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# Kaposi Julius-berg Syndrome on Congenital Ichthyosis, about a Case

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***Authors' contributions***

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

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***Case Study***

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## ABSTRACT

**Introduction:** Kaposi-Julius berg syndromeis a rare complication of infection with the virus of the 'herpes, often occurring on a pre-existing skin disease. It mainly affects children, the management of this syndrome is based on antiviral medication with antibiotic therapy. In the absence of diagnosis and early treatment, its prognosis is severe.
**Observation:** We present the first case of kaposi juliusberg syndrome on congenital ichthyosis described. This is a 5-year-old boy followed for congenital ichthyosis since birth, under keratolytic treatment and emollients. He was hospitalized for febrile rash made of lesions vesicular, pustular and crusty. He also presented with bilateral keratitis. The clinical examination showed a fever of 39◦C, an altered general condition with submandibular lymphadenopathy. The skin examination revealed a dozen vesicles and pustules in a bouquet on the face and scalp, meliceric crusts. antibiotic therapy with amoxicillin-clavulanic acid was instituted. The evolution was rapidly favorable. The clinical examination of the mother revealed a recurrence of oral herpes at the origin of the contagion, motivating the introduction of a preventive treatment for recurrences in her.
**Conclusion:** This observation shows that it is essential to inform the family and friends of patients with ichthyosis of the risk of severe herpes infection and to avoid all contact with the herpes virus as far as possible.

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## 1. INTRODUCTION

Hereditary ichthyosis is a disease due to gene mutations involved in the development of the skin barrier, the latter is therefore very deficient. Hereditary ichthyosis begins, in the majority of cases, at birth or in childhood. As with all pre-existing skin diseases, the occurrence of herpes simplex virus type 1 (HSV1) infection responsible for Kaposi-Juliusberg herpetiform pustulosis (PHKJ) is classic; the same is true in atopic dermatitis (AD) as in other dermatological pathologies such as Darier's disease, pemphigus or epidermolysis bullosa (EB) of the Dowling-Meara type and seborrheic dermatitis [1].

Kaposi-Juliusberg syndrome begins with a fever of 40°C, accompanied by severe fatigue and chills. Numerous skin lesions then appear, predominantly on the face (sometimes haemorrhagic pustules, edema and redness of the skin) [2-4].

These lesions can lead to peeling of the skin,necrosesskin and corneal damage. They have the particularity of spreading very quickly over the body, and they are associated with inflammation of thelymph nodes. Bacterial superinfections are common [5-7].

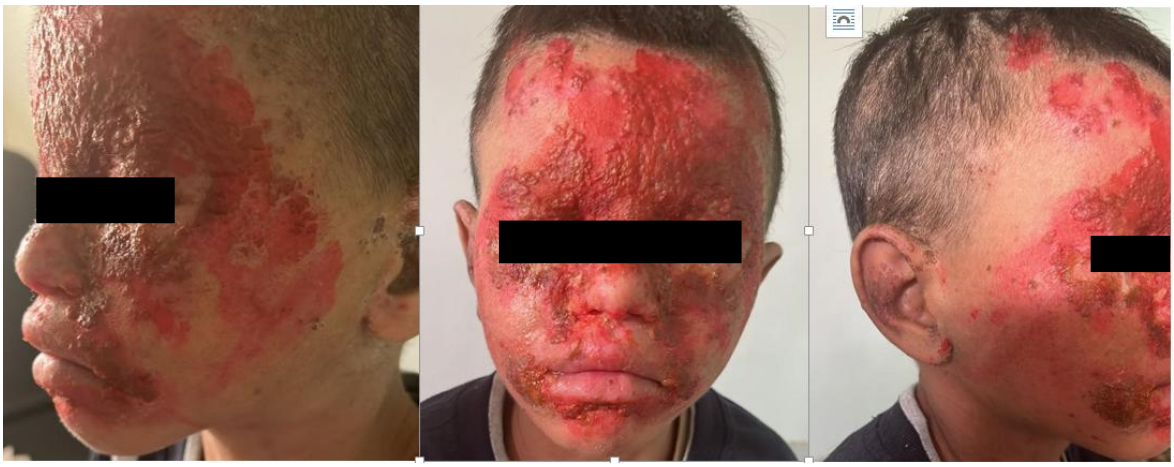
Treatment with aciclovir or its precursor remains very effective, but in congenital ichthyosis there seems to be a need to use these antivirals at an appropriate dosage and rigorous follow-up in order to avoid relapses and complications.

## 2. CLINICAL CASE

We report the singular observation of KAPOSI JULIESBERG in a patient followed for ichthyosis after the free and informed consent of his parents. This is a 5-year-old male child, followed for congenital ichthyosis diagnosed at birth, treated with emollients and moisturizing creams, with a history of herpetic keratitis 3 years ago. He was hospitalized for a vesiculo-pustular febrile eruption, the clinical examination objectified an altered general condition made of anorexia and lethargy, a fever of 39°C, 40°C, the skin examination objectified pustules and vesicles in clusters, crusty, oozing, painful lesions of the face, scalp and pinnae of the ears with a limitation of the opening of the eyes, very evocative of kaposi Julius berg syndrome (Fig. 1). There were infra-centimetric submandibular and retro-auricular adenopathies. Clinical symptoms revealed Kaposi Julius berg syndrome.

