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Vogt-Koyanagi-Harada Syndrome

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Vogt-Koyanagi-Harada (VKH) syndrome is a rare autoimmune multisystemic disease involving the melanocyte-containing organs; it is a diagnosis of exclusion. The disease is progressive and has undesired complications. We report the first case of VKH syndrome in the Kingdom of Bahrain. The report aims to describe this rare syndrome with more emphasis on the ocular manifestations and management.

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Vogt-Koyanagi-Harada (VKH) Syndrome, formerly known as uveoencephalitis, is a rare granulomatous autoimmune multisystemic disease[1]. It affects the melanocytes-containing organs, eyes, skin, inner ears, meninges, and central nervous system.

In the 9th century, the ocultist Ali Ibn Isa Al Kahhal (Baghdad) had described this inflammatory eye condition and associated with the cutaneous manifiestations[2].

In the early 20th century, scientists Vogt, Harada, Koyanagi, et al reported cases of uveitis with deafness, vitiligo, and/or alopecia separately[1]. The syndrome was named after the aforementioned scientists due to their contribution.

Although VKH is a rare disease, it has racial, sexual and age preferences. It is found to be more prevalent among Asians, Africans, Indians and Latinos. Females are 50% more prone to the syndrome compared to males. The disease usually affects young adults, second to fourth decade[3]. Unusual presentations and association with other autoimmune diseases have been reported[4]. If the patient presents in childhood, the disease could recur and become aggressive.

The aim of this report is to present rare case of Vogt-Koyanagi-Harada (VKH) syndrome, its management and to raise awareness about this rare and multisystemic disease.

THE CASE

A seventeen-year-old healthy Bahraini male student presented with bilateral painful loss of vision for one day. The patient woke up with painful red eyes and noticed marked decrease of vision associated with photophobia. The patient was seen two months earlier by an ophthalmologist for unilateral painful right eye, and advised frequent use of topical steroid. At the previous episode, vision was mildly affected and resolved with the treatment. The patient had vitiligo patches on the trunk and poliosis of scalp hair, eye lashes and trunk hair

which have appeared recently, see figures 1 and 2. Patient had no neurological and auditory symptoms. Review of systems was unremarkable for any associated autoimmune disease.

