

Vogt-Koyanagi-Harada Syndrome: A Case Report

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ABSTRACT

Vogt-Koyanagi-Harada Syndrome (VKH), a rare meningo-uveal syndrome is a diagnostic challenge. That there are no confirmatory diagnostic laboratory tests makes a diagnosis one by exclusion. A case of a 50 year old female patient managed as an outpatient at St. Mary's Mission Hospital Eye Clinic is presented. The objective of this presentation is to describe this rare disease entity, the challenges faced in arriving at the diagnosis, the management of the condition and subsequent outcome.

INTRODUCTION

Vogt-Koyanagi-Harada syndrome is one of the Meningio-uveitic syndromes – a constellation of clinical symptoms and signs for which no definitive confirmatory tests are available¹. It presents with a prodromal stage characterised by headaches, meningismus, dysacusis, tinnitus and flu-like symptoms. In the ocular stage, a granulomatous anterior and posterior uveitis occurs. A serous retinal detachment, disc hyperaemia and choroidal lesions may occur too. A third stage is characterised by worsening of the symptoms and signs together with skin and hair depigmentation, alopecia and hearing loss.

It is a systemic disorder and the extraocular findings are most important in securing the diagnosis.

This case is presented for its rarity and the challenges posed in securing a diagnosis.

CASE REPORT

A 50 year old African woman presented to St. Mary's Mission Hospital Eye Clinic with severe right sided headache, nausea and vomiting, painful right eye with blurred vision, tearing and photophobia on the same eye for two weeks. She had developed an itchy hypopigmented patch on her right forehead about two weeks prior to onset of ocular symptoms. The lashes on her upper lid had also turned white. There was neither prior history of ocular disease nor trauma.

Her uncorrected vision was 6/9 both eyes on the Snellen Chart. The intraocular pressure (IOP) by applanation tonometry was 16 and 12mmHg

for the right and left eyes respectively. The right eye showed a hypopigmented area from the brow, nasal 2/3 of the upper lid, nasal fold and the entire lower lid margin (Figure 1). The affected areas showed whitening of the hair and lashes. She had deep ciliary injection (Figure 2) on the right eye, fine keratic precipitates (Figure 3), and an anterior chamber (AC) activity characterised by a dense flare and moderate cells.

Figure 1: Vitiligo and Poliosis in VKH



Figure 2: Ciliary injection in VKH

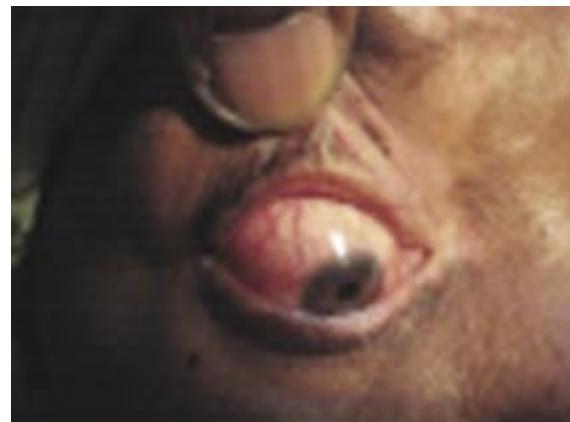
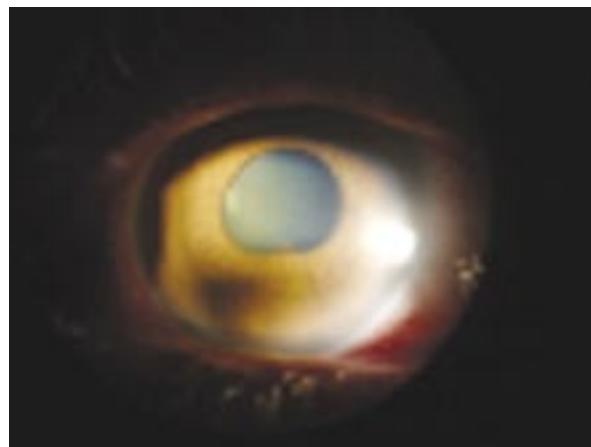


Figure 3: Keratic precipitates in VKH**Figure 4:** Posterior synechiae demonstrated after Tropicamide drops

The pupil was round, reacted sluggishly to light but had no relative afferent papillary defect (RAPD). Dilating the pupil using tropicamide 1% revealed some posterior synechiae (Figure 4). There was no vitreous activity and the disc was slightly hyperaemic.

