Case Report

A rare case of complete Vogt-Koyanagi-Harada disease presenting to a tertiary care hospital in late stage: Clinical features, diagnosis, and management

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ABSTRACT

Vogt-Koyanagi-Harada disease (VKHD) is a rare T-cell-mediated multisystemic autoimmune disorder affecting organs with high melanocytic concentrations such as uvea, skin, ear, and meninges. VKHD is difficult to diagnose because its clinical presentation is variable and multisystemic which often leads to late diagnosis and treatment allowing the appearance and progression of the disease sequelae. Here, we report the case of a young adult female who was previously undiagnosed and inadequately treated at multiple centers presenting to our hospital in the late stage of VKHD with panuveitis, retinal detachment, hearing loss, alopecia, and vitiligo, which was classified as complete VKH disease and successfully treated in our hospital.

Key words: Panuveitis, Rare autoimmune disease, Retinal detachment, Vogt-Koyanagi-Harada disease

Vogt-Koyanagi-Harada disease (VKHD) is a rare multisystemic granulomatous autoimmune disease affecting organs with high melanocyte concentrations such as the eye, central nervous system (CNS), inner ear, and skin [1]. VKHD has a predilection for dark-complexioned persons and more prevalent in certain ethnicities such as Hispanics, people from the Middle East, and Asian Indians but not the blacks of sub-Saharan African descent [2]. It is more common in adults and women are more affected than men [3]. In India, a few cases have been reported with prevalence in uveitic cohorts being 1.4–3.5% in the South Indian population [4]. Pathogenesis of VKHD involves T-cell-mediated autoimmune disorder targeting melanocytic self-antigens. It has a genetic predisposition associated with HLA-DRB1*0405 [5].

CASE REPORT

A 35-year-old female presented to our hospital with chief complaints of pain, redness, and progressive diminution of vision in both eyes for 2 years. It was associated with headache, vertigo, and hearing loss. She also complained of a gradual loss of hair, white patches over the scalp, and forehead with whitening of eyebrows and eyelashes over a period of 3 years for which she had taken homeopathic medications. She had a history of multiple episodes of pain, redness, and diminution of vision in both eyes for 1 year for which she was seen and treated at many periphery hospitals in line of conjunctivitis with topical antibiotic drops but the detailed evaluation was not done anywhere. Later on, she developed further loss of vision for which she came to our hospital.

On general examination, the patient was conscious, cooperative, and well oriented. Vitals were stable. Pallor was present. There was the presence of multiple vitiligo patches over the scalp and forehead (Fig. 1a). On ocular examination, there was the presence of hypopigmented patches over the eyebrows and poliosis (Fig. 1b). Slit-lamp examination showed circumcorneal congestion, mutton-fat keratic precipitates over corneal endothelium, Grade 1+ cells, and minimal flare in the anterior chamber, poorly reacting pupils, segmental posterior synechiae, iris atrophic patches, and immature cataract in both eyes. Visual acuity was CF 1 m in both eyes. The intraocular pressure (IOP) was normal in both eyes.

Posterior segment examination revealed mild vitritis and retinal detachment involving the macula. Fundus photograph of both eyes showed similar findings as above (Fig. 2a). Optical coherence tomography (OCT) showed separation of the neuroretinal layer from outer hyper-reflective layer, presence of pockets of subretinal fluid separated by fibrous septa, subretinal deposits of fibrin, and internal limiting membrane irregularities in both eyes (Fig. 2b). Dermatological examination revealed alopecia and vitiligo. ENT examination showed sensorineural

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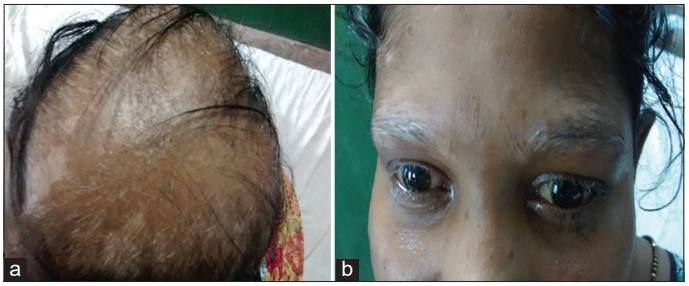

