SAS Journal of Surgery
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

Vogt-Koyanagi-Harada

Vogt-Koyanagi-Harada Syndrome Diagnosed at the Sequellar Stage in an 11-Year-Old Child: A Case Report

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DOI: 10.36347/sasjs.2023.v09i02.010 | **Received:** 11.12.2022 | **Accepted:** 29.01.2023 | **Published:** 17.02.2023

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Abstract **Case Report**

Vogt-Koyanagi-Harada syndrome is a severe granulomatous panuveitis associated with neuromeningeal, dermatologic, and auditory extraocular manifestations. Uveitis in children accounts for only 5-10% of all uveitis. Vogt-Koyanagi-Harada syndrome is an even rarer cause. The exact etiology remains unknown, however, a genetic predisposition to an autoimmune process directed against melanocytes is incriminated. We report the case of an 11-year-old child with a history of recurrent red eyes with episodes of headache accompanied by dizziness who consulted us for a painful red eye with decreased visual acuity. In view of the clinical and paraclinical anamnestic elements and based on the diagnostic criteria of the Vogt-Koyanagi-Harada syndrome of the American Uveitis Society revised in 2001, the diagnosis of a VKH syndrome in its complete form was retained. Treatment with 3 boluses of methylprednisolone (1 g/1.73m2) over three days was initiated, followed by oral therapy combined with topical corticosteroids and cycloplegic therapy. The long-term evolution was marked by recurrences of the previous uveitis, and the patient was put on immunosuppressive treatment with mycophenolate mofetil (30 mg/kg per day in two doses). Although rare, VKH syndrome exists in the pediatric population. Our observation highlights the diagnostic difficulties associated with this condition. We insist on the usefulness of a thorough history in order to retrace the history of the disease and on the necessity of an early diagnosis and treatment to guarantee a better prognosis.

Keywords: VKH disease, Childhood, retrospective diagnostic.

INTRODUCTION

Vogt-Koyanagi-Harada syndrome is a severe granulomatous panuveitis associated with neuromeningeal, dermatologic, and auditory manifestations [1].

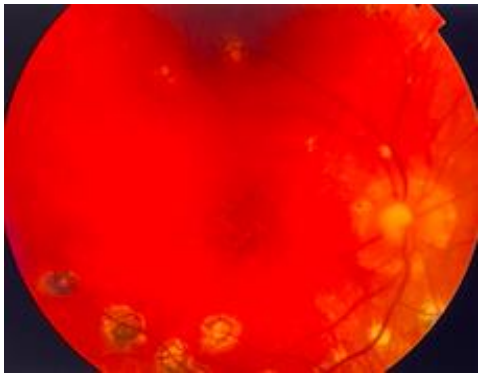
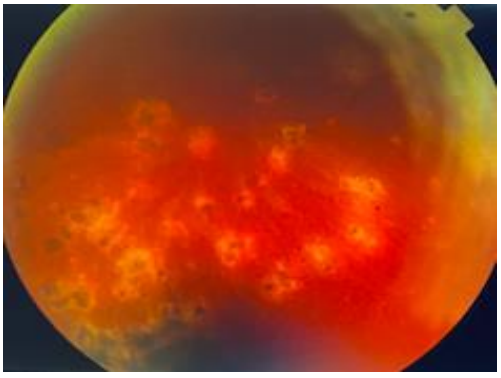
Uveitis in children accounts for 5-10% of all uveitis. Vogt-Koyanagi-Harada syndrome is an even rarer cause. The etiological factors remain unknown, but there is a genetic predisposition to an autoimmune process directed against melanocytes.

We report the case of an 11-year-old child with bilateral choroiditis in the context of Vogt-Koyanagi-Harada syndrome diagnosed in its chronic stage.

CLINICAL CASE

An 11-year-old child presented to the emergency room with a painful red eye and decreased visual acuity for 4 days. The interrogation reported repeated episodes of red eyes that resolved spontaneously, as well as episodes of headache accompanied by dizziness.

The ophthalmological examination revealed a visual acuity of 1/10 in the right eye and 8/10 in the left eye with conjunctival hyperemia only in the right eye. Examination of the anterior segment of the right eye revealed non- granulomatous retrodescemetic precipitates, banded keratopathy, 3-crossed cellular Tyndall, cyclitic membrane and iridocrystalline synechiae. There is no ocular hypertonia. The left eye has non-granulomatous retrodescemetic precipitates, a quiet anterior chamber and iridocrystalline synechiae. Fundus examination in both eyes showed yellowish-white deep choroidal nodules (Dalen Fuchs nodules) with diffuse retinal depigmentation giving a sunset appearance, the macula appeared reworked. There is no serous detachment (DSR) in both eyes (Figures 1 and 2).

Figures 1 and 2: Dalen Fuchs nodules with sunset appearance of the fundus in both eyes

The general examination revealed poliosis of the bregmatic region (Figure 3) and skin depigmentation of the back (Figure 4) and forefeet (Figure 5).

