Do you know this syndrome?*
*Você conhece esta síndrome?**

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CASE REPORT

A 32-year-old female patient has presented skin lesions for five months. Upon examination, there were well-delimited achromic patches, which were symmetrically located in the temporal and eyebrow regions, dorsum of hands and shoulders. She had poliosis circumscribed to eyelashes, eyebrows and scalp (Figures 1 e 2).

One month before the onset of skin lesions, she presented one episode of intense headache and fever, followed by dysacusia, conjunctival hyperemia and bilateral amaurosis. Therefore, she had to be admitted to hospital and bilateral uveitis was the confirmed diagnosis (Figure 3). CSF analysis revealed meningitis with mononuclear (lymphocytic) infiltration and brain CT scan showed diffuse cerebral edema. At hospital she was given high doses of systemic steroids and recovered from neurological and ophthalmologic conditions after 20 days.

WHAT IS THIS SYNDROME?

Vogt-Koyanagi-Harada Syndrome

The Vogt-Koyanagi-Harada (VKH) syndrome is an inflammatory disease that manifests as a bilateral panuveitis associated with involvement of the central nervous system, auditory system and skin at variable levels.

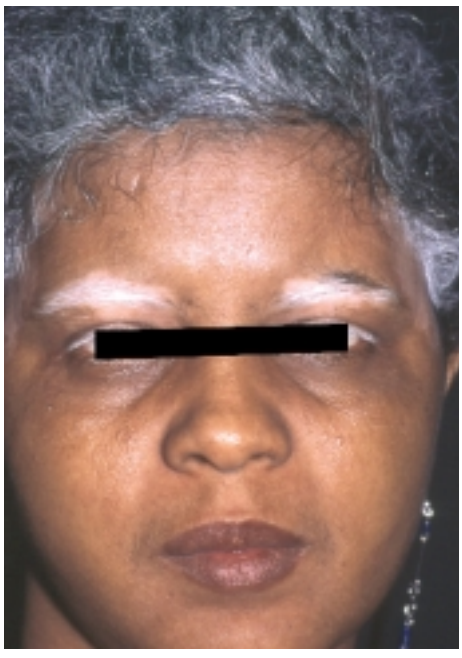

FIGURE 1: Achromic patches in the temporal and eyebrow regions; poliosis

The exact cause is still unknown, but there is some evidence indicating a T-lymphocyte-mediated autoimmune process, targeted against melanocyte-associated antigens, such as tyrosinase, tyrosinase-related proteins and S-100 protein.[1]

The disease is more common in females and most cases occur in the third and fourth decades of life.[2] The prodromes of VKH syndrome include headache, fever, nausea and vomiting and comprise the meningeal involvement phase, with pleocytosis in CSF analysis and a predominance of mononuclear (lymphocytic) cells. In this stage, diverse neurological symptoms may occur, ranging from behavioral alterations, such as psychosis and mental confusion, to significant motor deficits, such as hemiplegia and paraplegia.[3] The disease progresses with ocular alterations, such as photofobia, conjunctival hyperemia, reduced visual acuity and ocular pain. Bilateral panuveitis and specific retinal depigmentation are observed[1] (Figure 4). Fifty percent of patients present auditory symptoms, such as tinnitus and dysacusia.

The skin involvement usually appears within the first three months, soon after the initial ocular symptoms,[4] and the individuals may present poliosis, alopecia or vitiligo. Poliosis is the most frequent sign, followed by alopecia and vitiligo. Although poliosis is characteristically present in eyelashes and eyebrows,

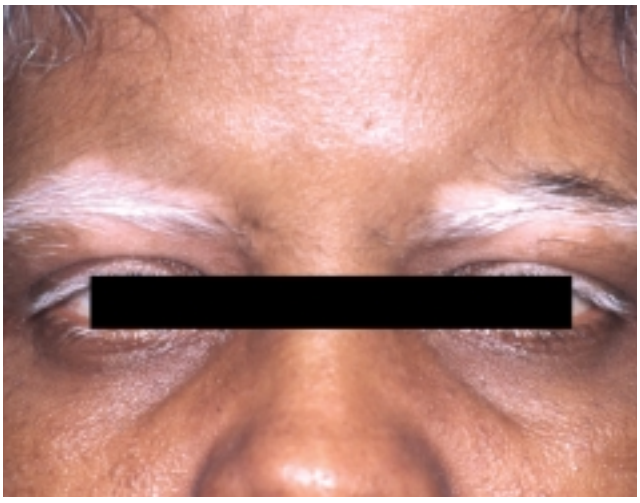

FIGURE 2: Eyelash and eyebrow poliosis

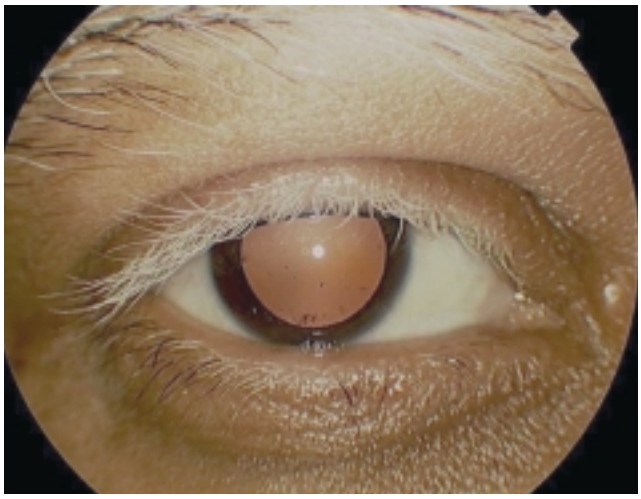

FIGURE 3: Anterior uveitis - keratic precipitates

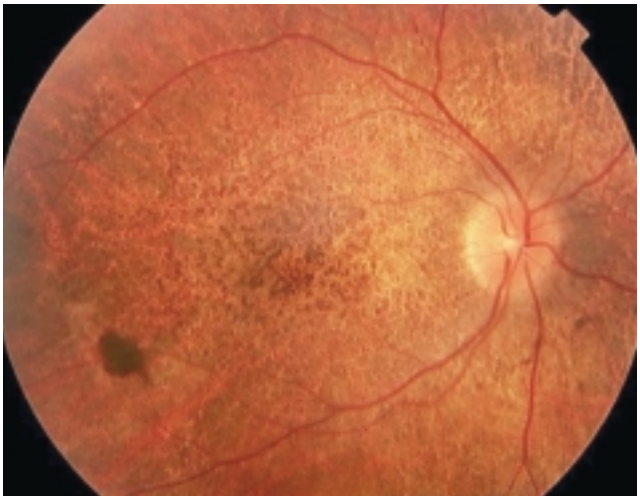

FIGURE 4: Altered retinal pigmentation

it may occur in any type of hair; if extensive, it represents a poor prognosis of skin repigmentation.[3] The vitiligo lesions are usually symmetrical and more often found in the face, neck and shoulders.[5-7]

Skin hypopigmentation tends to not resolve spontaneously, while hearing function is completely recovered. The eye condition progresses with complications, and may result in total amaurosis, but most cases improve.[8-10]

The disease frequently responds to systemic steroids, but this therapy must be aggressive and introduced early, with high doses of steroids in order to reduce possible sequelas.[1,8] The skin disease should be treated with drugs used in vitiligo.[3] ❑

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