

## An Unusual Presentation of Varicella

Geethu Gangadharan,\* MD, Sebastian Criton, MD

Address: Amala Institute of Medical Sciences, Thrissur

E-mail: drgeethugangadharan@gmail.com

\* Corresponding Author: Dr. Geethu Gangadharan, Amala Institute of Medical Sciences, Department of Dermatology, Amala Nagar, Thrissur, Kerala

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### Abstract

**Observation:** Atypical manifestations and complications are the most common cause of morbidity and hospitalization in commonly regarded self limiting infection like varicella. Hence early recognition of the same is crucial. We report a case of bullous erythema multiforme occurring in the prodrome of varicella.

### Introduction

Varicella caused by varicella zoster virus, is one of the common highly contagious, self limiting viral exanthema with an incidence of about 60 million cases per year worldwide. Besides its classical presentation, varicella infection can manifest with an array of atypical presentations and complications accounting for the morbidity and mortality caused by this infection [1, 2]. These atypical presentations often pose a diagnostic challenge for clinicians and early and prompt recognition of the same is crucial. We report a case of an unusual presentation of varicella as bullous erythema multiforme.

### Case Report

A 2 year old child was brought to our emergency department with a history of fever, fluid filled lesions and erosions over trunk, face and extremities. The child who was apparently normal, was noticed to have an erosion over his forearm. He later developed similar erosions and blisters over his trunk, face and extremities. Two days later, he developed few small vesicles over his body. There was no history of any drug intake prior to onset of symptoms.

But there was a history of chickenpox in the household in the previous month.

On examination, the child was febrile and irritable. Dermatological examination revealed multiple discrete ovoid and targetoid erosions over trunk, face and forearms. A few lesions showed central he-



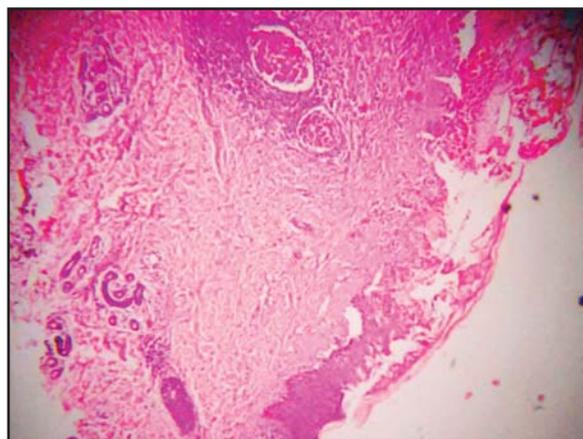
**Figure 1.** Multiple erosions over trunk, face and forearms



**Figure 2.** Ovoid and targetoid erosions, few with a central haemorrhagic crust and scattered haemorrhagic vesicles

morrhagic crust. There were also few scattered hemorrhagic vesicles over trunk (**Figures 1 and 2**). Palms, soles, distal extremities and mucous membrane were spared. Nikolsky sign was negative. Systemic examination was unremarkable. The clinical differential diagnosis considered were bullous erythema multiforme with varicella and childhood pemphigus triggered by varicella.

Hematological parameters were within normal limits. Since the patient had hemorrhagic vesicles, we considered the possibility of impending disseminated intravascular coagulation, but D-dimer value was only 1024 ng/ml. Tzanck smear taken from erosions showed multinucleate giant cells. Skin biopsy showed intraepidermal vesicle formation and necrosis of keratinocytes focally and extending to full thickness of epidermis, which was suggestive of erythema multiforme (**Figures 3 and 4**). Direct immunofluorescence study was found to be normal, thus ruling out the possibility of any immunobullous disease. Hence a diagnosis of bullous erythema multiforme with varicella was made. The patient was treated with intravenous acyclovir 20



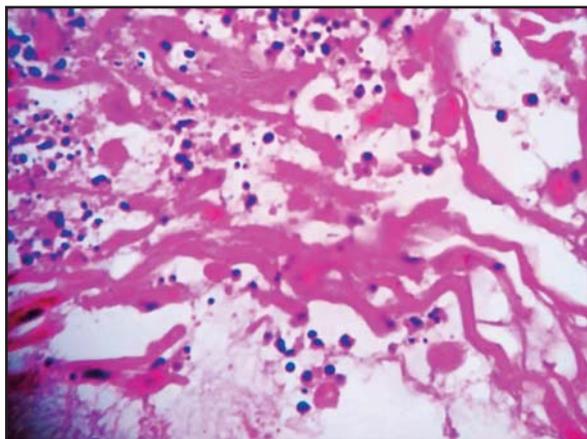
**Figure 3.** Scanner view showing intraepidermal vesicle formation and necrosis of keratinocytes

mg/kg every eight hours for five days and supportive care. The patient showed remarkable improvement with drying of lesions as soon as treatment was initiated. (**Figure 5**).

### Discussion

Although varicella is regarded as a self limiting disease, atypical manifestations and complications seldom occur, especially in immunocompromised individuals, constituting the main cause of morbidity and hospitalization due to this infection [3]. Various documented cutaneous complications of varicella are secondary bacterial infections, varicella gangrenosa, varicella bullosa, hemorrhagic varicella, Steven Johnsons syndrome and erythema multiforme [1, 4, 5]. To the best of our knowledge, only very few cases of varicella infection complicated by bullous erythema multiforme have been reported till now.

Erythema multiforme is a mucocutaneous manifestation of a distinct skin-directed immune reaction that occurs in the setting of an infection in certain predisposed individuals [6]. The common associated infectious agent is HSV, but there are few reports of erythema multiforme occurring few days before and after the onset of varicella rash. Though the exact pathogenesis is not clear, it might be hypothesized as similar to herpes associated erythema multiforme. Varicella DNA fragments maybe transported (by peripheral blood CD34+ Langerhans cell precursors) during the time of secondary viremia, to the keratinocytes and this may lead to the recruitment of varicella virus-specific CD4+ TH1 cells. The inflammatory cascade is initia-



**Figure 4.** High power view showing necrosis of keratinocytes

ted by interferon- $\gamma$  (IFN- $\gamma$ ), which is released from the CD4+ cells in response to viral antigens, and immune mediated epidermal damage begins subsequently [7]. Another probability is the presence of a danger signal like another infection like mycoplasma pneumonia, acting together with varicella, leading to development of erythema multiforme in the prodrome of varicella.

The disease course and sequence of events with regard to appearance of erythema multiforme and varicella rash varies in the previously reported studies. In the previous reports by *Hosoya et al*, *Choy et al* and *Kishore et al*, varicella rash preceded the occurrence of erythema multiforme by two days to more than 12 weeks [8]. Whereas in the report by *Prais et al* [9], erythema multiforme preceded the onset of varicella rash by a few days, which is similar to our case, were it was two days before onset of varicella rash.

### Conclusion

Early recognition of unusual presentations of common infections like varicella is paramount in reducing its morbidity. As in our case, bullous erythema multiforme can present in the prodrome of varicella or after the onset of rash. The rarity of this presentation should not exempt from including bullous erythema multiforme in the list of complications and atypical presentations of varicella.



**Figure 5.** Drying of lesions one day after the treatment was initiated

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