Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Internal Medicine

Vogt-Koyanagi-Harada Disease Diagnosed in a Black-Skinned Woman in Internal Medicine

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DOI: 10.36347/sjmcr.2023.v11i06.030 | **Received:** 01.05.2023 | **Accepted:** 07.06.2023 | **Published:** 12.06.2023

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Abstract **Case Report**

***Introduction*:** Vogt-Koyanagi-Harada disease, initially described as an uveomeningoencephalitic syndrome, is a systemic granulomatous autoimmune disease that targets melanocyte-rich tissues, such as the eye, inner ear, meninges, skin and hair. It is one of the most prevalent causes of noninfectious uveitis affecting, more frequently, individuals of pigmented skin, such as Asians, Middle Easterners, Hispanics and Native Americans. However, it is very infrequent among persons of African descent. Here, we report on a case in a black-skinned woman in internal medicine. ***Clinical Observation*:** A 45-year-old Malian black-skinned woman with no history of ocular trauma or intraocular surgery presented at internal medicine outpatient clinic with 3-months history of achromic skin patches over the face and trunk, bilateral visual disturbance, bilateral hearing loss with tinnitus and headache. Dermatological examination revealed bilateral, symmetrical achromic lesions over the back, buttocks and eyelids, with scalp poliosis; and perioral pityriasis versicolor lesions. She had no alopecia or other dermatological lesions. Ophthalmological examination revealed a profound drop in visual acuity. Oto-rhino-laringologic examination revealed a discreet hearing loss. Ophthalmological investigations revealed bilateral posterior panuveitis on funduscopic examination, bilateral chronic iridocyclitis on slit- lamp examination, and bilateral choroidal thickening on ocular ultrasound. The diagnosis of Vogt-Koyanagi-Harada was retained, based on the diagnostic criteria established by the American Society for the Study of Uveitis (UAS) in 1978. Prednisone at a dose of 1.5 mg per kilogram tapering courses with adjuvant treatment was initiated. The ophthalmologist's prescription consisted of 1% atropine eye drops, 0.25% timoptol eye drops and dexamethasone eye drops which had been discontinued because of systemic cortitherapy; and that of the dermatologist, Hydrocortisone butyrate 0.1% cream, ciclopirox olamine 1% cream. On seven months follow-up visit, systemic signs and hearing disorders disappeared, vitiligo lesions had slightly regressed and visual acuity had improved. ***Conclusion*:** It appears from our observation that the diagnosis and management of this granulomatous and systemic autoimmune disease must be based on a multidisciplinary approach, including the ophthalmologist, dermatologist, neurologist, otolaryngologist, and internist.

Keywords: Vogt-Koyanagi-Harada disease, vitiligo, panuveitis, internal medicine, Mali.

INTRODUCTION

Vogt-Koyanagi-Harada disease, initially described as an uveomeningoencephalitic syndrome, is a systemic granulomatous autoimmune disease that targets melanocyte-rich tissues, such as the eye, inner ear, meninges, skin and hair. This disease is mainly a

Th1 lymphocyte mediated aggression to melanocytes after a viral trigger in the presence of HLA-DRB1*0405 allele [1, 2].

Vogt-Koyanagi-Harada disease is an important cause of noninfectious uveitis affecting, more frequently, individuals of pigmented skin, such as Asians, Middle Easterners, Hispanics and Native Americans. However, it is very infrequent among persons of African descent [3]. Here, we report on a case in a black-skinned woman in internal medicine.

CLINICAL OBSERVATION

A 45-year-old Malian black-skinned woman with no history of ocular trauma or intraocular surgery presented at internal medicine outpatient clinic with 3-months history of achromic skin patches over the face and trunk, bilateral visual disturbance, bilateral hearing loss with tinnitus and headache. On physical examination, the blood pressure was 130/80 mmHg, the heart rate was 100 beats per minute, the respiratory rate 28 cycles per minute, the temperature was 37.1°C and the Body Mass Index (BMI) was 18.58 kilogram per square meter. Dermatological examination revealed bilateral, symmetrical achromic lesions over the back, buttocks and eyelids, with scalp poliosis (figs. 1 and 2); and perioral pityriasis versicolor lesions. She had no alopecia or other dermatological lesions. Ophthalmological examination revealed a profound drop in visual acuity. Oto-rhino-laringologic examination revealed a discreet hearing loss. Neurological examination revealed no meningoencephalitis syndrome.

The complete blood count showed a non-regenerative microcytic hypochromic anemia (the hemoglobin level was 8.0 g per deciliter, the mean corpuscular volume was 59.1 femtoliters, the mean corpuscular hemoglobin concentration was 17.9 picograms and the reticulocyte rate was 98 170 cells per cubic millimeter . The hemolysis and martial workup was normal. The inflammatory markers was elevated, the erythrocyte sedimentation rate at 99 millimeter at the first hour (normal range, 0 to 29 millimeter), and the blood C-reactive protein level at 56,5 mg per liter (normal value, < to 6 mg per liter). The antinuclear antibody test was negative, and no other autoantibodies testing were performed. Ophthalmological investigations revealed bilateral posterior panuveitis on funduscopic examination, bilateral chronic iridocyclitis on slit-lamp examination, and bilateral choroidal thickening on ocular ultrasound. No fluorescein or indocyanine green angiography or optical coherence tomography were performed. Skin biopsy was also not performed. The infectious assessment was unremarkable.

