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Clinical Case

A Case of Rare Autoimmune Disease: Vogt-Koyanagi-Harada Disease

La Maladie de Vogt-Koyanagi-Harada, une Maladie Auto-Immune Rare : À Propos d'un Cas

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ABSTRACT

Vogt-Koyanagi-Harada (VKH) disease is a rare autoimmune disorder. It is exceptional in black sub-Saharan Africans. A 27-year-old dermatology patient with progressive segmental vitiligo associated with bilateral visual acuity loss, headache and hearing loss. Hypocromic macules were noted extending to the inner surface of the upper lip and to the right jugal region. Examination of the skin revealed poliosis of the moustache and scalp. Slit-lamp inspection revealed numerous corneal keratic precipitates. Optical coherence tomography (OCT) revealed chorioretinitis scarring in the right eye and macular and papillary atrophy in the left. Tone luminance audiometry revealed a 1st degree major hearing loss of the bilateral mixed type. We made the diagnosis of Vogt-Koyanagi-Harada syndrome. The patient received a bolus of methylprednisolone followed by prednisone.. Vogt-Koyanagi-Harada disease is not common in sub-Saharan Africa. It is essential to consider this disease in all cases of segmental vitiligo.

RÉSUMÉ

La maladie de Vogt-Koyanagi-Harada (VKH) est une maladie auto-immune rare. Elle est exceptionnelle chez les noirs d'Afrique subsaharienne. Nous rapportons un cas de VKH chez un sujet de phénotype noir. Il s'agit d'un patient de 27 ans reçu en consultation de dermatologie pour un vitiligo segmentaire survenue progressive associée à une baisse de l'acuité visuelle bilatérale, des céphalées et une hypoacousie. On notait des macules hypocromiques s'étendant au niveau de la face interne de la lèvre supérieure et à la région jugale droite. Les muqueuses étaient d'aspect sain et l'examen des phanères retrouvait une poliose au niveau de la moustache et du cuir chevelu. L'inspection à l'aide d'une lampe à fente a révélé la présence de nombreux précipités kératiques cornéen. La tomographie en cohérence optique a retrouvé à l'œil droit une cicatrice de chorioretinite et à l'œil gauche une atrophie maculaire et papillaire. L'audiométrie tonale luminaire a retrouvé un déficit auditif majeur de 1er degré de type mixte bilatéral. Devant ce tableau clinique nous avons retenu le diagnostic du syndrome de Vogt-Koyanagi-Harada devant le vitiligo, la surdité et la panuveite bilatérale. Le patient a bénéficié d'un bolus de methylprednisolone 240 mg pendant 3 jours puis relais avec de la prednisone 1 mg/kg par voie orale pendant 6 mois. La maladie de Vogt-Koyanagi-Harada n'est pas fréquente en Afrique subsaharienne. Il est essentiel d'envisager cette maladie devant tout vitiligo segmentaire uvéite bilatérale, qu'elle soit ou non accompagnée de signes neuroméningés ou de l'oreille interne.

INTRODUCTION

Vogt-Koyanagi-Harada (VKH) disease is a rare systemic autoimmune disease with manifestations mainly affecting the eyes, ears, skin and nervous system. [1]. The disease is mediated by Th1 lymphocytes targeting melanocytes, so all body tissues made up of melanocytes can be affected. [2]. The origin of this condition remains unknown, although numerous infectious triggers have been suggested. VKH disease is uncommon and mainly affects dark-skinned Asian, Middle Eastern, Hispanic and Native American populations. [1]. However, it is rarely described in black sub-Saharan Africans [3]. In this case report, we describe VKH disease in a black sub-Saharan African subject of Burkina Faso origin, discovered during follow-up of vitiligo.

CASE PRESENTATION

Mr DD was a 27-year-old patient who had been regularly treated in dermatology for 5 years for vitiligo of undetermined aetiology and who had no other specific pathological history. He was seen in a dermatology consultation for a progressive extension of the vitiligo lesions associated with a bilateral decrease in visual acuity, headaches and hypoacusis. Examination of the skin and appendages revealed hypocromic macules extending to the inner surface of the upper lip and the right jugal region (Figure 1).