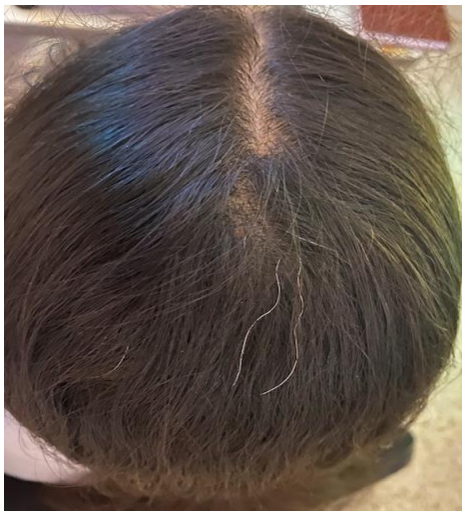


Figure 3: Poliosis of the bregmatic region

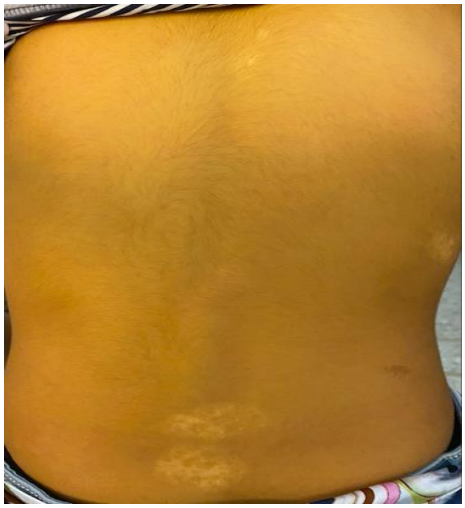


Figure 4: Depigmentation of the back

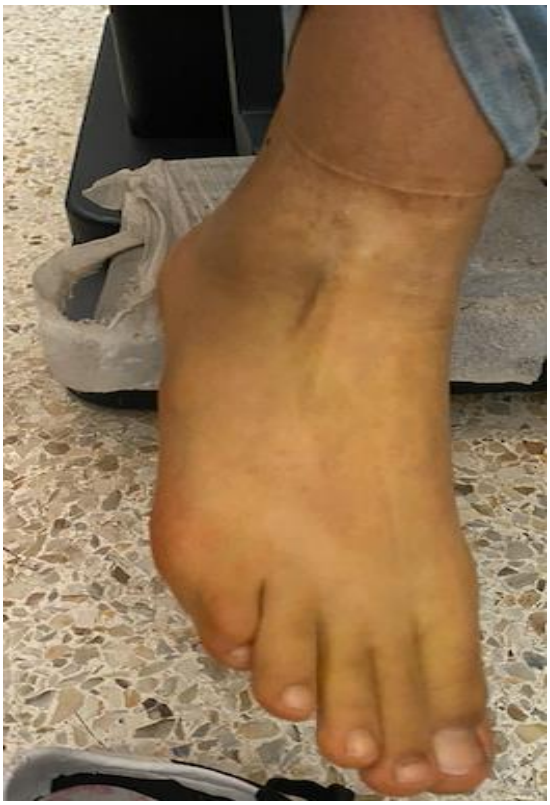
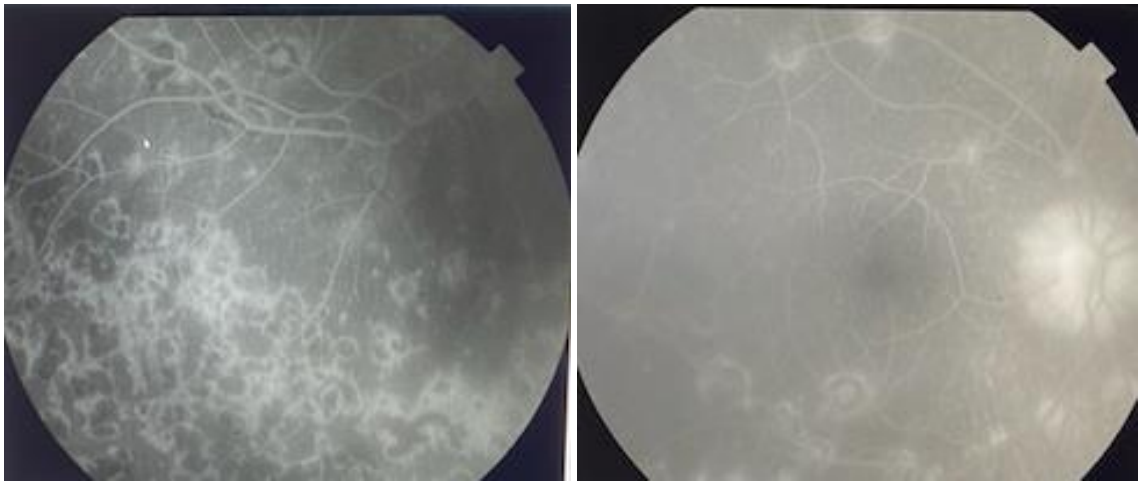


Figure 5: Depigmentation of the forefoot

The child underwent retinal angiography with fluorescence, which showed heterogeneous choroidal filling, atrophy of the pigment epithelium and peripheral pigmentary changes without papillitis and without retinal serous detachment (RSD) (Figures 6 and 7). We completed the study with an optical coherence tomography (OCT), which showed a macular profile at the limit of normal in both eyes, notably no RSD (Figure 8). A lumbar puncture was performed in the child and showed a pleocytosis. A complementary audiogram was done but came back normal.



