

CASE REPORT



Smoldering Vogt-Koyanagi-Harada disease with extra ocular involvement — A case report

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Abstract

To report a case of smoldering vogt-koyanagi-harada disease with extra-ocular involvement. A 45-years-old male presented with diminished vision in BE for one 1 — week associated with redness, pain, watering, intolerance to light with tinnitus also gives a history of similar complaints, in the past 2years. Examination revealed both eye visual acuity CF1/2 meters. Slit-lamp examination showed conjunctival ingestion, mutton fat KP's, sluggishly reactive pupil with multiple broken posterior synechiaie. Fundus showed grade 2 vitreous cells with multiple opacities, diffuse retinochoroidal thickening with disc edema. Tonometry was 16 and 29 mmhg respectively. B-scan showed multiple vitreous echo's, diffuse retino-choroidal complex thickening with multiple shallow serous retinal detachment. After routine investigations patient was started on topical and systemic steroids. Lumbar puncture showed 82% lymphocytic pleocytosis. The patient recovered well with 6/24 vision in BE with a resolution of fundus finding at 6week followup. High index of suspicion, timely diagnosis and management are crucial to prevent vision threatening complications in VKH disease.

Keywords: Vogtkoyanagiharada disease; retinochoroidal thickening; serous retinal detachment; tinnitus; CSF pleocytosis; systemic steroids

Introduction

The uveomeningeal syndromes are a heterogeneous group of disorders or conditions that share a common characteristic, the affection of the retina, uvea and meninges,⁽¹⁾ their causes can be infectious, due to neoplastic process or to systemic diseases. This is a systemic autoimmune disease characterized mainly by

a bilateral granulomatous panuveitis, in which antibodies are produced against tissues rich in melanocytes such as the retina, the inner ear, the meninges, skin and hair,⁽²⁾ which explains its extraocular manifestations and its clinical presentation. Occurs more commonly in darkly pigmented races. Onset is usually in the 2nd to 5th decade of life VKH disease has 4 clinical phases of presentation

consisting of the following.⁽³⁾

Prodromal phase — Nonspecific symptoms among which are fever, headache, nausea, meningism, vertigo and disacusia, rarely include ataxia, confusion and neurological focality. It usually lasts 3 to 5 days.

Acute uveitic phase — Decreased visual acuity, usually unilateral onset, then contralateral with a difference of 1-3 days, photophobia, iridocyclitis, retrokeratic deposits, ocular hypertension, vitritis. Choroiditis that results in rupture of the external hematorretinal barrier (pigmented epithelium of the retina), causing exudative retinal detachments. It can cause acute glaucoma. It lasts several weeks.

Convalescence phase — Retinal detachments gradually subside and uveitis begins to subside. There is a pigmentary redistribution that gives rise to an eye fund "like sunset" (sunset glow fundus). It can last for months.

Chronic recurrent phase — It can interrupt the convalescent phase with recurrent anterior uveitis. There is the formation of synechiae, pupillary block, iris atrophy, cataracts and glaucoma.

Systemic manifestations

Neurological symptoms: Headache, meningism, encephalitis, paralysis of the cranial nerves, aphasia, loss of consciousness and seizures. They usually appear in the prodromal phase.

Auditory symptoms: Tinnitus, hipoacusia and vertigo. They can be present in all phases.

Dermatological manifestations: Vitiligo, alopecia and poliosis; generally in the convalescence phase.

The diagnosis is made through clinical criteria established and published by the American Uveitis Society in 2001.⁽²⁾ The treatment is mainly based on systemic corticosteroids during the acute phase. Other treatment options include immunosuppressants such as azathioprine, cyclosporine, methotrexate and the use of immunoglobulins,^(3,4) and the use of monoclonal antibodies such as adalimumab has been documented, which is associated with a lower risk of recurrence and visual impairment.⁽⁵⁾ The complications that occur are related to the duration of the disease, recurrences and the age of the patient at the time of suffering from the disease. The complications that usually occur are cataracts, glaucoma, choroidal neovascularization and subretinal fibrosis.⁽⁶⁾

Case report

A 45-years-old male, farmer presented with c/o diminished vision in BE for 1 week associated with redness, pain, watering, intolerance to light and tinnitus which increases during night hours. Also, gives a history of similar complaints in the past 2years. Examination revealed both eye visual acuity CF1/2 meters. Slit lamp examination showed conjunctival ingestion, mutton fat KP's on the posterior cornea, sluggishly reactive pupil with multiple broken posterior synechiae. Fundus showed hazy media with grade 2 vitreous cells, multiple vitreous opacities, diffuse retinochoroidal thickening, and multiple shallow serous retinal detachment with disc edema. Tonometry was 16 and 28 mmhg respectively.