Figure 1: Hypocromic macules extending to the inner surface of the upper lip and the right jugal region

The same placard-like lesions were found on the forehead. The mucous membranes were healthy and examination of the skin revealed poliosis of the moustache and scalp, with no alopecia. Visual acuity was 1/10 in the right eye and 3/10 in the left. Bilateral conjunctival congestion was noted. Slit-lamp inspection revealed numerous mediumsized keratic precipitates on the corneal endothelium, with extensive cellular infiltration and marked dilatation of the anterior chamber. Both pupils showed synechiae. Intraocular pressure was 17 mmHg in both eyes. Otoscopy was normal on ENT examination. There was no involvement of other pairs of cranial nerves, and examination of the nervous system and other systems was unremarkable. Biochemical examination of the cerebrospinal fluid revealed pleocytosis, with lymphocytes accounting for 80%, and normal glycorrhachia and proteinorrhachia. The haemogram was unremarkable, as were the renal and hepatic functions. Papillary and macular OCT revealed a chorioretinitis scar with an interpapillomacular membrane in the right eye and macular and papillary atrophy in the left (Figure 2).

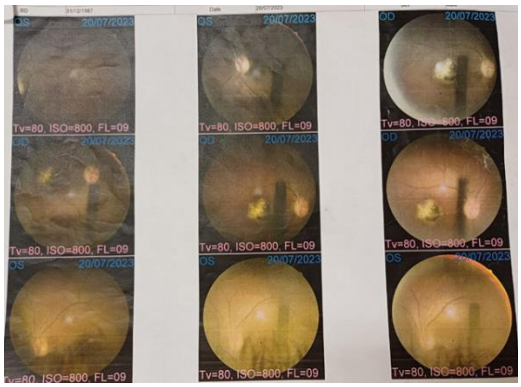

Figure 2: Optical coherence tomography revealed chorioretinitis scarring in the right eye and macular and papillary atrophy

Luminal tone audiometry revealed a major hearing loss of 1er degrees of mixed type on both sides (Figure 3).

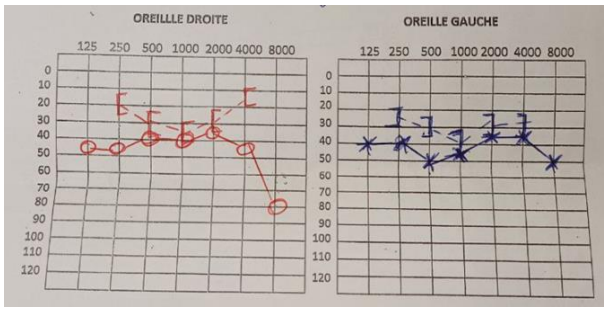

Figure 3: Luminal tone audiometry revealed a major hearing loss of 1er degrees of mixed type on both sides

Cerebral CT was unremarkable. Paraclinical examinations for other autoimmune diseases were inconclusive. Given this clinical picture, we made the diagnosis of an acute attack of VKH disease in view of the vitiligo, deafness and bilateral panuveitis. The patient received a 240 mg bolus of methylprednisolone for 3 days, followed by oral prednisone at a dose of 1 mg/kg/day, with a 10% regression every 10 days for 6 months.

DISCUSSION

As far as we know, this is the first case of VKH disease to be reported in Burkina Faso and one of the rare cases described in sub-Saharan Africa because this disease is rare in black African patients. VKH disease, also known as Vogt-Koyanagi-Harada syndrome, was described by Vogt in 1906, Harada in 1926 and Koyanagi in 1929. It is a bilateral, chronic and diffuse granulomatous panuveitis, presenting with serous retinal detachment and often associated with neurological, auditory and dermatological involvement [4]. The incidence of the disease is estimated at 1:40,000, with a variable geographical distribution. [5]. VKH disease accounts for 4-11% of endogenous uveitis and predominates in young adults and women [6]. It generally manifests itself against a particular genetic background, as shown by the family cases documented in the literature. The socio-demographic data from the series by Alaoui et al, which reports an average age of 36 years, correspond to that of our patient. [6]. However, the male gender of our patient differs from the cases in this series, which was predominantly female, as was the series of Moroccan patients [6] [7].