Figure 1: (a) Multiple vitiligo patches and alopecia on the scalp of the patient; (b) whitening of eyebrows and eyelashes

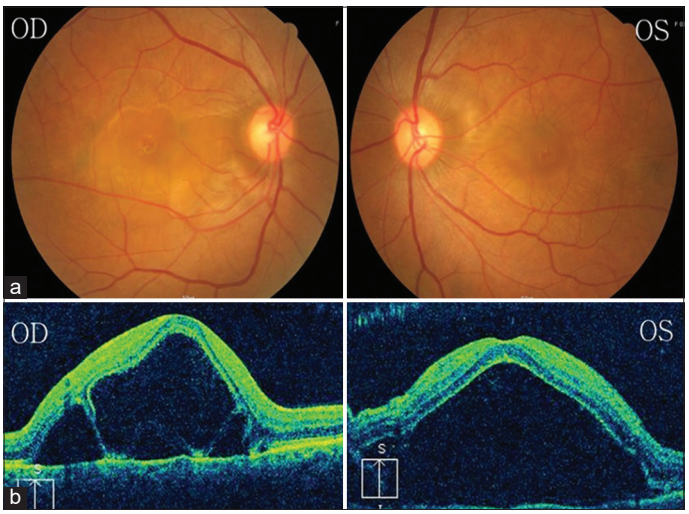

Figure 2: Fundus photo (a) and optical coherence tomography image (b) showing retinal detachment with subretinal fluid in both eyes

hearing loss of moderate degree (53 db) in both ears. CNS examination was normal. The rest of the systemic examinations were within normal limits.

Serological tests showed mild anemia (HB%: 8 g%) and elevated liver enzymes. Cerebrospinal fluid (CSF) study showed lymphocytic pleocytosis (lymphocytes: 85%). Chest X-ray, magnetic resonance imaging scan brain, and orbit were normal. Based on these findings, a diagnosis of complete VKH disease was done according to the revised diagnostic criteria for VKHD [6-8].

The patient was started on IV methylprednisolone (500 mg) for 3 days followed by oral prednisolone (1 mg/kg body weight) which was subsequently tapered at weekly intervals. She was also prescribed instillation of topical 1% prednisolone drop and 1% atropine 3 times/day.

Following treatment, pain and redness resolved over a week and there was a decrease of subretinal fluid over a period of 2 weeks. Follow-up at the end of 2 months of corticosteroid therapy showed complete resolution of subretinal fluid and complete reattachment of the retina on fundus photograph (Fig. 3a) and OCT (Fig. 3b) in both the eyes. However, there was a progression of cataract in both the eyes and worsening of alopecia. Best-corrected visual acuity at the end of 2 months was 20/200 in both eyes. The patient is now maintained on low-dose topical steroids. The patient has been educated about the nature of the disease and its complications and advised for timely follow-ups.

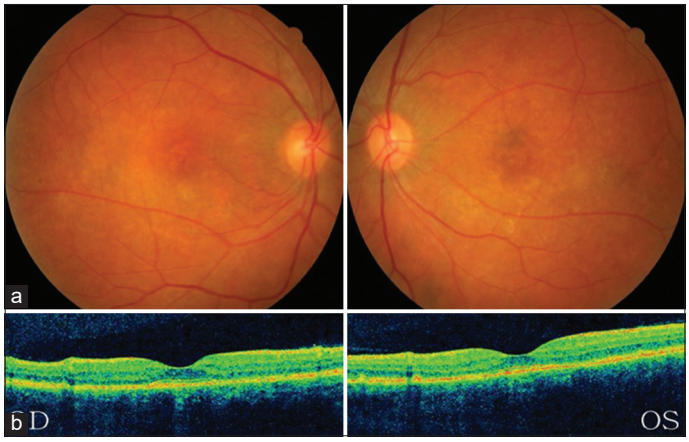

Figure 3: Fundus photo (a) and optical coherence tomography image (b) showing resolution of subretinal fluid and reattachment of retina post-treatment in both eyes