Figures 6 and 7: Fluorescent angiography showing heterogeneous choroidal filling, atrophy of the pigment epithelium and peripheral pigmentary changes

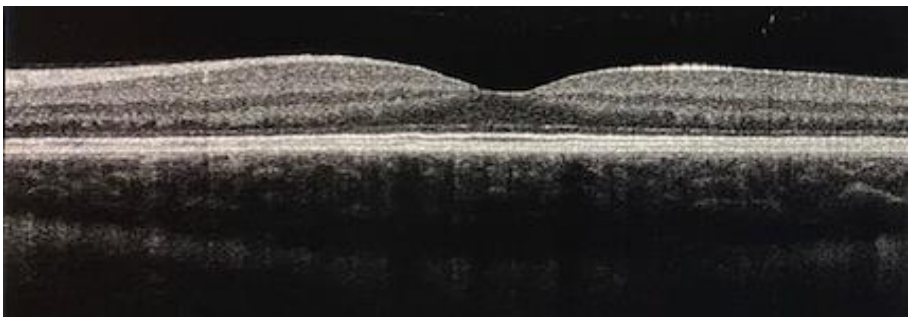


Figure 8: Macular OCT without features

Before deciding on a diagnosis, we carried out an etiological investigation in order to eliminate another disease. Thus, we performed a routine blood test including a blood cell count, C reactive protein, and renal check-up which came back normal. Serology for syphilis, toxoplasmosis and toxocariasis also came back normal. Given the epidemiological context of the country, we completed the test with a quantiferon assay, which came back normal.

In view of these clinical and paraclinical anamnestic elements, based on the diagnostic criteria of the Vogt- Koyanagi-Harada syndrome of the American Uveitis Society revised in 2001, the diagnosis of a complete VKH syndrome was retained [2].

Treatment with three boluses of methylprednisolone (1 g/1.73m2) over three days was initiated, followed by oral therapy combined with topical corticosteroids and cycloplegic therapy.

The short-term evolution was marked by the disappearance of the Tyndall at the end of the 6th day and an improvement of the visual acuity. The long-term evolution was marked by episodes of recurrence of the anterior uveitis. The patient was then put on immunosuppressive treatment with mycophenolate mofetil (30 mg/kg per day in 2 doses).

DISCUSSION

VKH syndrome is a well described cause of uveomeningitis in adults. It is particularly common in women aged 20 to 50 years. It is relatively rare in children [3, 4].

The characteristic manifestation of VKH is a severe bilateral panuveitis associated with retinal serous detachments and neuromeningeal, auditory and dermatological signs [5].

Ocular signs dominate the picture and are the most serious feature of the disease. Lesions may predominate in the anterior or posterior uvea.

Cutaneous signs, often late, occur in 8 to 16% of cases and include poliosis, canitis, alopecia, vitiligo, or hyperpigmented spots [6, 7].

The meningoencephalitis syndrome may be inaugural and go unnoticed as in our patient.

This syndrome classically evolves in four phases: a prodromal phase characterized by the presence of neuromeningeal signs, an acute uveitis phase, a convalescence phase characterized by the appearance of depigmentation of the choroid and the integuments and a recurrence phase. This succession of phases is of major diagnostic interest because it allows

to correct the diagnosis in forms with atypical onset. Currently, new diagnostic criteria seem to allow an earlier diagnosis [2].

VKH syndrome can be difficult to diagnose in children because of its rarity in this field. In the presence of neuro-meningeal signs [8], it may pose the problem of differential diagnosis with a cerebral pathology, in particular a tumor, which should be eliminated by MRI. Similarly, other more frequent causes of uveitis in children, especially infectious causes, must be eliminated. Finally, it is important to emphasize the importance of regular follow-up of any uveitis whose etiology could not be determined in children, because the appearance of late local and/or general signs may lead to reconsideration of the diagnosis [9]. This applies to our patient whose diagnosis was made only in the chronic stage of the disease.

High-dose corticosteroid therapy is the treatment of choice in VKH syndrome in children. This treatment should be given early and in the form of bolus corticosteroids followed by oral corticosteroids in decreasing doses over a long period of time up to two years [10]. In case of corticoresistance or corticodependence, recourse to immunosuppressants and/or immunomodulators is necessary.

The severity of uveitis lies in the frequency of recurrences, in the corticoresistance and in the high incidence of complications such as cataract, glaucoma, epithelial atrophy, which condition the prognosis and explain the poor final visual acuity.

CONCLUSION

Although rare, VKH syndrome exists in the pediatric population and should be considered in the setting of uveomeningitis with multifocal RSD.

Treatment is based on high-dose bolus corticosteroid therapy, mainly followed by oral administration.

Our observation highlights the diagnostic difficulties related to this pathology. We insist on the usefulness of a thorough history in order to retrace the history of the disease and on the necessity of an early diagnosis and treatment to guarantee a better prognosis.

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