Further inquiry on past medical history revealed nothing of significance. The parameters of a complete blood count (CBC) were all within normal, an erythrocyte sedimentation rate (ESR) of 14mm/Hr, as well as a negative VDRL for syphilis.

A diagnosis of severe anterior uveitis with concomitant vitiligo was made and treatment for uveitis using prednisolone acetate 1% q2hr and atropine sulphate 1% OD was commenced.

The patient was reviewed four days later and both the symptoms and signs had worsened. The patient complained of increased pain and poorer vision. She had frontal headaches and

moderate neck pains especially on flexion and extension. She was not vomiting but said she was generally tired.

Ocular examination revealed reduced visual acuity on the RE, 6/24, an increase of flare and cells and a mid dilated pupil. IOP in the affected eye remained normal at 14mmHg. There was marked disc oedema with pale-white linear retinal lesions (Figure 5) in the outer temporal fundus that appeared raised above the adjacent retina. The left eye remained quiet.

Figure 5: Retinal lesion in VKH

There were no focal or generalised CNS signs. The cranial nerves were also normal. At this point she was started on high dose oral steroids: prednisolone 40mg b.i.d. The topical treatment continued with the frequency of atropine drops upped to q.i.d. Prophylactic ranitidine 150mg b.i.d was also added to the treatment. She was asked to return on the fourth day for further review.

On the third visit, the patient reported a marked relief on ocular pain. The headaches, neck pain and the general malaise had also disappeared. Visual acuity RE was 6/18 and LE 6/9 uncorrected. The keratic precipitates were less and there was a marked reduction in the amount of flare and cells in the AC. The retinal lesions remained as in the previous visit. IOP right eye was normal at 12mmHg. A decision was made to continue the steroid therapy at the same dose for a further five days before tapering it off gradually to 30mg b.i.d for 5 days.

She was reviewed a week after the third visit when she complained of glare but no pain. Her VA remained stable at 6/18 and IOP 14mmHg.

The anterior segment had no activity but the retinal appearance remained the same. Steroid therapy was continued at 30mg b.i.d for 5 days to taper off to 30mg OD there after. Atropine drops were discontinued at this time.

Total daily dose of the steroids was tapered down to 20mg OD when she developed dyspepsia and abdominal bloating at 3 weeks. She tolerated this dose well into the 5th week post presentation when she reported that her vision had cleared and she was not in any pain or discomfort (VA 6/9 BE). The anterior segments were quiet. Disc swelling was still present but reduced. Retinal lesions however remained the same.

After this encounter, unfortunately, she was lost for follow-up.

DISCUSSION

Uveitis is a common presentation in eye clinics though a definitive aetiology is seldom found. The case presented shows the empirical way uveitis management is frequently undertaken and a call for a high index of suspicion in case of unfamiliar associated symptoms and signs.

In 1906, Vogt described a case of anterior uveitis associated with poliosis and dysacusis. In 1929, Koyanagi discussed the details of six cases in Japan, all with symptoms similar to the case described by Vogt, but also including alopecia and vitiligo. In 1926, Harada reported on a second condition that included posterior uveitis and exudative retinal detachment associated with a cerebrospinal fluid pleocytosis and meningeal signs. Because of the similarities and overlapping features of both clinical conditions, many investigators considered these diseases to be related, hence the name Vogt-Koyanagi-Harada syndrome (uveo-encephalitis)².