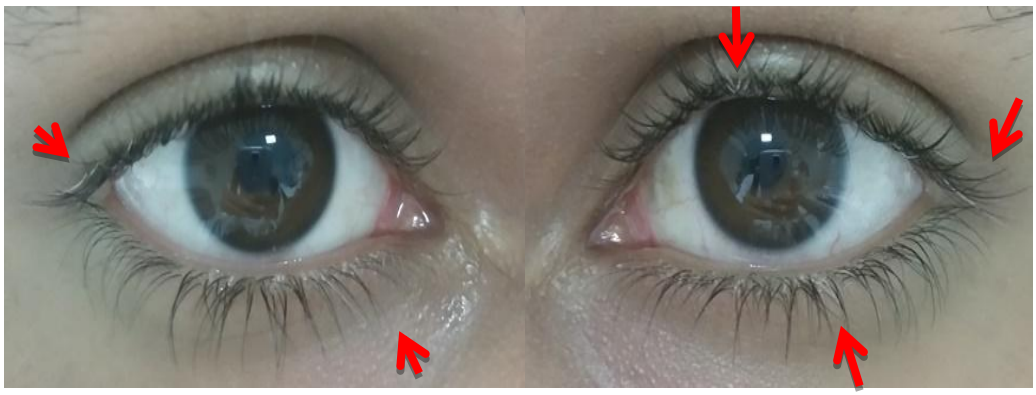

Figure 1: Right and Left Eye Lashes Poliosis

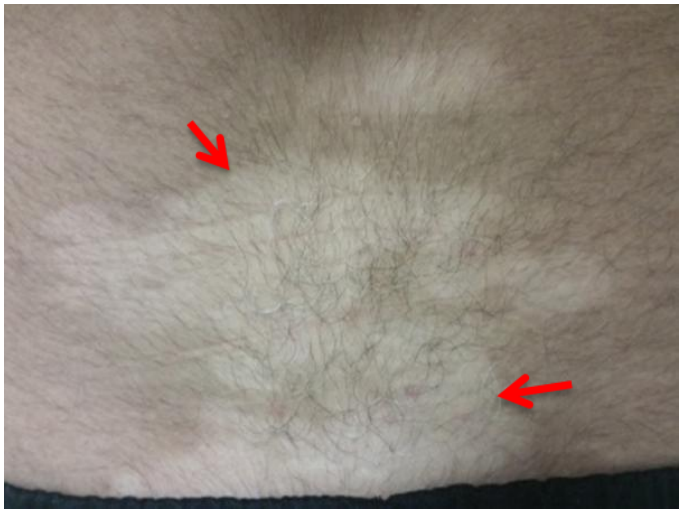

Figure 2: Patient's Lumbar Area Showing Vitiligo Lesions and Poliosis

On examination, the visual acuity in the right eye was 6/18 and the left eye was 6/21. There was bilateral conjunctival injection. Both eyes showed anterior chamber and the vitreous cells 3+ and 2+ respectively. The pupils were reacting sluggishly due to the presence of posterior synechia. The intraocular pressure was 10 mmHg bilaterally. Fundus examination and optical coherence topography showed bilateral disc hyperemia and edema, and exudative retinal detachment involving the macular area with areas of active chorioretinitis, see figure 3. Diagnosis of incomplete Vogt-Kayonagi-Harada syndrome was made. Adjuvant uveitis investigations and MRI were normal. Estimated sedimentation rate was elevated.

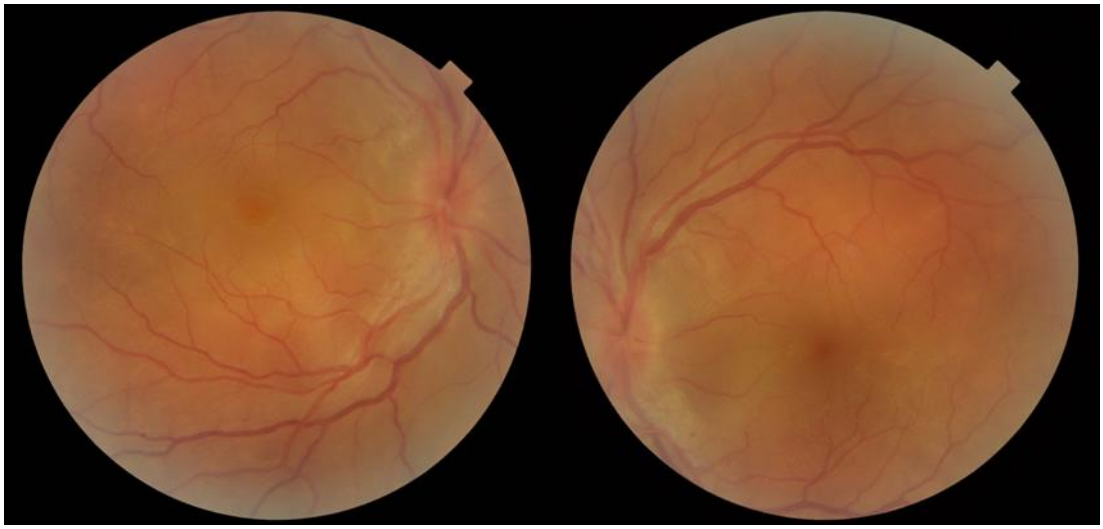

Figure 3: Right and Left Fundus at Presentation

The patient refused admission and was treated as an outpatient with oral prednisolone 1 mg/kg/day for one week with a tapering plan -5 mg/week, topical Pred Forte 1%, and topical atropine 1%. The patient was referred to dermatology and rheumatology. Follow-up of 6 months revealed that the patient is in a remission period with 6/6 vision bilaterally, see figure 4. He was stable on maintenance low-dose steroid course and methotrexate (started by rheumatologist).