The pathogenesis of this condition is correlated with an immune dysfunction targeting melanocytes, causing cytotoxicity and apoptosis mediated by T [6]. As melanocytes are neural crest cells and contribute to the formation of tissues such as the skin, meninges, retina, uvea, cochlea and labyrinth, the disease can affect these various organs. [5]. It is also potentially associated with the detection of a melanotropic virus, Epstein Barr virus [6]. Several authors have highlighted the presence of anti-retinal antibodies, in particular anti-S-arrestin, which could be one of the preferred immune targets of this syndrome [6]. However, VKH disease is thought to occur in a genetic background linked to the association with HLA DR4/HLA DRB1-04*05, which has been reported in the Japanese population. [2]. The presence of this allele is associated with a higher risk of developing this disease. The diagnosis of VKH disease was based on the diagnostic criteria of The International Committee on

Nomenclature Classification and the American Uveitis Association of VKH syndrome of 2001 with a complete VKH syndrome found in our patient. [8]. This syndrome is said to be complete when there is no history of trauma or ocular surgery and there are no clinical or biological abnormalities suggestive of other ocular pathologies. In addition, there is bilateral ocular involvement such as panuveitis, neurological involvement such as hypoacusis and skin involvement such as poliosis and vitiligo. In the literature, hearing loss is asymptomatic and found in 75% of cases with an average loss of 30 dB. However, it was symptomatic in our patient in whom hypoacusis was one of the reasons for consultation. [2]. In fact, dysacusis is generally found in the prodromal phase, as well as in other neurological disorders of VKH disease, preceding the acute uveitis phase. In addition, like our case, 63% of patients in the series by Lavezzo et al had lymphocytic meningitis, which is in line with the literature. [9]. Skin involvement is found in 10 to 63% of patients with pigmented skin and in the cohort of Diallo et al it represented 93% of patients. [10]. These skin lesions are generally of the poliosis, vitiligo or alopecia type, but they occur in the late stages known as convalescence phases and are rarely revealing. [10]. Ocular involvement is generally the first reason for consultation in VKH disease and is a key factor in the severity of the disease. They may be located in the anterior uvea, representing the Vogt-Koyanagi variety of the disease, or in the posterior uvea, representing the Harada variety of the disease. In our patient, the ocular and ocular damage progressively developed over 3 years during the follow-up of the vitiligo. Our patient had no cerebral involvement on cerebral CT as in most cases described in the literature. Although CT cannot be used to diagnose VKH disease, it can be used to rule out differential diagnoses of neurological involvement.

The treatment of VKH disease is not well codified due to its rarity. Its treatment is a challenge for the practitioner, especially in an African context where technical facilities are limited. However, all authors agree that high-dose corticosteroid therapy improves disease activity and improves visual and ocular prognosis. [10]. This prednisone- or methylprednisolone-based corticosteroid therapy may be administered for 6 to 12 months as a first-line treatment. [10]. However, in cases of corticoresistance or corticodependence, some authors recommend the use of immunosuppressants such as aziathropine, mycophenolate mofetil and tacrolimus in 2ème lines. [6]. In the event of failure, rituximab has shown results in some cases described in the literature [6]. In terms of the prognosis of VKH disease, certain poor prognostic factors have been identified, such as advanced age, a chronic inflammatory state with long-term corticosteroid therapy and the presence of subretinal neovessels. [7]. Ocular damage is the most serious and hearing damage generally regresses within 2 or 3 months, while dermatological damage remains permanent. [11].

CONCLUSION

VKH disease is not common in Burkina Faso or sub-Saharan Africa. It is essential to consider this disease in all cases of bilateral uveitis, whether or not accompanied by neuromeningeal or cutaneous signs. A high index of suspicion is recommended for early detection and prompt treatment to avoid irreversible visual loss.

CONFLICT OF INTEREST

We have no conflict of interest.

DATA AVAILABILITY

All data present in this study are available from the corresponding author upon a reasonable request.

INFORMED CONSENT

We have obtained the informed consent of the patient.
We have obtained consent to publish this article.

AUTHORS' CONTRIBUTIONS

Diabri Banyama Marie, Bayala Yannick Laurent Tchenadoyo, contributed to the study design, data collection, data analysis, and manuscript writing. Marcellin Bonkoungou contributed to manuscript writing. Zabsonre/Tiendrebeogo Joëlle Wenlassida Stéphanie, Ouedraogo Dieu-Donné were responsible for the design and supervision of the study.

ETHICAL CONSIDERATIONS

Our study is not unethical.
We have not funder.
We confirm that the current manuscript is not under consideration in other journals - as a whole or in part.

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