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Atypical presentation of ARN syndrome during primary varicella infection in an adult

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Abstract: **Purpose**: To report an unusual case of acute retinal necrosis (ARN) during primary varicella infection in an adult. **Methods**: A 33-year-old man with a history of a recent primary varicella infection transmitted by his son, consulting with redness, pain and reduced visual acuity in the right eye. The eye examination revealed a panuveitis with a whitish retinal focus, located at the extreme periphery. VZV serology shows positive anti-VZV antibody. The search for the chickenpox genome in aqueous humor also came back positive. **Results**: Given the context, the diagnosis of varicella panuveitis retinal necrosis is therefore established. The patient is put on oral valaciclovir (1g every 8 hours), topical corticosteroids and mydriatic eye drops. The evolution is marked by regression of inflammation, the gradual improvement of visual acuity up to 20/20 and healing of the necrotic lesion. **Conclusion**: Earlier diagnosis and effective treatment improves the prognosis of ocular inflammation. In the context of chickenpox with ocular manifestations, acute retinal necrosis should be rigorously investigated. A dilated fundus with a good examination of the retinal periphery is essential.

Index Terms **- Chickenpox; Acute retinal necrosis.**

I. INTRODUCTION

The ocular manifestations associated with the varicella zoster virus (VZV) are mainly related to the reactivation of the virus. Inflammatory ocular involvement in chickenpox is rarely reported [1]. It usually occurs in children and adolescents [2-3]. We describe the case of an adult with uveitis associated with acute retinal necrosis following primary chickenpox virus infection.

II. OBSERVATION:

This is a 33-year-old patient with a history of a primary varicella infection in his 4-year-old son followed by a vesicular rash in the father who does not recall having had a similar episode before (Fig.1a). The patient presents for an ophthalmologic consultation with redness, pain and reduced visual acuity in the right eye that has progressed for 15 days, with an interval of 3 weeks from the onset of the rash. Visual acuity is 20/20 on the right and 20/100 on the left. Biomicroscopic examination of the right eye found conjunctival hyperemia, white granulomatous retro-descemetic precipitates, a three-cross cell tyndall. The vitreous is the seat of a hyalite with a 2-cross cell tyndall and a 2-cross flare. The fundus, hampered by the disturbance of the surroundings, revealed a whitish retinal focus, barely visible, located in the extreme periphery around six o'clock. Examination of the left eye did not show any abnormalities. Dermatological examination found generalized varicella lesions in the process of healing (Fig.1b).

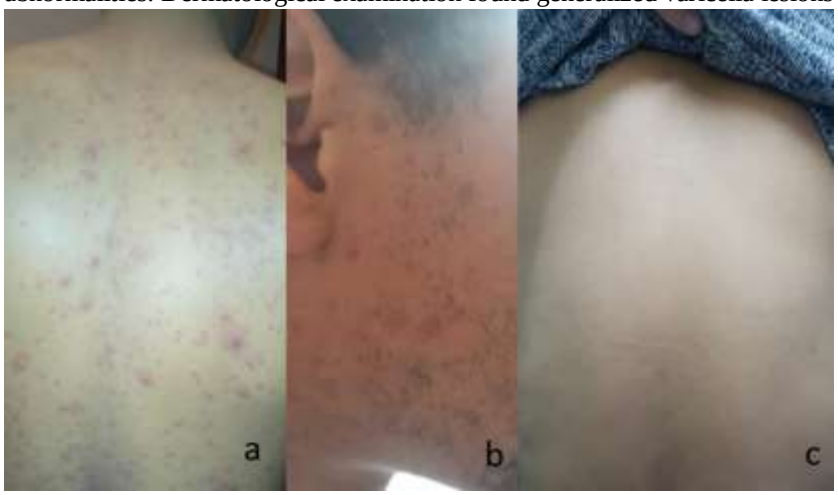

Fig.1: Images of the rash. a: At the initial stage. b: At the time of hospitalization. c: At the scarring stage after 10 months.