The diagnosis of Vogt-Koyanagi-Harada was retained, based on the diagnostic criteria established by the American Society for the Study of Uveitis (UAS) in 1978 [4] (table 1).

Prednisone at a dose of 1.5 mg per kilogram tapering courses adjuvant treatment was initiated. The ophthalmologist's prescription consisted of 1% atropine eye drops, 0.25% timoptol eye drops and dexamethasone eye drops which had been discontinued because of systemic cortitherapy; and that of the dermatologist, Hydrocortisone butyrate 0.1% cream, ciclopirox olamine 1% cream.

On seven months follow-up visit, systemic signs and hearing disorders disappeared, vitiligo lesions had slightly regressed and visual acuity had improved.

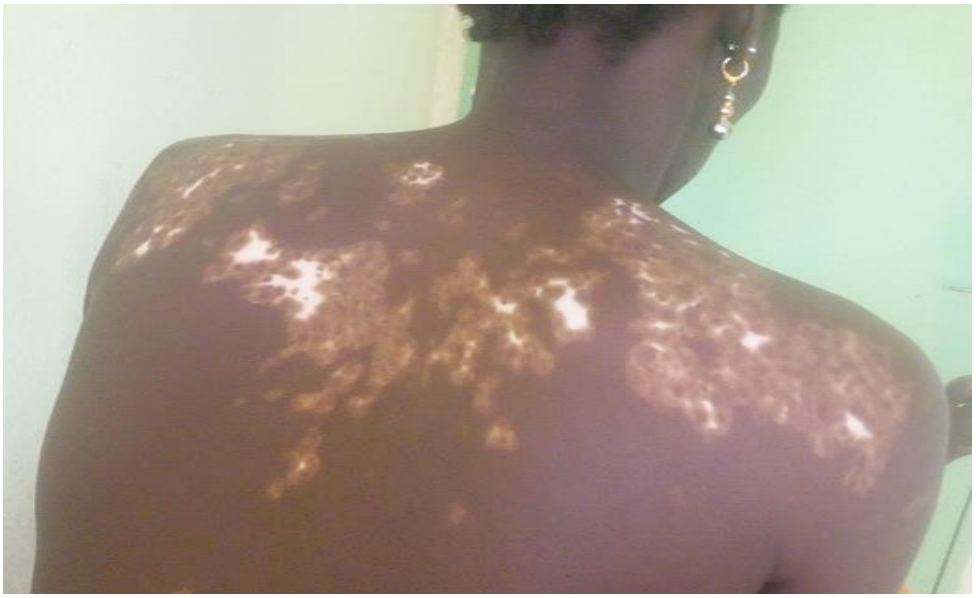

Figure 1: Symmetrical, bilateral vitiligo lesions over the upper back

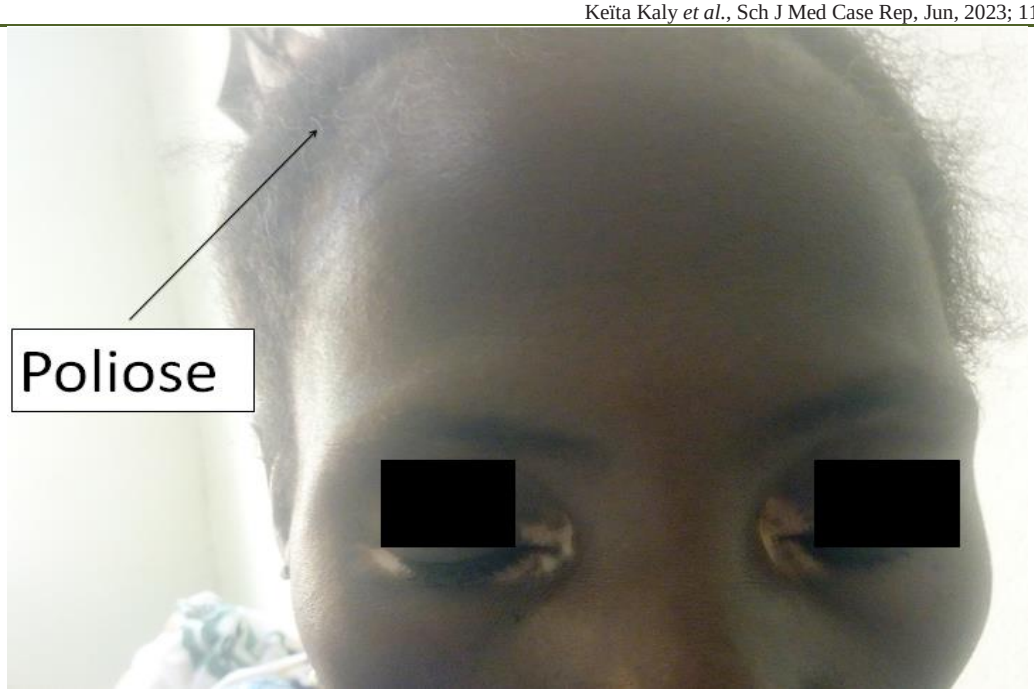

Figure 2: Symmetrical, bilateral vitiligo lesions over the eyelids with scalp poliosis

