References


and located superiorly. Based on the modified Callendar classification for the cytologic classification of uveal melanomas, a diagnosis of mixed-cell-type choroidal melanoma was made (Figure 1).

The 2004 report of the Collaborative Ocular Melanoma Study (COMS) classified posterior uveal melanomas according to basal diameter and thickness measured by ultrasound. An anteroposterior thickness of 9.9 mm, which this patient had, was classified as large. Several treatment options are available, primarily based on tumor size. Enucleation is indicated for tumors greater than 15 mm in diameter and greater than 10 mm in thickness, and in patients who already have irreversible loss of useful vision. Neo-adjuvant and adjuvant therapies offer no 5-year survival advantage over enucleation alone.

Follow-up plans for this patient included liver-function test every 3 months, liver ultrasound every 6 months, and artificial-eye implantation after 2 months.

Figure 1. Low-power view showing darkly pigmented choroidal mass (A) and high-power field showing mixed population of cells (B).

Vogt-Koyanagi-Harada in a Kadazan female

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ABSTRACT

Objective
To report a case of Vogt-Koyanagi-Harada in a Kadazan girl, a member of an indigenous race of the state of Sabah, Malaysia.

Methods
This is a case report.

Results
A 23-year-old Kadazan female presented with bilateral sudden blurring of vision of two days duration associated with ocular pain, metamorphopsia, and severe headaches. Examination revealed bilateral visual acuities of 6/18 correctable to 6/12, 1+ anterior-chamber cells, and multifocal areas of exudative retinal detachments. A diagnosis of Vogt-Koyanagi-Harada (VKH) syndrome was made after excluding other differential diagnoses. She was treated with intravenous methylprednisolone with good outcome.

Conclusion
The treatment for VKH is well established, requiring the use of oral steroids in most cases. In severe cases, high-dose intravenous methylprednisolone is recommended. Early diagnosis and aggressive treatment improve outcome in VKH.
A 23-YEAR-OLD Kadazan female consulted at the Penang Hospital Eye Clinic for sudden blurring of vision in both eyes of 2 days duration, associated with redness and dull eye pain, metamorphopsia, and severe headaches. There was no history of tinnitus, hearing difficulty, ocular trauma, surgery, or menstrual problem. Family history was unremarkable. Except for gastritis, there were no other health problems.

Physical and neurological examinations were unremarkable. Ocular examination revealed bilateral visual acuity of 6/18 improved to 6/12. Pupils were round and equally reactive to light and accommodation. Corneas were clear, intraocular pressures were normal, and the conjunctivae were mildly injected. The anterior chambers contained 1+ cells.

Posterior-segment examination revealed clear vitreous and normal-looking optic discs. Both posterior poles exhibited multifocal areas of exudative retinal detachment (Figure 1). B-scan ultrasonography showed diffuse choroidal thickening (Figure 2). Fluorescein angiography (FA) showed early multiple pinpoint areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments.

Extensive laboratory examinations were done to rule out other ocular diseases: tuberculosis, syphilis, and sarcoidosis were excluded via chest X-ray and rapid-plasma-reagent test; connective tissue diseases via a thorough medical evaluation, negative-serum-rheumatoid factor, and antinuclear antibodies; breast and cervical cancers by gynecologic evaluation. Computed tomography (CT) of the brain and lumbar tap were normal. Hearing assessment and pure-tone audiometry were also normal.

An initial diagnosis of Vogt-Koyanagi-Harada (VKH) syndrome was made based on the presence of bilateral uveitis, multifocal serous retinal detachment, and severe headache. Under the criteria set following the first international workshop on VKH in 1999 (Table 1), the diagnosis of VKH requires the exclusion of penetrating ocular trauma or intraocular surgery (which may suggest sympathetic ophthalmia) and other ocular diseases, both of which were excluded in this patient. Since neurological, auditory, and dermatological features of VKH were absent in this patient, a diagnosis of “probable VKH” was made based on three of the five criteria (1, 2, and 3). The features in this case were similar to those described by Harada, with mild anterior uveitis, multifocal choroiditis, and possible meningismus. The missing diagnostic criteria were the integumentary features. However, skin lesions are often not present in Harada’s (<10%). Moreover, visual recovery generally is good in contrast to the poor visual outcome of Vogt-Koyanagi syndrome. This patient’s visual acuity improved to 6/9 with treatment.