Prevalence: The worldwide prevalence of Vogt-Koyanagi-Harada syndrome is not easy to assess because this syndrome occurs more frequently among Asians. In Japan, approximately 8% of uveitic patients have Vogt-Koyanagi-Harada syndrome, with an incidence of 800 new cases per year. Ohno and co-workers³ reported that 1% of 5500 uveitic patients were diagnosed with Vogt-Koyanagi-Harada syndrome, of whom 41% were Asian, 29% were white, 16% were Hispanic, and 14% were African-American. Snyder and Tessle⁴ found that 3.7% of 455 patients seen in their uveitis clinic had Vogt-

Koyanagi-Harada syndrome, 75% of whom were either of Native American or Hispanic decent; none were Asian. In the National Eye Institute⁵ a series of 78 patients, 44% of the patients with this syndrome were African-American, 37% white, 11% Hispanic, and 6% Asian. Of the 65 patients with Vogt-Kayanagi-Harada syndrome seen in Doheny Eye Institute⁶, 51 (78%) were Hispanic, 7 (10%) Asian, 4 (6%) African-American, 2 (3%) white, and one Native American. Overall, no significant sex differences have been found in this syndrome. North America, however, females seem to be affected more frequently than males. This syndrome typically affects adults between the ages of 20 and 50 years.

Clinical manifestations: The progression of Vogt-Koyanagi-Harada syndrome used to be divided into three stages, but is now divided into four:

Stage 1: The prodromal stage

This occurs a few days before the ocular symptoms and is characterized by headaches, deep orbital pain, vitiligo, nausea, slight fever, and occasional photophobia and lacrimation. Cerebrospinal fluid may show pleocytosis.

Stage 2: The ophthalmic or uveitis stage

This is characterized by bilateral uveitis that may affect one eye first with sudden blurring vision, photophobia, impairment of ocular movement, dysacusis, and meningism. This stage can last weeks to months. On ocular examination, panuveitis can be recognized by mutton-fat keratic precipitates, aqueous cells and flare, iris nodules, synechiae, vitreous haze and cells, optic disc swelling, retinal edema, haemorrhages, exudate and non-rhegmatogenous detachment, choroidal infiltrates, and infiltrates at the retinal pigment epithelial level. Approximately 15% of patients may note skin hyperesthesia.

Stage 3: The convalescent (recovery) or chronic stage. This stage may last from weeks to months, or it may continue chronically. It is characterized by subsiding uveitis with possible visual and neurologic improvement, but continuous dysacusis (especially to high-frequency sound), poliosis, vitiligo, and alopecia. Ocular examination reveals depigmentation of the perilimbus (Sugiura's sign) and a pale fundus (sunset-glow fundus). In general, the retina has reattached with a mottled appearance and with white spots interspersed with a pattern of mottled pigmentation. The periphery

and equator show focal depigmentation. Sometimes strands of chorioretinal scarring with pigmentation or depigmentation can be observed. Occasionally, sub-retinal neovascular membrane and subretinal fibrosis develop.

Stage 4: The recurrent stage. This stage of Vogt-Kayanagi-Harada syndrome usually involves complications such as mutton-fat keratic precipitates, iris Koeppe's nodules, synechia, cataract, glaucoma, subretinal neovascularization, and subretinal fibrosis.

Treatment: Currently, the most valuable therapeutic medication for Vogt-Koyanagi-Harada syndrome is systemic corticosteroids⁷. As in sympathetic ophthalmia, an early and aggressive high-dose steroid regimen at the beginning of the disease, followed by gradual tapering of the dosage, is recommended for successful treatment. The early administration of systemic steroids may prevent the progression of the disease, shorten the duration, and decrease complications and other systemic involvement. Other immunosuppressive medications, such as chlorambucil and cyclosporine, have had positive effects in patients in whom steroid therapy failed. Before one chooses any of these

medications, however, their side effects must be considered and carefully monitored. Topical steroids and cycloplegics alone do not suffice.

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INVESTING IN PEOPLE IS AN INVESTMENT FOR THE FUTURE FRED HOLLOWES EASTERN AFRICA MOVES A NOTCH HIGHER

Kimani G –Regional Manager FHEA

"Dependency should not occur. Being a parent consists of raising a dependent person and creating an independent person. In running a blindness programme in the developing world, the aim is to have it so that it can run itself at the highest possible level." Fred Hollows said this over a decade ago; it stands true today as it will some hundreds of years to come.