Table 1: Diagnostic criteria of American Uveitis Society (AUS) in 1978 for the diagnosis of Vogt Koyanagi Harada Disease in our patient

Diagnostic criteria of American Uveitis Society (AUS) in 1978 [4]	**In our patient**
No history of ocular trauma and/or surgery	She had no history of ocular trauma and/or surgery
At least three of the following four signs	
Bilateral chronic iridocyclitis	Her slit-lamp examination concluded a bilateral chronic iridocyclitis
Posterior uveitis	Her funduscopic examination revealed bilateral posterior panuveitis
Neurologic signs	She presented also with 3-months history of tinnitus and headache.
Cutaneous findings (alopecia, poliosis or vitiligo).	bilateral, symmetrical achromic lesions on the back, buttocks and eyelids, with scalp poliosis were noted on dermatological examination

DISCUSSION

Vogt-Koyanagi-Harada disease is a rare granulomatous inflammatory disease that affects pigmented structures, such as eye, inner ear, meninges, skin and hair [2]. It is one of the most prevalent causes of noninfectious uveitis affecting, more frequently, individuals of pigmented skin, such as Asians, Middle Easterners, Hispanics and Native Americans. However, it is very infrequent among persons of African descent [3]. Its incidence among all cases of uveitis varied 1% – 7% [5-8]. In the United States, this incidence is approximately 1.5 to 6 per 1 million patients, while in Japan approximately 800 new patients each year [1, 9]. In Africa, few cases are reported [10, 11]. Most studies have found that women were affected more frequently than men and that most patients were in the second to fifth decades of life at the onset of the disease [2]. This case report describes the onset of Vogt-Koyanagi-Harada disease in Malian young adult black-skinned woman revealed by achromic skin patches on the face and trunk, bilateral visual disturbance, bilateral hearing loss with tinnitus and headache and retained according to the diagnostic criteria of American Uveitis Society (AUS) in 1978 [4] well-fulfilled.

Vogt-Koyanagi-Harada disease is classically divided into four stages: prodromic, acute uveitic, convalescent and chronic/recurrent [1]. The patient had presented the prodromal, acute uveitic and convalescent stage signs but not chronic/recurrent stage (table 2).

Furthermore, the extraocular manifestations involved mainly integument and the central nervous system at various stages of the disease. Except the mingoencephalitic syndrome, our patient extraocular manifestations (table 3).

The main differential diagnosis of Vogt-Koyanagi-Harada disease is sympathetic ophthalmia secondary to the trauma and/or previous intraocular surgery. Anamnesis revealed no history of trauma and/or previous intraocular surgery in our patient.

Table 2: Signs of different stages of disease found in our patient

Stages of disease [2]	Our patient
Prodromal stage (fever, headache, nausea, vertigo, orbital pain, photophobia, tearing, tinnitus, vertigo and neurologic symptoms and cerebrospinal fluid examination findings)	Headache, tinnitus, cerebrospinal fluid examination findings are not performed
Acute uveitic stage (acute blurring of vision in both eyes, a diffuse choroiditis with exudative detachment of the neurosensory retina, Hyperemia and edema of the optic disk, bilateral granulomatous iridocyclitis with mutton fat keratic precipitates, iris nodules and shallow anterior chamber)	Bilateral visual disturbance, bilateral posterior panuveitis on funduscopic examination, bilateral chronic iridocyclitis on slit-lamp examination, and bilateral choroidal thickening on ocular ultrasound.
Convalescent stage (depigmentation of the integument and choroid: vitiligo, alopecia and poliosis)	Bilateral, symmetrical achromic lesions on the back, buttocks and eyelids, with scalp poliosis
Recurrent or chronic stage (Ocular complications observed in convalescent and chronic stages: cataract, glaucoma, choroidal neovascularization and retinal/choroidal fibrosis)	Not yet

Table 3: Extraocular manifestations seen in our patient

Extraocular manifestations [2]	Our patient
Central nervous system involment (neck stiffness, confusion and headache, cranial neuropathies, hemiparesis, aphasia, acute transverse myelitis and ciliary ganglionitis, pleocytosis on cerebral spinal fluid)	Only headache
Inner ear involvement (dysacusis, hearing loss and vertigo, tinnitus)	Hearing loss, tinnitus
Skin and appendages involvement (vitiligo, alopecia and poliosis of the lashes, eyebrows and scalp hair)	Bilateral, symmetrical achromic lesions on the back, buttocks and eyelids, with scalp poliosis

CONCLUSION

It appears from our observation that the diagnosis and management of this granulomatous and systemic autoimmune disease must be based on a multidisciplinary approach, including the ophthalmologist, dermatologist, neurologist, otolaryngologist, and internist.

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