Section: Ophthalmology

Case Report

VOGT-KOYANAGI-HARADA DISEASE WITH HASHIMOTO'S THYROIDITIS-A RARE CASE REPORT

Leshna Kopparapu[1], B.Dharma Raju[2], Md.Ruksana[3], I.Mounika, Y.Roja.[4]

[1]*Junior Resident, Department of Ophthalmology, NRI Institute of Medical Sciences, Visakhapatnam, Andhra Pradesh, India.*
[2]*Professor, Department of Ophthalmology, NRI Institute of Medical Sciences, Visakhapatnam, Andhra Pradesh, India.*
[3]*Junior Resident, Department of Ophthalmology, NRI Institute of Medical Sciences, Visakhapatnam, Andhra Pradesh, India.*
[4]*Junior Resident, Department of Ophthalmology, NRI Institute of Medical Sciences, Visakhapatnam, Andhra Pradesh, India.*
[5]*Junior Resident, Department of Ophthalmology, NRI Institute of Medical Sciences, Visakhapatnam, Andhra Pradesh, India.*

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Corresponding Author:
Dr. Leshna Kopparapu
Junior Resident, Department of Ophthalmology, NRI Institute of Medical Sciences, Visakhapatnam, Andhra Pradesh, India.
Email: leshnachinnu17@gmail.com.

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ABSTRACT

Background: Vogt–Koyanagi–Harada (VKH) disease is a rare uveitic entity which could be associated with autoimmune thyroid disease. This report was aimed to investigate with optical coherence tomography for the diagnosis of VKH disease and associated autoimmune thyroid disease confirmed by anti thyroperoxidase (TPO) antibodies.

Case study: A 20-year-old young female came with a chief complaint of defective vision both eyes since 1 day. Past history of viral illness, fever since 1 week associated with headache, orbital pain and photophobia. No history of any ocular trauma or ocular surgery in the past. Ophthalmic evaluation revealed visual acuity in both eyes 6/12, fine pigmented keratic precipitates KP's over corneal endothelium and aqueous grade 1 cells, along with bilateral serous macular detachment, retinal pigment epithelium(RPE) changes and choroidal thickening. Patient is a known case of hypothyroidism and she is on Tab. Thyroxine sodium 50mcg.To exclude any association with other autoimmune disorders, Anti TPO and anti-nuclear antibody(ANA) were done. Thyroid stimulating hormone(TSH) levels were elevated, anti TPO were positive confirming hashimoto's thyroiditis and ANA weak positive for the exclusion of lupus choroidopathy.

Conclusion: VKH, essentially a clinical diagnosis may be associated with other autoimmune disorders like Hashimoto's, Grave's disease which need to be evaluated. Visual prognosis depends on early treatment with steroids and immunosuppressants, preventing recurrence of the disease process. This case highlights the importance of evaluation of VKH disease and other associated autoimmune disorders.

Keywords: Vogt-Koyanagi-Harada Disease, Hashimoto's Thyroiditis.

INTRODUCTION

VKH disease is B/L granulomatous panuveitis associated with serous retinal detachment with or without extraocular manifestations affecting young adults. In 1932, Babel suggested to call entity VKH disease. VKH disease is T-cell mediated autoimmune reaction associated with HLADR1, HLADR4 (subtypes 0405). VKH disease presents in 3 forms as Complete VKH, Incomplete VKH and Probable VKH according to revised diagnostic criteria. Complete VKH includes 1. Bilateral ocular involvement - diffuse choroidal thickening, bullous serous retinal detachment, recurrent anterior uveitis and multifocal areas of pinpoint leakage on fundus fluorescein angiography.2. Neurologic-meningismus, tinnitus, CSF pleocytosis3. Integumentary-alopecia, vitiligo, poliosis. Incomplete VKH includes point 1 and either 2 or3 must be present, while Probable VKH includes only point 1 of bilateral ocular involvement.[1,2] The Autoimmune thyroid disease (AITD0 is characterized by the dysfunction of thyroid tissue by antibody-mediated immune inflammation and the etiology of AITD is multifactorial and occurs predominantly in females.[3] In this study, we reported a rare case of probable VKH disease accompanied with AITD. There are few reports about association of both diseases.

CASE REPORT

A student of 20year-old female residing at Visakhapatnam has presented to our hospital with defective vision of both eyes, sudden onset, gradually progressive since 1 day. Past history of viral illness, fever since 1 week associated with headache, orbital pain and photophobia. No history of any ocular trauma or ocular surgery in the past. Her best corrected visual acuity (BCVA) was 6/12 in botheyes. Slit-lamp examination revealed circumcorneal congestion, fine pigmented KP's over corneal endothelium and aqueous grade 1 cells. Dilated fundus examination demonstrated bilateral serous macular detachment, RPE alteration and choroidal thickening. There was subretinal fluid accumulation with retinal detachment and thickening of choroid confirmed as evidenced on optical coherence tomography (OCT) and B-scan.