In a world full of competing priorities, making the right choice of where to invest and when to invest is important in determining the end results as well as the impact of the available resources. In the past five years, Fred Hollows Eastern Africa (FHEA) has invested in one of the most valuable pillars of the V2020 initiative, - Human Resource Development. FHEA recognises that the training and equipping eye care workers is the engine oil that will effectively drive the V2020 partnership in the attainment of the V2020 goal; a world where needless blind is eliminated.

Ten years ago, FHF revolutionised cataract surgery through the introduction and training of eye surgeons in the Intra Ocular Lens implantation. This simple technology has over the years pushed the quality of cataract surgery outcome to high standards with more cataract patients having the benefit of discarding the once familiar aphakic glasses. In the four Eastern African countries (Kenya, Tanzania, Rwanda and Burundi) FHEA has partnered with the respective governments and relevant institutions in equipping different cadres of eye care with skills that are positively affecting the outputs/outcomes at their respective work place.

In Tanzania and over the years, FHEA has worked closely with Kilimanjaro Centre for Community Ophthalmology (KCCO) in training eye care managers and ophthalmologists drawn from East African countries. The objective of training the eye care managers is to develop a cadre of eye care staff that manages the day to day running of the eye programmes; therefore releasing the much needed skills and valuable

time of the clinical officers and ophthalmologists to treat eye diseases. The aim of training the ophthalmologists in the management of eye care programmes is to equip them with skills that enable them to support the managers and ensure the latter use their skills optimally. To this end and over the last four years, the Foundation in partnership with KCCO has trained approximately 70 eye care managers and ophthalmologists.

Similarly and with KCCO, FHEA has been involved in the training of eye care workers who have expressed interest in research methodologies. Over the years the interest in research in eye care has grown. The practitioners as well as the donors have been keen to plan their eye care programmes on evidence. There has also been a growing need to ensure that lessons learnt during the implementation of eye care programmes within the region are documented and used as a resource in planning eye care programmes. To this end, the Foundation has supported KCCO in training the researchers in writing and publishing their work. This skill is as important as the research itself, since without publishing research findings, the research is rendered useless. Evidence based planning is arguably the best tool of advocacy and with this recognition on hand, FHEA has recently appointed a full time researcher based in Melbourne to work closely with the field teams.

With Trachoma being a major threat among the avoidable blindness diseases in Eastern Africa, FHEA has taken a keen interest in supporting the training and equipping of clinical officers in lid surgery. While lid surgery is simple and straight, the lack of skills on the part of health care providers as well as lack of appropriate instruments may hinder the progress of combating blindness due to trachoma. Over the past five years, the Foundation has been involved in training and equipping 26 lid surgeons in Kenya.

As part of ensuring that the cataract surgeons graduating from Kenya Medical Training College

are well equipped whilst posted in the field, the Foundation has ensured that where applicable, the surgeons are equipped with operating microscopes. This ensures that they not only perform the surgeries, but their skills are utilised whilst in the field.

While it is fashionable to have new equipment and instruments in place when the old ones break down, the Foundation with other partners have over time realised that the costs of new equipment is prohibitive in running sustainable eye care programmes. Subsequently, we have invested in the training and equipping of a mobile Government technician who undertakes the repair work as the need arises in the static eye units. The Foundation will in the coming year in collaboration with the Ministry of Health in Kenya establish a static equipment/instrument repairs workshop in Mbagathi

District Hospital in Nairobi, Kenya. This will be a one stop garage for eye care equipment and instrument repairs.

Over the years, the Foundation has supported the Kenya National Ophthalmic Workshop (KNOW). This workshop brings together once in a year, the mid level eye workers (MLEW) in Kenya for planning and continuous medical education. With other participating partners, the Foundation is committed to support the KNOW and the OSEA conference in a thrust to ensure that skills development in eye care is continuous.

As a Foundation we seek to promote sustainable eye care programmes and we are committed to investing in the training of eye care workers to ensure a solid foundation of eye care in the region.

