Vogt Koyanagi Harada Syndrome Presenting as Isolated Ocular Disease—A Case Report

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INTRODUCTION

Vogt–Koyanagi–Harada (VKH) syndrome is an idiopathic multisystem autoimmune disease featuring inflammation of melanocyte-containing tissues such as the uvea, ear and meninges. VKH disease occurs more commonly in patients with a genetic predisposition to the disease. Several human leukocyte antigen (HLA) associations have been found in patients with VKH disease, including HLA-DR4, HLA-DR53, and HLA-DQ4 [1]. VKH can be subdivided into Vogt–Koyanagi disease, characterized mainly by skin changes and anterior uveitis, and Harada disease, in which neurological features and exudative retinal detachments predominate. Diagnosis is made by findings of fundus fluorescein angiography and B-scan. Treatment involves high dose oral corticosteroids pulsed with intravenous corticosteroids [2]. Prognosis depends on early recognition and aggressive control of the early stages of the disease. Our aim of study is to identify patients with vögkoyanaghairada disease presenting with limited clinical features.

CASE REPORT

A 20 year old male patient presented to our out-patient department on the 8th of March, 2014 with the chief complaint of diminished vision of a gradually progressive and painless onset in both eyes since a week. On examination, the anterior segment was normal except that both his pupils reacted sluggishly to both direct and indirect light reflexes. His fundus in both eyes showed peripapillary and macular edema about 3 disc diameters in size with mild perivascular sheathing. Fundus fluorescein angiography (FFA) showed multifocal hyper fluorescent dots at the level of RPE and accumulation of dye in the sub retinal space. B-scan showed localized retinal detachments in both eyes. It was diagnosed as Vogt Koyanagi Harada syndrome presenting as isolated ocular disease. VKH syndrome is an idiopathic multisystem autoimmune disease featuring inflammation of melanocyte-containing tissues such as the uvea, ear and meninges.

Key words: VKH syndrome, peripapillary edema, multiple serous retinal detachments
Figure 1: FFA showing multifocal hyperflourescent dots at the level of RPE and accumulation of eye in the subretinal space

Figure 2: B-Scan showing localized retinal detachments and choroidal thickening in both eyes

Figure 3: Visually evoked potential showed normal pattern reversal stimulation, P100 latencies and amplititude on bilateral sides
It was diagnosed to be Vogt Koyanagi Harada syndrome based on the fundus signs, fundus fluorescein angiography and B-scan. He was treated with intravenous corticosteroids i.e, Inj Methyl prednisolone 1 gm IV for 3 days followed by oral steroids. His vision improved to 6/9 on the 3rd day of treatment and 6/6 two weeks later.

**DISCUSSION**

The diagnosis of VKH which can be defined as chronic, bilateral granulomatous[5] ocular and multisystem autoimmune[6] inflammatory condition, of unknown cause, is usually made on the basis of clinical findings and by excluding the other possible uveomeningoencephalitic syndromes. For this reason clinical criteria recommended by the American Uveitis Society can be used. This case also met the criteria. The diagnosis of VKH syndrome was made depending on the history, clinical signs [7] and laboratory findings and our case presented as isolated uveal involvement with papilloedema and thickening of posterior choroid without anterior uveitis and accumulation of subretinal fluid and serous retinal detachment is the hallmark feature of vogt-koyanagi-harada disease

**Modified diagnostic criteria[8] for VKH syndrome**

1. Absence of a history of penetrating ocular trauma
2. Absence of other ocular disease entities
3. Bilateral uveitis
4. Neurological and auditory manifestations
5. Integumentary findings, not preceding onset of central nervous system or ocular disease, such as alopecia, poliosis and vitiligo.

In complete VKH, criteria 1–5 must be present.

In incomplete VKH, criteria 1–3 and either 4 or 5 must be present.

In probable VKH (isolated ocular disease), criteria 1–3 must be present.

In our case criteria 1-3 are met,so our case presented as isolated ocular disease according to modified diagnostic criteria.

Treatment involves high-dose oral prednisolone (60–100 mg/day) that may be augmented with 3-day intravenous pulse therapy with methylprednisolone (500–1000 mg/day).

Prognosis [9] depends on early recognition and aggressive control of the early stages of the disease. Late diagnosis or incorrect initial therapy is more likely to be associated with a guarded prognosis with only 50% of patients having a final visual acuity better than 6/12.

Differential diagnosis of bilateral exudative retinal detachments

- Carcinoma metastatic to the choroid.
- Uveal effusion syndrome.
- Posterior scleritis.
- Eclampsia.
- Central serous retinopathy.
- Age-related wet macular degeneration.

**CONCLUSION**

VKH syndrome can present as an isolated ocular disease. Treatment involves the use of high dose oral corticosteroids pulsed with intravenous corticosteroids. Prognosis depends on early recognition and aggressive control of the early stages of the disease. Late diagnosis or incorrect initial therapy is more likely to be associated with a guarded prognosis.

**CONFLICT OF INTEREST**

The authors declared no conflict of interest.

**FUNDING:** None

**REFERENCES**