Patient had history of prodromal illness 5 days before defective vision associated with headache, orbital pain, light sensitivity and mild fever. She is a known case of hypothyroidism. Fundus examination with indirect ophthalmoscopy(IO) & 90D shows

both eyes media clear, Optic disc is normal size, shape, colour with well-defined margins, cup disc ratio(CDR) is 0.4:1, normal arteries: veins (AV) ratio 2:3, macula with serous macular detachment +, foveal reflex dull and peripheral retina normal. Optical coherence tomography macula showed multiple pockets of subretinal fluid, neurosensory retinal detachment with RPE alteration in both eyes. B-scan showed both eyes choroidal thickening. Fundus fluorescein angiography (FFA) shows Early Hypo with multiple pin point Hyper fluorescence with RPE alteration establishing our diagnosis of VKH disease. Elevated levels of TSH 10 uIU/ml and Anti Thyroperoxidase antibody 6.9 IU/ml suggestive of Hashimoto's thyroiditis. ANA- 1:40 Sample dilution Weak positive, ESR-116mm/1st hr. ENT is pure tone audiometry are within normal limits, No Dermatological Integumentary signs noted. Based on the above findings, we came to a clinical diagnosis of Probable Vogt Koyanagi Harada disease with Hashimoto's thyroiditis.

Patient was given both eyes periocular transeptal triamcinolone acetonide 40mg injections and started on oral tab prednisolone 60mg once daily for one week.

Review after one-week visual acuity improved in right eye 6/9 and left eye6/6. Optical coherence tomography shows reduced and resolved subretinal fluid with RPE changes. Tab Mycophenolate mofetil 500mg once daily is added to the treatment regimen followed by incremental dose of 500mg weekly till 4 weeks in combination with steroids on tapering doses.

DISCUSSION

The VKH disease is an auto immune multisystem disease and it may be combined with neurologic, auditory, and cutaneous manifestations.[1] It is common for VKH disease to occur in patients with genetic predispositions to the disease, including those from Asia, Latin America, and the Middle East.[1] The role of genetic factors in the development of VKH, such as HLA alleles, CTLA-4, and other polymorphisms of genes had been identified and studied. However, the importance of these genomic findings are currently unclear. There are 4 phases in the presentation of VKH disease, including prodromal, acute uveitic, convalescent, and chronic recurrent phases. Symptom such as dermatologic changes commonly occur in late convalescent in chronic phase. In 2011, Rao et al,[4] compared 1147 patients with bilateral uveitis in 10 centres, 180 of these patients were diagnosed with VKH disease with extremely high positive association with exudative retinal detachment. More than half of these patients with acute VKH disease presented only with ocular manifestations.[5] However, there is still need to differentiate VKH disease from other causes of panuveitis, including, sympathetic ophthalmia, infectious intraocular inflammation, intraocular lymphoma, posterior scleritis, uveal effusion syndrome, sarcoidosis, and other systemic disorders.[1] Therefore, it is always necessary to evaluate the patient's complete history, systems and physical examinations, and laboratory results. In our case, systemic evaluation and laboratory testing results were normal for exclusion of other panuveitic entities. The clinical and angiographic features in both eyes were determined to be typical for the acute phase of VKH syndrome.

In literature reviews, the association of VKH syndrome with AITD has been reported in 6 cases (5 woman out of 6, aged between 20 and 65 years). In these reports, 4 of them were determined with hypothyroidism and elevated TPO-Ab/Tg-Ab titers, and 2 of them were of hyperthyroidism.[6,7,8] Drug-induced hypersensitivity is considered as a triggering factor for T cell-mediated autoimmune dysfunction to induce Grave disease followed by VKH ssyndrome. It was also reported that VKH syndrome and Hashimoto thyroiditis may share a basic defect in the autoimmune pathogenetic process.[4] The HLA alleles was also considered as a factor that can trigger VKH syndrome associated with autoimmune polyglandular syndrome type 1. In addition, another report has demonstrated that a 45-year-old Saudi man of VKH syndrome associated with hypothyroidism and diabetes mellitus. They considered that the pathogenesis of VKH syndrome can include microbial agent or antigen to alter cell surface component of skin, uvea, follicular epithelial cell of thyroid, and pancreas that can trigger autoimmunogenic organ dysfunctions. Compared to the previous studies,[9,10,11] In our case good visual acuity is achieved by early diagnosis with optical coherence tomography, fundus fluorescein angiography followed by aggressive therapy with periocular and oral steroids in acute uveitis stage and maintenance therapy with a combination of oral steroid and antimetabolites.

Mycophenolate mofetil being safe and effective steroid sparing drug is preferred antimetabolite in combination with steroids on long term treatment as it prevents progression of VKH disease to chronic recurrent stage. Diagnosis of Hashimoto's thyroiditis with positive Anti TPO antibodies, exclusion of other autoimmune diseases like lupus choroidopathy and referral to ENT, Dermatology departments for evaluation of extra ocular manifestations is important in our case management.

CONCLUSION

Visual prognosis depends on early treatment with steroids and Immunosuppressants, which results in fair prognosis with 60-70% retaining vision 6/9 or better, as achieved in this patient. Prevention of recurrence of disease leading to choroidal neovascular membrane and subretinal fibrosis is important to restore good visual potential. This case highlights the importance of evaluation and management of VKH disease in detail to find out any other associated autoimmune diseases like Hashimoto's thyroiditis which can lead to serious systemic complications like heart failure, thyroid lymphoma.

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