■ *Case report*

Probable Vogt-Koyanagi-Harada's Syndrome associated with tonic pupils

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Abstract

Background: Pupillary abnormalities with complete or incomplete form of VKH have rarely been reported . We report a case of "probable" Vogt- Koyanagi-Haradas (VKH) disease associated with tonic pupils.

Case: A young healthy male presented with 15 days' history of bilateral decrease in vision. The patient had bilateral panuveitis with exudative retinal detachment associated with tonic pupils. There were no other systemic associations. The ultrasonography and fluorescein angiography findings were consistent with VKH disease.

Conclusion: Pupillary reaction abnormalities though uncommon could be seen in association with "probable" VKH disease.

Keywords: probable Vogt-Koyanagi-Haradas disease, pupillary abnormalities

Introduction

Vogt-Koyanagi-Harada (VKH) disease is an autoimmune response of the body against melanocytes of the uveal tissue, skin, inner ear and meninges. Ocular findings usually occur in association with signs and symptoms of meningeal irritation. Rarely have pupillary abnormalities with complete or incomplete form of VKH been reported. We report a case of "probable" VKH disease in association with tonic pupils. A computerized medline search did not reveal any previous report of such association.

Case report

A twenty-eight-year-old male presented with complaints of sudden painless decrease in vision of 15 days duration in the right eye followed by the left eye. There was no history of any trauma or surgery in the past. There was no other remarkable ocular or systemic history. Systemic examination with special reference to the central nervous system, skin and auditory system was essentially normal. Ocular examination revealed that the patient could count fingers at 1meter distance with both eyes with accurate projection of rays. The pupils were dilated and not reacting to light or near stimulus. The corneal sensations, as tested by wisp of cotton, were normal. The ocular movements were full. Slit-lamp biomicroscopy showed a clear cornea, anterior chamber inflammatory cellular reaction of + 2 and flare of +2 in both the eyes. There were no keratic precipitates, granulomas or synechiae. Posterior segment examination using indirect ophthalmoscopy revealed vitreous cellular reaction of + 2 and media clarity of grade II. The optic discs were normal, vessels were mildly tortuous, and retina showed exudative retinal detachment with shifting of fluid. (Figure 1a & 1b).

Fig 1a: Fundus montage picture of the right eye showing clear media, normal disc and inferior exudative retinal detachment, which can be appreciated as inferior retinal vessel blur.

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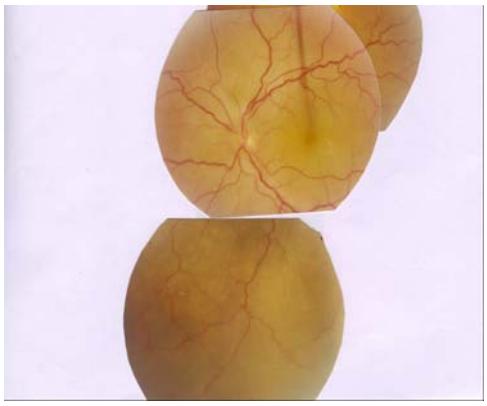

Fig 1b: Fundus montage picture of the left eye showing clear media, normal disc and inferior exudative retinal detachment.

Fundus fluorescein angiography showed multiple hyperfluorescent spots in the early-dye transit phase with pooling of the dye corresponding to exudative retinal detachment in the late phase (Figure 2a & 2b).

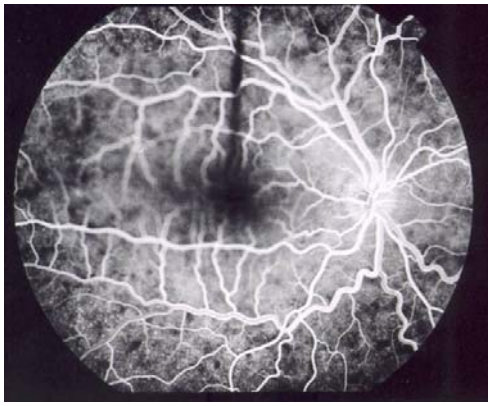

Fig 2a: Fundus fluorescein angiography of the right eye showing point like hyperfluorescent lesions in the early-dye transit phase.

