respond very well, vision becomes 2/60 to 6/12 in both eyes, whereas previous studies have found complete or partial recovery of vision with high dose of intravenous corticosteroid. The limitations are no HLA typing and genetic analysis were done.

Conclusion

Favorable clinical course in our patient suggests that initial treatment with high-dose oral

corticosteroid therapy with slow tapering might be beneficial in pediatric patients with Probable VKH disease.

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