Fundus examination showed a dendritic-looking corneal ulcer that is pathognomonic for herpetic keratitis.

Laboratory tests including complete blood count, CRP, kidney and liver function were done to watch for bacteremia or secondary visceral involvement and were found to be normal.



**A)** **B)** **C)**

**Fig. 1. A): Vesiculo-pustular lesions grouped in a bouquet on the eczematous eczematous skin of the face, pavilion eyelids; B): Vesiculo-pustular lesion of the face with ocular pus; C): Vesicular-papular lesions on the pinna of the ears and scalp with ulceration and secondary impetigination**

Injectable treatment with aciclovir at 10 mg/kg/8 h allowed an improvement in the general condition with gain of apyrexia in 48 hours, disappearance of vesicular and bullous skin lesions in 22 days, it was necessary to continue the treatment long-term preventive based on emollients and keratolytics to preserve the skin barrier (Fig. 2).

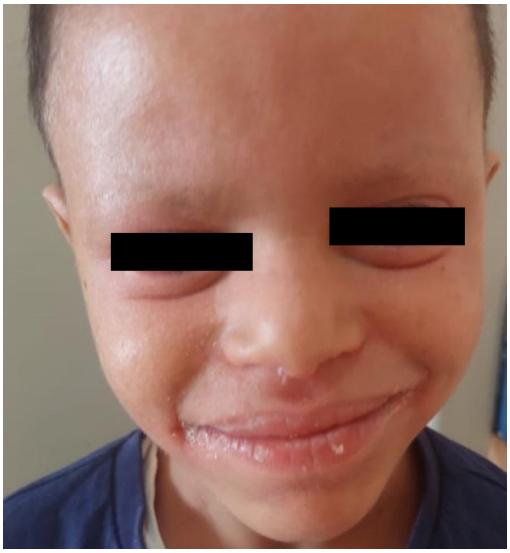


**Fig. 2. Healing of facial lesions after a 22-day follow-up**

Emollients and keratolytics were applied once daily every day of the week + No topical corticosteroids were applied during the active period of the disease.

## 3. DISCUSSION

To our knowledge, no case of Kaposi Julius-berg syndrome on congenital ichthyosis was mentioned in the literature.

Superinfection with HSV1 or VZV is a classic complication of pre-existing skin diseases, cases are currently rarely reported except for a recent publication trying to correlate the intensity of MD (DARIER disease) to the frequency of occurrence of PVKJ (kaposi juliusberg varicella pustullosis) [8]. Most of the publications relate to the occurrence of PVKJ during AD (atopic dermatitis), thus trying to correlate the occurrence of PVKJ with signs of severity of AD or with specific genetic factors, the dosage of aciclovir and valaciclovir are more important than usual [8].

Atopic dermatitis (AD) is the most common inflammatory skin disease in children. The worldwide prevalence of AD is estimated to be 8 to 20%. It is characterized by recurrent chronic pruritic lesions with a different distribution according to age. Eczema herpeticum (EH) (Kaposi's varicelliform eruption) is an acute disseminated viral infection, usually Herpes simplexvirus (HSV) type 1, which develops in the presence of an existing skin lesion, often on the ground of the AD. Eczema herpeticum was first described in children and occurs with higher prevalence in childhood [9].

Psoriasis is fertile ground for a herpetic transplant, however there are only a few cases in the literature, it is a very rare pathology in children [10].

There is a single described case of Dowling-Meara type EB complicated by Kaposi-Juliusberg syndrome [1].

Eczema herpetiformis is associated with a flu-like syndrome, fever, malaise, lymphadenopathy and may be complicated by keratoconjunctivitis, meningitis and encephalitis [9] which testifies to the severity of the prognosis.

These cases show the importance of preventing herpes superinfections linked either to HSV1, VZV or coxackies virus in children with congenital ichthyosis, in the same way as in children with atopic dermatitis [9]. Although there are few reported cases, the prevalence is probably higher. The information of the parents concerning the risk of contagion and the need for eviction must be systematic because there can exist serious clinical forms. This complication should be considered in any child with ichthyosis presenting with a febrile attack of the disease without an infectious point of infection found. In addition, watch out for any ocular symptoms that could be the cause of a complication, such as herpetic keratitis and blepharitis [11].

In our observation, the treatment of herpetic keratitis consisted of ocular washing with isotonic saline serum, intravenous aciclovir and the use of artificial tears without ocular corticosteroid therapy [11]

Morbidity and mortality from Kaposi Julius berg syndrome can be minimized by starting antiviral therapy as early as possible in diagnosis. The main treatment is acyclovir [9]. Systemic antivirals and hospitalization are recommended for severe illness and immunocompromised patients [9] Secondary bacterial infection (mainly

due to Staphylococcus aureus, Streptococcus pyogenes and Pseudomonas which may mask the underlying viral pathogenesis and sepsis are the most important complications, and therefore systemic antibiotic therapy should be added if necessary.

## 4. CONCLUSION

A careful history taken with awareness of risk factors and characteristic skin signs pointing to HE will be helpful for an accurate diagnosis. Laboratory evaluation is not diagnostic.

But useful to support the clinic. Early diagnosis and timely antiviral treatment can prevent major complications and death.

## CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

## ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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