VKH syndrome is a disorder affecting mostly Asians (Japanese and Chinese), Hispanics, and Native Americans (particularly Cherokee Indians). It generally occurs in individuals between 20 and 50 years old. The youngest reported case involved a 4-year-old. Both sexes are affected, but Rubsamen and Gass found more females in their case series.

VKH is a multisystem, granulomatous inflammatory disorder affecting the eyes, meninges, skin, and auditory system. Vogt and later Koyanagi described patients with bilateral anterior uveitis, vitiligo, poliosis, alopecia, and dysacusia; Harada described patients presenting with posterior uveitis, exudative retinal detachment, and cerebrospinal-fluid pleocytosis. It is now accepted that these two entities are spectrums of the same disease, currently termed VKH syndrome. The etiology is largely unknown. The pathogenesis is related to an immune reaction against uveal tissue, similar to sympathetic ophthalmia. The underlying mechanism of tissue damage is probably a Type IV autoimmune response against uveal antigens, possibly associated with melanocytes. Histopathology shows uveal infiltration with B and T-lymphocytes, multinucleated giant cells, and lymphoid nodules. The most common features seen are patchy choroidal atrophy, with focal areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments. Histopathology shows necrotizing uveal, subretinal, and choroidal inflammation with granulomatous infiltrates of lymphocytes, monocytes, and histiocytes. The pathogenesis is related to an immune reaction against uveal antigens, possibly associated with melanocytes. Histopathology shows uveal infiltration with B and T-lymphocytes, multinucleated giant cells, and lymphoid nodules. The most common features seen are patchy choroidal atrophy, with focal areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments. Histopathology shows necrotizing uveal, subretinal, and choroidal inflammation with granulomatous infiltrates of lymphocytes, monocytes, and histiocytes. The pathogenesis is related to an immune reaction against uveal antigens, possibly associated with melanocytes. Histopathology shows uveal infiltration with B and T-lymphocytes, multinucleated giant cells, and lymphoid nodules. The most common features seen are patchy choroidal atrophy, with focal areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments. Histopathology shows necrotizing uveal, subretinal, and choroidal inflammation with granulomatous infiltrates of lymphocytes, monocytes, and histiocytes. The pathogenesis is related to an immune reaction against uveal antigens, possibly associated with melanocytes. Histopathology shows uveal infiltration with B and T-lymphocytes, multinucleated giant cells, and lymphoid nodules. The most common features seen are patchy choroidal atrophy, with focal areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments. Histopathology shows necrotizing uveal, subretinal, and choroidal inflammation with granulomatous infiltrates of lymphocytes, monocytes, and histiocytes. The pathogenesis is related to an immune reaction against uveal antigens, possibly associated with melanocytes. Histopathology shows uveal infiltration with B and T-lymphocytes, multinucleated giant cells, and lymphoid nodules. The most common features seen are patchy choroidal atrophy, with focal areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments. Histopathology shows necrotizing uveal, subretinal, and choroidal inflammation with granulomatous infiltrates of lymphocytes, monocytes, and histiocytes. The pathogenesis is related to an immune reaction against uveal antigens, possibly associated with melanocytes. Histopathology shows uveal infiltration with B and T-lymphocytes, multinucleated giant cells, and lymphoid nodules. The most common features seen are patchy choroidal atrophy, with focal areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments. Histopathology shows necrotizing uveal, subretinal, and choroidal inflammation with granulomatous infiltrates of lymphocytes, monocytes, and histiocytes. The pathogenesis is related to an immune reaction against uveal antigens, possibly associated with melanocytes. Histopathology shows uveal infiltration with B and T-lymphocytes, multinucleated giant cells, and lymphoid nodules. The most common features seen are patchy choroidal atrophy, with focal areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments. Histopathology shows necrotizing uveal, subretinal, and choroidal inflammation with granulomatous infiltrates of lymphocytes, monocytes, and histiocytes. The pathogenesis is related to an immune reaction against uveal antigens, possibly associated with melanocytes. Histopathology shows uveal infiltration with B and T-lymphocytes, multinucleated giant cells, and lymphoid nodules. The most common features seen are patchy choroidal atrophy, with focal areas of hyperfluorescence and late pooling of fluorescein indicating multiple serous detachments. Histopathology shows necrotizing uveal
cells, epitheloid cells, and plasma cells suggesting that both cell-mediated and humoral immune mechanisms are involved.\(^9\) Norose and Yano demonstrated the involvement of cytotoxic T lymphocytes.\(^10\) Belfort et al. showed that T lymphocytes are the predominant cell type in aqueous of patients with VKH.\(^11\) Association with several leukocyte antigens such as HLA-DR4, HLA-DRw52, HLA-DRw53 and HLA-DRw54 have been demonstrated.\(^3\)–\(^5\), 12, 13 Genetic predisposition is also suggested by cases of monozygotic twins presenting with the disease.\(^14\)

VKH follows four clinical stages: prodromal, uveitic, chronic, and recurrent.