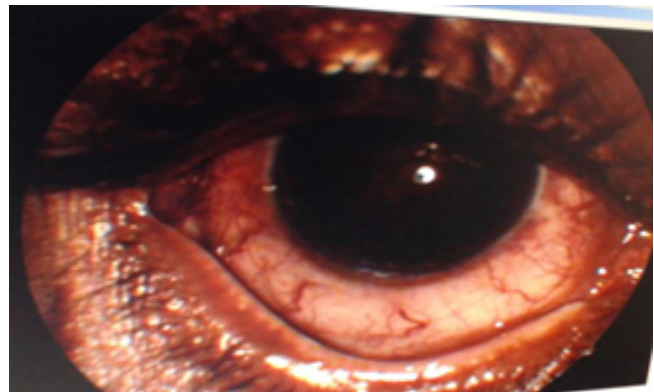


Fig 1. Anterior segment showing conjunctival ingestion



Fig 2. Slit lamp picture showing broken posterior synechiae

B-scan showed multiple vitreous echo's, diffuse retinochoroidal complex thickening with multiple shallow serous retinal detachment, with optic disc edema.

Lumbar puncture showed 82% lymphocytic pleocytosis.

After routine investigations patient was started on topical and systemic steroids, cyclopegics and antiglaucoma drugs.



Fig 3. Slit lamp picture showing multiple mutton fat KP'S

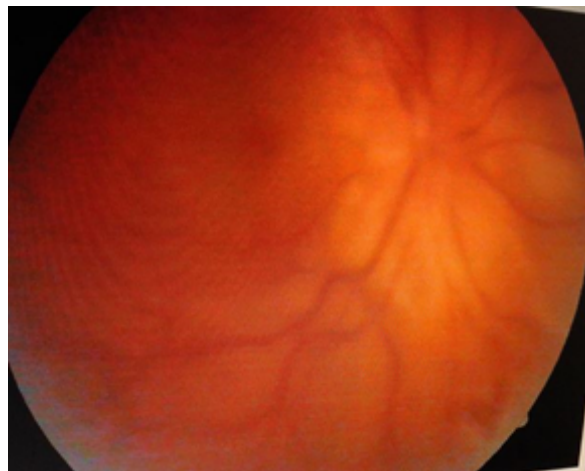


Fig 4. Fundus picture showing diffuse retinochoroidal thickening

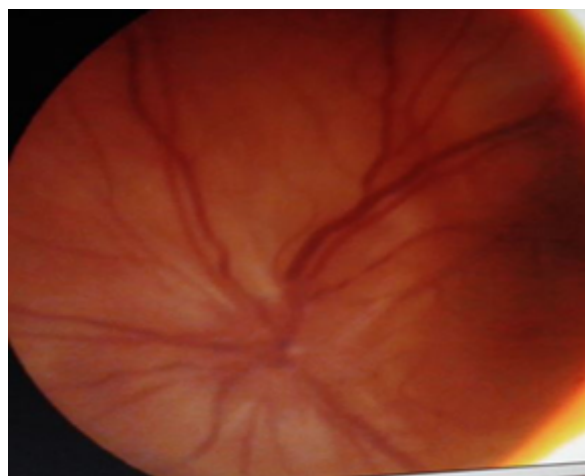


Fig 5. Fundus picture showing disc edema

IV methyl prednisolone 1mg/kg for 3days followed by oral steroids in a tapering dose. The patient recovered well with 6/24 vision in both eyes with resolution of fundus and b-scan findings at 6week followup.

