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CASE REPORT

Vogt-Koyanagi-Harada disease and bilateral simultaneous Adie's pupil

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Vogt-Koyanagi-Harada (VKH) syndrome is an uncommon autoimmune inflammatory malady commonly accompanied by neurological, auditory, and integumentary signs. We report a case of a teenage girl who complained of persistently decreased vision for the last 15 days. There was no systemic association. After 2 weeks, the patient developed tonic pupils. The pupil showed a tonic response to a 0.125% pilocarpine eye drop. Upon examinations, the patient was diagnosed with Vogt-Koyanagi-Harada disease and bilateral Adie's pupil, which is a rare conditions associated with VKH disease. Early treatment is crucial for preserving vision.

Keywords: Vogt-Koyanagi-Harada diseases, Adie's pupil, Pilocarpine, Autoimmune, Vision

Introduction

Vogt-Koyanagi-Harada (VKH) is an autoimmune condition that causes inflammation in tissues containing melanocytes, leading to depigmentation and vision problems. Timely diagnosis and treatment are effective in managing the condition's symptoms. Findings show inflammation in both eyes, fluid buildup in the retina, skin depigmentation, hair loss, tinnitus, hearing loss, and signs of meningitis (1). Pupillary abnormalities with VKH are rare. They're commonly associated with symptoms of meningeal irritation (2). Initial ophthalmic findings may be edema of the optic disk, diffuse choroiditis, and an exudative pattern of retinal detachment. Delayed ophthalmic features usually present as depigmentation of the retina (sunset glow fundus) and, less frequently, subretinal neovascularization (3). Very recently, studies reported that a new subset of T cells, termed Th17, may initiate and maintain this disease pattern (4). VKH malady occurs in four separate phases. Meningeal irritation and encephalopathy symptoms are the most common in the prodromal phage. A reduction in visual acuity in both eyes characterizes an acute uveitis phage. Patients typically develop vitiligo and poliosis due to depigmentation of the skin presentation in the chronic phage. In some cases of

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ocular inflammation, it can become intermittent, known as chronic recurrent phage (5).

Damage to the postganglionic fibers resulting from parasympathetic denervation causes pupillary constriction, which causes Adie's tonic pupil. Clinical characteristics of the tonic pupil include poor reaction to light, dissociation between near and light pupillary reflexes, tonic miosis and dilatation, segmental sphincter paralysis, and cholinergic hypersensitivity (6). The following are some reasons for a tonic pupil: migraine, vasospasm, surgery, trauma, and, rarely, ocular malignancies in the orbit or eye. Connective tissue and autoimmune diseases have been reported to be associated with tonic pupils (7). When bilateral involvement is present, it implies either autonomic neuropathy or widespread peripheral nerve dysfunction, such as diabetes, Riley-Day syndrome, paraneoplastic syndrome, or amyloidosis (8). Here, we attempt to describe an unusual clinical malady known as Vogt-Koyanagi-Harada disease with bilateral simultaneous Adie's pupil.

Case report

A teenage (16-year-old) girl from Bangladesh visited the Bangladesh Eye Hospital and Institute, reporting painless vision loss and floaters in her eyes for the previous 15 days. The young girl did not report any previous trauma or surgery in her clinical history. The patient had myopia, but there were no significant ocular or systemic medical conditions in the history. The overall examination of the body systems was within normal limits, with specific attention to the central nervous system, auditory system, and skin. The aided visual acuity was 6/12p by Snellen chart; slit lamp biomicroscopic examination reveals a moderate number of aqueous cells, flares in the anterior chamber, and a moderate amount of vitreous cells. The intraocular pressure was normal on both sides, at 17 mm Hg by non-contact air-puff tonometer. The retina was evaluated meticulously by an indirect ophthalmoscope on dilated pupils. There was shallow retinal detachment and blurred disk margins in both eyes.

The B-scan revealed an elevated choroidal thickness. Optical coherence tomography (OCT) showed multiple septate-making compartments with the presence of sub-retinal fluid (**Figure 1a, b**). Systemic investigations reported normal studies, including a complete blood count including erythrocyte sedimentation rate, random blood sugar, Mantoux test, Treponema pallidum hemagglutination assay test, antinuclear antibody test test, serum glutamic pyruvic transaminase test, serum creatinine, human leukocyte antigen B-27, and chest X-ray. A preliminary diagnosis has been made for VKH disease.

Abbreviations: AC, Anterior chamber; ANA, Antinuclear antibody test; SGPT, Serum Glutamic Pyruvic Transaminase test; TPHA, Treponema pallidum hemagglutination assay; VKH, Vogt-Koyanagi-Harada.

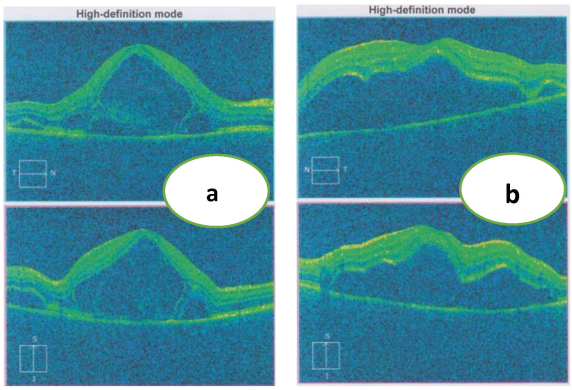

FIGURE 1 | OCT macula shows serous retinal detachment. **(a)** Right eye; **(b)** Left eye.