DISCUSSION

Vogt-Koyanagi-Harada syndrome is an uncommon autoimmune disorder having multisystemic involvement. The clinical course of VKHD includes four stages: (a) Prodromal stage characterized by non-specific symptoms such as malaise, fever, headache, and neck stiffness. Neurological involvement such as cranial nerve palsies, hemiparesis, transverse myelitis, optic neuritis, and hearing loss can also occur. CSF shows lymphocytic pleocytosis [9]. (b) Acute uveitic stage: It follows the prodromal phase and lasts for several weeks. Bilateral posterior uveitis is the most common finding; however, unilateral cases have also been reported. Fundus findings include multiple serious retinal detachments, thickening of the posterior choroid, and optic disc edema [10]. (c) Convalescent stage shows progressive depigmentation of the skin leading to vitiligo [9] and uvea leading to "Sunset glow" appearance of the fundus. Perilimbal vitiligo is the earliest depigmentation to occur, often within 1 month of onset of disease [11]. Alopecia and poliosis can also occur. (d) Chronic recurrent stage: Recurrent anterior granulomatous uveitis and ocular complications such as glaucoma, cataract, and choroidal neovascular membrane [9].

At present, VKHD is classified as per "The Revised diagnostic criteria" (2001) by the International Committee on Nomenclature as follows: [6-8] (a) Complete VKH disease – in which ocular, integumentary, and neurologic/auditory involvement are present. (b) Incomplete VKH disease – ocular with either neurological/ auditory involvement is present. (c) Probable VKH disease is uveitis consistent with VKH without any extraocular manifestations.

Although multiple cases of VKHD have been reported, yet the prevalence of this disease is low in our region. Due to dermatological manifestations of the disease such as vitiligo and alopecia which are very bothersome to the patients, these patients first seek treatment for such condition. The patient, in this case, was treated with multiple homeopathic and Ayurvedic medications for alopecia and vitiligo for 3 years and was not referred to seek any medical advice. This was the reason for the late diagnosis of the entity. Further, the beneficial and harmful effects

of such medications in the disease progression are not known. Multifaceted presentation of the disease, lack of awareness, and improper diagnosis, all these commonly lead to late presentation of this disease to the ophthalmological department. This case also was treated at multiple peripheral centers in line of conjunctivitis but posterior segment evaluation and systemic evaluation were not done anywhere which is another reason for the late diagnosis of this case.

The goal of treatment in VKHD is to suppress active inflammation, prevent disease relapse, and avoid complications. Treatment mainly includes administration of systemic immunosuppressive supplemented by local corticosteroids [12]. Early administration of oral prednisone at a dose of 1–2 mg/kg/day followed by slow tapering to avoid recurrences is the generally accepted regimen, while pulse intravenous corticosteroid therapy of 1 g/day of methylprednisolone for 3–5 days followed by oral prednisolone is usually reserved for cases with severe inflammation. The cases not responding to steroids, other immunosuppressive such as cyclosporine, azathioprine, and methotrexate, can also be added. Our patient showed a good response to steroids alone. Slow tapering of the corticosteroid dose, along with frequent follow-up examinations, is necessary to avoid the recurrence of inflammation [12,13]. IOP should be monitored at each follow-up visit, as glaucoma is a known secondary complication of this disease [9]. Recurrence of the disease is also very common so all the patients of VKHD should be counseled for regular follow-up to prevent further complications.

CONCLUSION

As VKHD is a progressive, rare, and multisystemic disease with variable outcomes, an early diagnosis and treatment play a significant role in deciding the fate of the patient. Primary health workers, being the ones to have the first contact with such patients, should be more aware of such syndromic multisystemic diseases and diligent while dealing with such patients. Early referral to tertiary centers should be the golden rule, as delay in initiation of treatment can lead to secondary complications such as serous retinal detachment, cataract, glaucoma, choroidal neovascularization, and sensorineural hearing loss, some of which may be irreversible. Moreover, in all the cases presenting with pain and redness to the eye department, fundoscopy should be done invariably, to rule out posterior segment involvement.

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