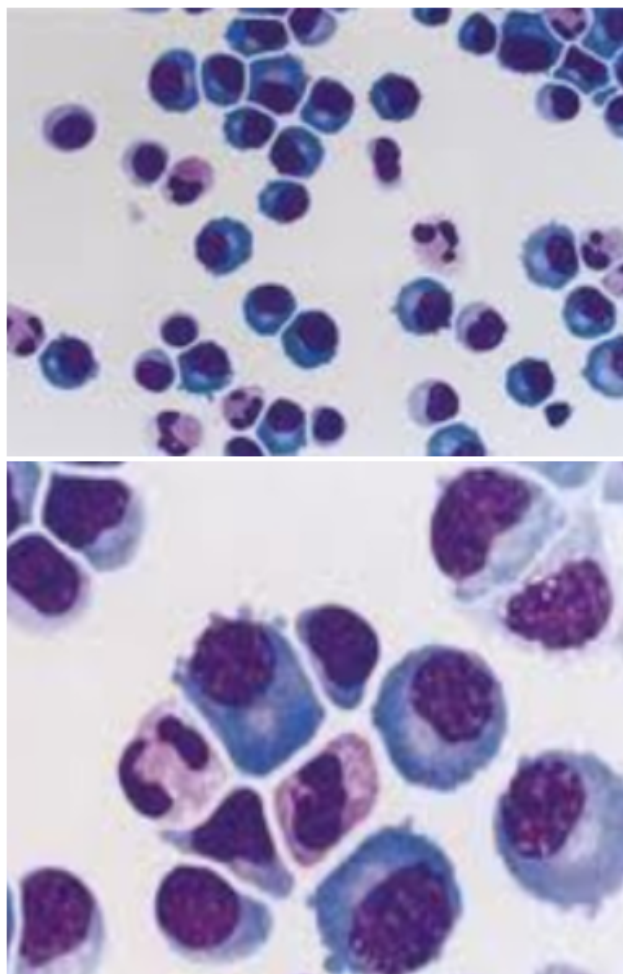


Fig 6. CSF pleocytosis

Discussion

The reported clinical case meets the criteria to classify it as incomplete VKH disease, as it has ophthalmological implications that do not have a history of trauma or ocular surgery prior to the onset of symptoms, absence of evidence suggestive of another ocular disease, bilateral ocular involvement, and auditory manifestations, supporting the diagnosis in the presented imaging studies and intense pleocytosis in the cerebro-spinal fluid. It is not possible to detect dermatological manifestations since these are usually characteristic of the convalescence phase and are almost always detected at follow-up appointments.^(7,8)

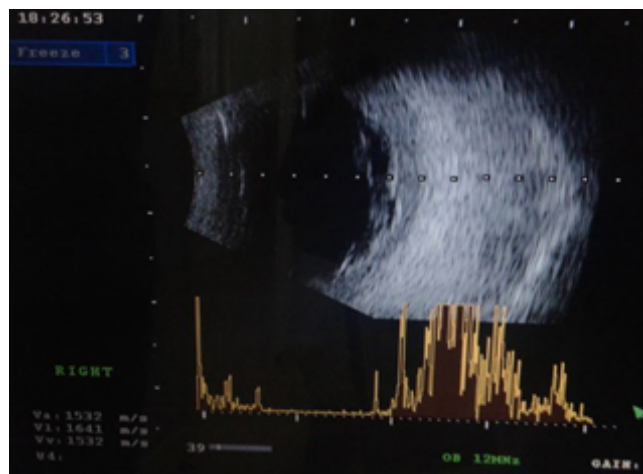


Fig 7. Diffuse RCC thickening with multiple serous RD

Inability to follow the patient deprives us of prognostic data of the particular case, as well as the detection of late-onset sequelae and their subsequent study.

Conclusion

High index of suspicion, timely diagnosis and management are crucial to prevent vision threatening complications in

VKH disease. The rarity of this syndrome makes its diagnosis a challenge, the presence of tinnitus and CNS findings must be considered. Long term followup is crucial for the detection of late onset sequelae and subsequent diagnosis of complete VKH disease.

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