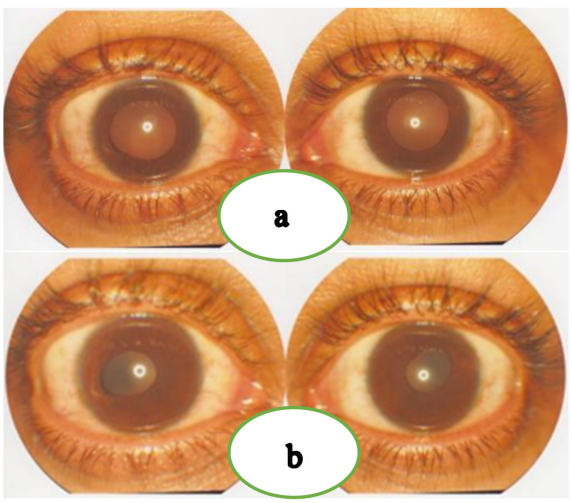

FIGURE 2 | (a) Pupil size before pilocarpine 0.125% drop instillation; **(b)** Pupil size 30 min after pilocarpine 0.125% drop instillation.

The patient received medication with both oral and topical steroids, which were gradually tapered based on the clinical response of the patient. The initial daily dose of oral prednisolone was 1 mg/kg body weight, gradually reduced to a maintenance dose of 15 mg over 3 months. After the first week of starting treatment, the patient's aided vision was 6/6 and there were no complaints of near vision. The anterior chamber (AC) is quiet, and the exudative retinal detachment (ERD) is resolving. At 2 weeks of follow-up, the patient complained of decreased near vision and a headache. Distant visual acuity 6/6 aided and near N6 with + 1.00 D addition. The AC is quiet, with a pupil 7 mm in size (**Figure 2a**) and a sluggish reaction in both eyes. The fundus examination revealed no ERD. The pupil showed a tonic response to near stimulus and continued to 0.125% pilocarpine eye drop (**Figure 2b**). Based on the overall clinical manifestations, the patient is confirmed to have bilateral Adie's pupils associated with VKH disease.

Discussion

Here is a report on a patient in Bangladesh who has VKH disease and bilateral Adie's pupil. In this case, there was only inflammation in the eye, with no signs of other systemic involvement or meningeal irritation. Our patient developed the acute uveitis stages. A possible diagnosis for VKH disease was considered using diagnostic criteria proposed by Read et al. (9).

When diagnosing VKH disease, the following criteria should be considered:

1 The inflammation should be present in both eyes.
2 There should be no prior history of surgery or trauma before the onset of uveitis.
3 There should be no clinical or laboratory evidence indicating the presence of any other ocular disease.
4 Skin changes or hair loss (alopecia) should be present.
5 Neurological symptoms should be present.
6 Auditory symptoms should also be present.

Complete VKH disease involves all six criteria, while incomplete VKH disease involves one to four criteria and neurological or auditory symptoms. Probable VKH disease only shows ocular involvement without any neurological or auditory symptoms (9–11).

The disease usually impacts people between 30 and 40, although it has been reported in children. It seems to impact women more often than men (3). VKH syndrome has been most frequently reported among people with more pigmentation, including Asians, Hispanics, and Native Americans (5). There is a rare association between tonic-pupil syndrome and VKH disease, and the reason for this relationship is still unclear. Kim and colleagues have suggested a potential explanation for the pupillary tone observed in certain patients (1, 12). They suggest that this change in tone may occur due to degeneration in the ciliary ganglion or damage to the short ciliary nerves, which can result from widespread inflammation within the eye. This damage may lead to abnormal re-innervation, which can perpetuate pupil abnormalities (6). No common mechanism may exist, and the occurrence may be a chance event. About 80% of Adie's tonic pupil is responsive to 0.125% pilocarpine (13). In this study, the patient developed tonic pupils and showed a response to 0.125% pilocarpine eye drop.

During the active phase, high-dose corticosteroids are administered intravenously for three consecutives or alternate days (13). This is followed by a moderate dose of oral steroids, which are gradually reduced. In our study, we advised only oral high dose corticosteroids initially, and gradually tapered depending on the treatment outcome. A long-term maintenance dose is helpful to reduce the recurrence of the disease. Some clinicians prefer to administer immunosuppressive agents as the first line of treatment (12, 14).

Complications like cataract, glaucoma, choroidal neovascularization, and optic atrophy may occur in up to 51% of patients with VKH disease. Early management and high clinical suspicion are crucial for maintaining good vision. Shorter duration oral corticosteroid is providing the greater risk of recurrence of VKH and poor visual outcome. It is better to tapper the dose of oral corticosteroid gradually and continued the maintaining dose at least 6 months for the management of VKH. The prognosis depends on the duration and frequency of ocular inflammation episodes (9, 10, 15, 16).

Conclusion

Although it is uncommon, bilateral simultaneous Adie's pupil could be associated with Vogt-Koyanagi-Harada disease. A thorough patient assessment and high suspicion index can help with diagnosis.

Author contributions

Conception and design of the study: MSA, SMUK, and RS. Acquisition of data: MSA, RS, and SR. Analysis and/or interpretation of data: MSA, RS, SR, and MZR. Drafting the manuscript: MSA and RS. Critical review/revision: SMUK and RPM.

Declaration of the consent of the patient

The authors have obtained appropriate patient consent forms for publishing clinical images and information in the journal.

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