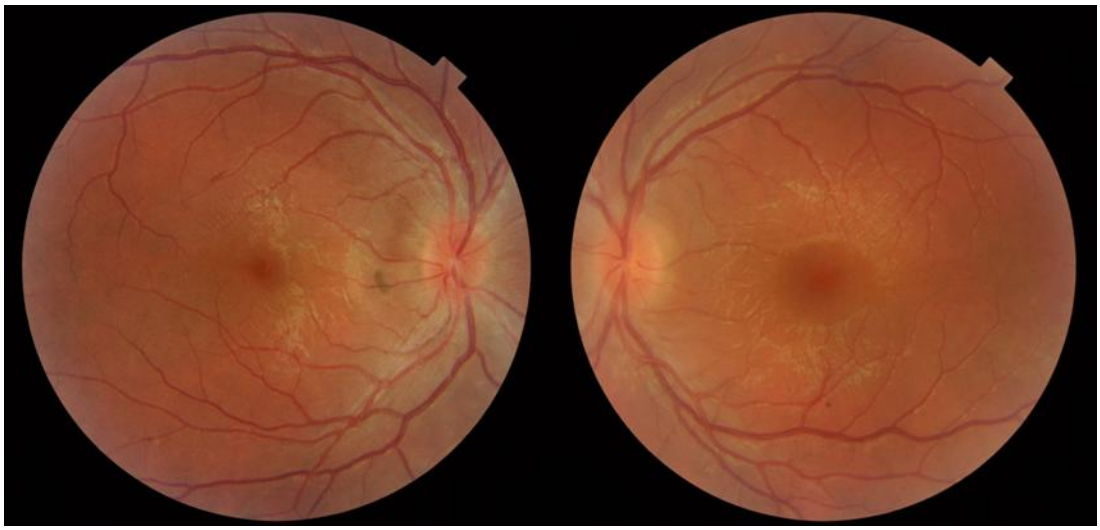

Figure 4: Right and Left Fundus after 3 Weeks of Treatment

DISCUSSION

Up until now, there is no known etiology of VKH syndrome, it is still undetermined. However, many researchers support the theory of autoimmune response to melanocytes[4].

VKH has ocular, neurological and cutaneous manifestations. As a syndrome, it is diagnosed based on criteria recommended by the America Uveitis Society[3].

An international committee on nomenclature published the revised VKH syndrome diagnostic criteria in 2001. They have classified VKH syndrome into three types: complete,

incomplete (either integumentary or neurological involvement) and probable (isolated ocular disease)[1].

VKH syndrome's stages are prodromal or meningeal stage, pan uveitis or ocular stage, convalescent or chronic stage and recurrence stage; auditory symptoms could occur at any of the four stages[3,5].

The diagnosis is made clinically and there are no serological tests available to confirm the diagnosis. However, ocular imaging instruments are used to assess the extent of the disease and to follow up the cases, which include optical coherence tomography, fluorescein angiography and B-scan ultrasonography.

Patients are treated through multidisciplinary approach (Ophthalmology, Neurology, Medicine or ENT). It is principally treated with immunosuppressants. In the acute VKH syndrome, the treatment should start early and aggressively with high dose intravenous prednisolone for 3 to 5 days (1 g daily) or oral dose (1-2 mg/kg/day). The steroid is tapered according to the inflammatory response. Steroids could be used as a maintenance therapy for VKH syndrome. Topical prednisolone acetate 1% is useful only in cases of anterior uveitis[6].

Other immune suppressants could be used in cases of steroid resistance. This might include intravenous immunoglobulins, azathioprine, cyclophosphamide, methotrexate, and cyclosporine[6].

The prognosis of the disease is favorable if treatment started early. Visual outcomes depend on the disease progression to glaucoma and subretinal neovascularization. However, well-treated VKH could preserve normal visual acuity[7].

CONCLUSION

Vogt-Koyanagi-Harada syndrome is a granulomatous autoimmune disease involving central nervous system, eyes, skin, and ears. It is clinically diagnosed according to the criteria set by the America Uveitis Society. Atypical presentation and association with other diseases have been reported. Usually, ocular symptoms follow the neurological symptoms and cutaneous manifestation. If treated early and sufficiently with immunosuppressant, undesired outcomes could be avoided.

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