Prodromal stage is characterized by headaches, meningismus, vertigo, deep orbital pain, nausea, slight fever, photophobia, and lacrimation lasting for a few days. Cerebrospinal-fluid (CSF) pleocytosis occurs in 80% of patients during this stage. Melanin-laden macrophages are responsible for the pleocytosis.\(^15\)

Uveitic stage, which follows the prodromal stage, is characterized by sudden bilateral blurring of vision with choroiditis, exudative retinal detachments as seen in this patient, disc swelling, mutton-fat keratic precipitates, and iris nodules, which may last for several weeks.

Chronic stage, which may last for months to years, is the convalescent phase when uveal and dermatologic depigmentation occurs. Sugita’s sign or perilimbal vitiligo is the earliest depigmentation seen. Depigmentation of the choroid may occur resulting in the “sunset glow” fundus. Dalen–Fuchs nodules may also be present.

Recurrence, which may interrupt the chronic stage, is often associated with anterior, rarely posterior, uveitis.

Investigations useful in diagnosing VKH are FA showing multiple areas of punctate hyperfluorescence and B scan demonstrating thickening of the choroid, sclera, or episclera posteriorly.\(^16\) Indocyanine green (ICG) shows dark background in the early phase, indicating delayed choriocapillary perfusion and nonuniform hypofluorescent lesions in midphase that persist in the recovery.\(^17\)

After infective etiologies were excluded, the patient was given intravenous methylprednisolone 500 mg every 12 hours 3 days after admission, together with topical betamethasone and homatropine 2%. After 5 days of methylprednisolone, there was complete resolution of the subretinal fluid in the right eye and near-complete resolution in the left. The headaches were also relieved.

The patient was subsequently discharged and prescribed oral prednisone at tapering doses.

After 6 months, her vision was 6/9 in both eyes. Some
pigmentary disturbances at the posterior pole were present with no evidence of active choroiditis or subretinal fluid.

The treatment for VKH is well established, requiring the use of a steroid in some cases for up to 1 year to prevent recurrence. Oral prednisone may be used, but in severe cases high-dose intravenous methylprednisolone is recommended. In steroid-resistant cases, cyclosporin A has been successfully used. Refractory cases have also been documented to respond to newer immunosuppressive agents such as FK506, which has been isolated from the fermentation broth of Streptomyces tsukubaensis. Long-term studies, however, are needed to fully evaluate the efficacy and safety of this drug.

To our knowledge, this is the first reported case of VKH among the Kadazans, the largest ethnic group in Sabah, a state of Malaysia north of Borneo.

Early diagnosis and aggressive treatment improve outcome. Treatment must be initiated early to prevent chronicity, which may result in such complications as retinal and disc neovascularisation that may lead to vitreous hemorrhage and tractional retinal detachment, choroidal neovascularisation, cataract, and glaucoma.

References

ABSTRACT

Objective
To report a case of acute post-cataract-surgery endophthalmitis after suture removal.

Methods
This is a case report.

Results
A 77-year-old Chinese male presented with sudden painless blurring of vision in the left eye (OS) of 3 days duration 22 days after cataract surgery and 15 days after corneal-suture removal. OS was injected with corneal striae and had a visual acuity of 1/60; 3+ anterior-chamber cells with small hypopyon, and hazy vitreous. Endophthalmitis was considered and immediate vitreous tap with intravitreal antibiotics were given. Intensive topical antibiotics were instituted, followed by a repeat intravitreal antibiotic injections 3 days later. Postoperatively, there was massive fibrin formation with cyclitic membrane and sequelae pupillae that required two peripheral iridotomies. Visual acuity slowly recovered from hand movement to 6/18.

Conclusion
Endophthalmitis can be successfully treated without pars plana vitrectomy, following the Endophthalmitis Vitrectomy Study (EVS)