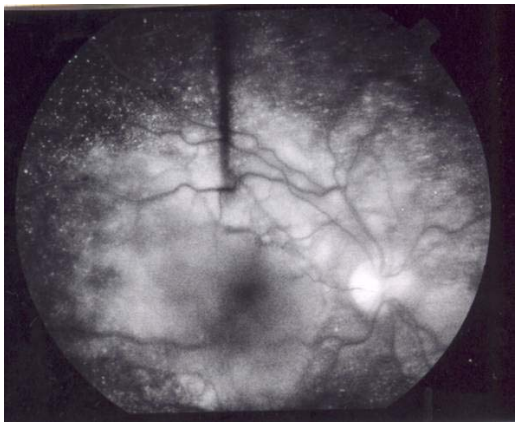

Fig 2b: Fundus fluorescein angiography of the right eye showing pooling of dye corresponding to exudative retinal detachment in the late phase.

Ultrasound A and B-scan showed increased choroidal thickness of 2.51mm in the right eye and 2.12 mm in the left eye. Systemic investigations including erythrocyte sedimentation rate, haemogram, blood counts, mantoux test, VDRL, TPHA, antineutrophilic antibodies, urine examination, fasting blood sugar and audiometry were unremarkable. The patient was diagnosed to be a case of "probable" VKH disease. Intravenous methyl prednisolone was administered for three consecutive days in a daily dosage of one gram. The patient was started on oral prednisolone (1.5mg/ kg body weight) which was slowly tapered according to the ocular response. At 4 weeks follow-up examination, the patient had unaided visual acuity of 6/9 for distance and N6 for near with +2.00 Diopter spherical in both the eyes. There was complete resolution of macular edema, exudative retinal detachment and retinochoroidal thickness. However the pupils were still 6 mm and nonreacting to light. The pupils showed tonic response to near stimulus and also constricted to a single drop of 0.16% pilocarpine. The patient had exacerbation of symptoms 3 months after the first episode when the oral prednisolone had been tapered to 40 mg once a day. The distance visual acuity was 6/12 in right eye and 6/9 in left eye and the near vision was N6 with + 2.00 dioptre spherical. The pupils were dilated and tonic reaction to near persisted. Slit-lamp biomicroscopy showed anterior chamber cellular reaction of + 2 and vitreous reaction of +1. Fundoscopy revealed macular edema in the right eye and whitish well defined placoid lesion with overlying edema at the level of retinal pigment epithelium and pigmentary changes in macula of the left eye. The steroids were increased to 100 mg once a day along with topical betamethasone. The reactivation of uveitis resolved in 2 weeks and the patient recovered vision. The steroids were gradually tapered over a period of 9 months. At 28 months follow-up, though the accommodation paresis had recovered, the pupils were 5mm dilated with absent light reaction and tonic pupillary reaction to near stimulus.

Discussion

Panuveitis in VKH disease is rarely associated with pupillary abnormalities. Review of literature using computerized 'Medline' search revealed three previous reports of the same (Brouzas D et al 1997, Kim JS et al 2001, Levy NS et al 1970). In all these

cases, panuveitis was associated with other signs of meningeal irritation and tonic pupils. In the present case however, the inflammation was limited to the eye only and no signs of meningeal irritation or other systemic involvement were seen. Thus this case falls in the category of "probable" VKH disease (Reed RW et al 2001).

Tonic pupil can be found in post ganglionic parasympathetic damage in cases of ciliary ganglionitis, autonomic and peripheral neuropathies or chronic diffuse ocular inflammations of the eye involving short posterior ciliary nerves. The recovery of accommodative paresis and constriction to 0.16 % pilocarpine is suggestive of partial recovery of post-synaptic fibres and denervation hypersensitivity of receptors. The ciliary ganglion lies in close proximity to meninges around the optic nerve and lateral rectus muscle. The diffuse inflammation in the former can involve the ganglion. The point of interest is that in our case, the optic disc did not show edema on clinical examination or fundus fluorescein angiography. However, the patient was not subjected to visual evoked potential. Diffuse inflammation has been documented even in quiescent cases of chronic VKH which could have lead to involvement of intraocular course of the short posterior ciliary nerves (Lubin JR et al 1982).

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