The initial fluorescein angiogram, difficult to interpret because of the extent of the inflammation, revealed retinal vasculitis (Fig. 2). Given the context, the diagnosis of varicella panuveitis with very probable retinal necrosis is therefore established. The patient is put on oral valaciclovir (1g every 8 hours), topical corticosteroids and mydriatic eye drops.

Fig.2: The initial fluorescein angiography, difficult to interpret because of the extent of the inflammation, shows a significant leak of fluorescein indicating retinal vasculitis.

In the biological assessment, an inflammatory syndrome is noted with a sedimentation rate of 80 mm the 1st hour. VZV serology shows positive anti-VZV antibody (IgM and IgG type) (Fig.3a). The search for the chickenpox genome in aqueous humor also came back positive (Fig.3b).

Fig.3: a: VZV serology shows positive anti-VZV Ab (IgM and IgG type). b: The search for the chickenpox genome in the aqueous humor is positive.

In The blood serology for HIV is negative. The resumption of the examination did not find any carcinological history or immunosuppressive treatment. The patient is therefore considered immunocompetent. The rest of the serologies are negative (syphilis, toxoplasmosis, lyme disease, cat scratch disease). The quantiferon is also negative. Taking into account the clinical and biological elements, the diagnosis of varicella panuveitis complicated by retinal necrosis in an immunocompetent is retained.

The evolution is marked by the gradual improvement of visual acuity and regression of inflammation at the end of the first week. Oral corticosteroid therapy in decreasing doses is introduced, accelerating the cleansing of the inflammation. The focus of peripheral retinal necrosis, becoming more visible, completes its healing from the center to the periphery (Fig.4a).

Further angiography shows moderate hyperfluorescence in the area of necrosis with minimal leakage of fluorescein into the perivascular space (Fig.4b). Two months later, visual acuity was 10/10 with a fundus showing complete healing of the focus and the persistence of a true flange after hyalitis (Fig.5). Valaciclovir is stopped after 12 weeks of treatment. The patient remains asymptomatic with a follow-up of 10 months (Fig.1c and 4c).

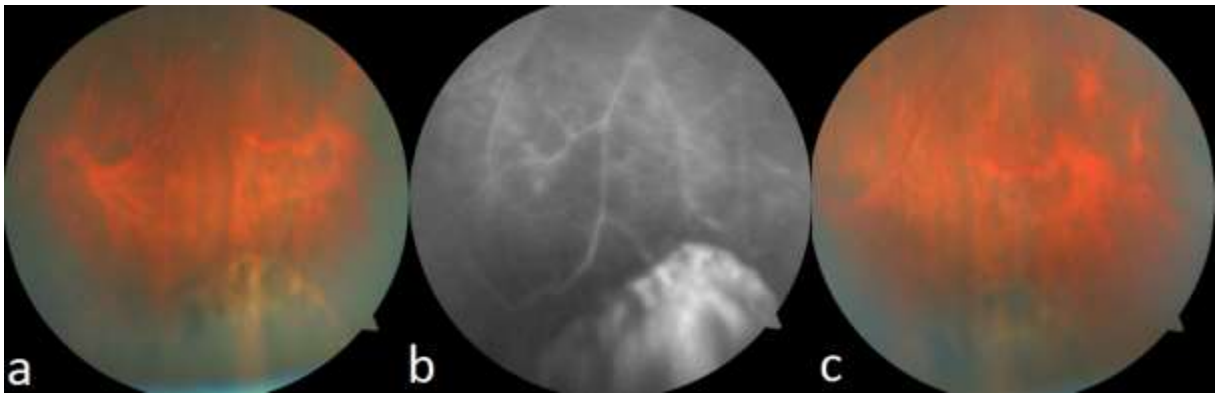

Fig.4: a: The focus of retinal necrosis, in the extreme periphery, is in the process of healing. b: The follow-up angiogram shows moderate hyperfluorescence in the area of necrosis with minimal leakage of fluorescein into the perivascular space. c. Appearance of the focus of retinal necrosis at 10 months of evolution.

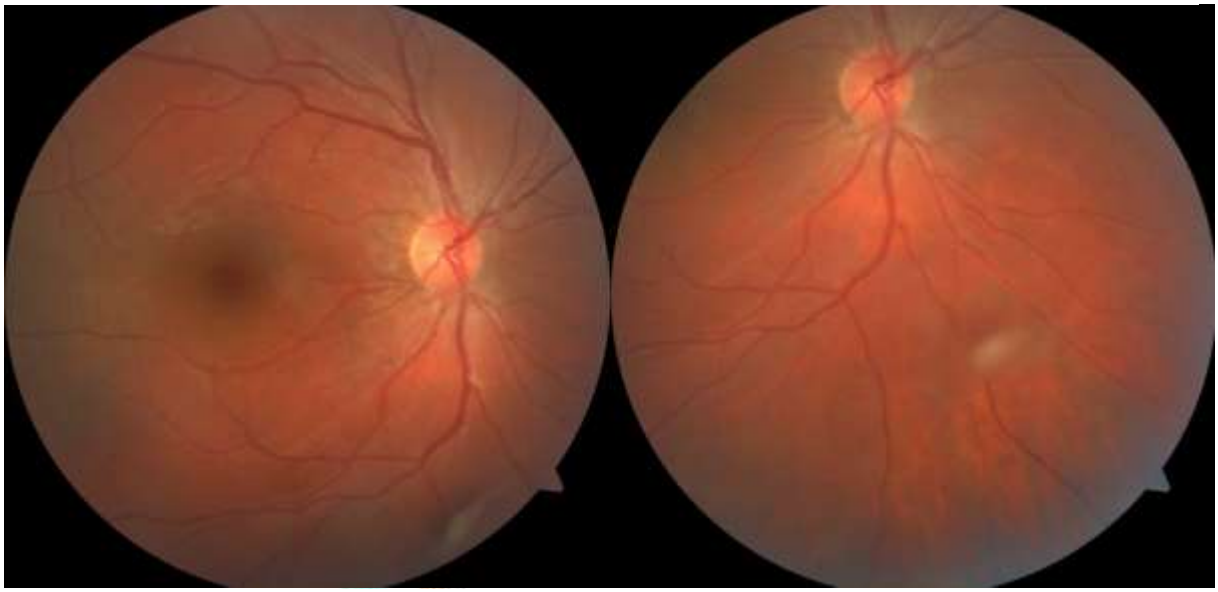

Fig.5: Image showing a true bridle, at 5 o'clock, after hyalite.

III. Discussion

Ocular complications are rarely reported in chickenpox [1, 4-9]. They typically appear within a few days to several weeks of the onset of the rash [10]. Ocular involvement may be related to viral replication or to an immunologic reaction following systemic viral infection [10].

Ocular manifestations associated with chickenpox are diverse including conjunctivitis, scleritis, keratitis, anterior uveitis, acute retinal necrosis, retinal vasculitis, optic neuritis and ophthalmoplegia [1,10]. The involvement can be unilateral or bilateral [1, 4, 8]. Our patient presented with panuveitis with acute unilateral retinal necrosis.

Anterior uveitis in children is usually minimal. It usually requires simple treatment with cycloplegic drugs, sometimes combined with topical corticosteroids to reduce the immune response [10]. In adults, there is little data on the treatment modalities of uveitis associated with chickenpox, given the rarity of reported cases. In all cases, antiviral treatment should be started in the face of severe forms. Acute retinal necrosis (ARN) is rarely reported in chickenpox [11].

The patients described include adults and children, immunocompetent or not [1]. ARN associated with chickenpox appears to be quieter than typical ARN, with slower progression of retinitis, better visual acuity, and a rarer occurrence of retinal detachment [1]. However, regular monitoring is necessary for recurrence of intraocular inflammation or retinal necrosis [12].

IV. Conclusion

Earlier diagnosis and effective treatment improves the prognosis of ocular inflammation. In the context of chickenpox with ocular manifestations, acute retinal necrosis should be rigorously investigated. A dilated fundus with a good examination of the retinal periphery is essential.

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