ACUTE GENERALISED PUSTULAR PSORIASIS OF VON ZUMBUSCH – A RARE CASE REPORT

Jayakar Thomas¹, ShriSindhuja T², Chinthaamani KPR², Manoharan D³

¹Professor & Head, ²Junior Residents, ³Professor, Department of Dermatology, Venereology & Leprosy, Sree Balaji Medical College & Bharath University, Chennai 600044, Tamil Nadu, India.

ABSTRACT

Von Zumbusch is the most severe form of generalized pustular psoriasis (GPP) that usually starts as multiple erythematous, tender plaques later becoming studded with tiny sterile pustules that coalesce to form lakes of pus. It can precede the onset of psoriasis or can be a continuum of the disease. We report a case of acute generalized pustular psoriasis of Von Zumbusch in a 30year old Asian male who presented with acute onset of reddish raised lesions associated with pustules all over the body.

INTRODUCTION

Generalized pustular psoriasis is an extreme form of psoriasis that presents with fiery erythematous plaques studded with pustules all over the body. It is classified as: Acute generalized pustular psoriasis of Von Zumbusch, Generalized pustular psoriasis of pregnancy, Infantile and juvenile type, Circinate and Localized (not hands and feet) (Griffiths & Barker, 2008). Von Zumbusch, a type of generalized pustular psoriasis is named after a dermatologist from Germany, Leo Ritter von Zumbusch, who first described the earliest recorded case of generalized pustular psoriasis in 1910 (Sandra Philipp, 2014). It is more commonly seen in adults and the disease runs a more benign course in children. The etiology is unknown but genetic correlation with HLA-B27 has been well established in the past. Clinically it is characterized by fever that lasts for several days associated with sudden generalized eruption of sterile pustules of size 2-3 mm in diameter distributed all over the body including the nail bed, palms and soles.

Case Report: A 30 year old male came to our OPD with complaints of reddish raised lesions associated with pustules all over the body for the past 2 weeks. There was history of fever following which he developed pus filled lesions over the right leg which later progressed to involve all over the body. He had a similar episode 2 years back, for which he had been hospitalized. He had been diagnosed with psoriasis vulgaris 10 years ago and has been on T. Methotrexate for the past 2 years. History of seasonal variation was present (lesions aggravated in the winter season). There was no history of drug intake, topical application prior to the onset of the lesions. No history of itching, oliguria, jaundice or joint pain.

Dermatological examination revealed multiple erythematous scaly plaques with pustules studded over the periphery of the plaque distributed all over the body. Few plaques coalesce to form larger plaques. Sparing of palms, soles and face noted. Scalp, nails and oral mucosa were normal.

Systemic examination revealed normal study. Complete blood analysis done was normal. Punch biopsy taken from the site of the lesion revealed hyperkeratosis, extensive neutrophilic infiltrates in the subcorneal layer with spongiotic pustules.
**DISCUSSION**

Von Zumbusch, a type of acute generalized pustular psoriasis is of two types. In the first, typical psoriasis of early onset progresses to pustular psoriasis after some years often provoked by withdrawal of steroids or other factors and the second type where the onset of psoriasis occurs later in life and is usually atypical, acral or flexural in distribution (Harvey Baker & Terence J. Ryan, 1968). Attacks are characterized by fever that last several days followed by a sudden generalized eruption of sterile pustules which occurs all over the body including the nail bed, palms and soles. The pustules usually arise on the highly erythematous skin lesions and then become confluent as the disease becomes severe leading to erythroderma. Triggering factors are irritant topical therapy such as coal tar, anthralin (Raghavendra Rao et al., 2007) and drugs such as sudden withdrawal of steroids, iodide, lithium, progesterone and terbinafine. Involvement of oral mucosa is not uncommon, with the tongue appearing clinically and histologically synonymous with geographic tongue. Nails may become hyperkeratotic or become separated by subungual lakes of pus and shedding of the nail soon follows. In the absence of effective treatment, death can occur in the acute stage. Complications linked with acute generalized pustular psoriasis are hypoalbuminaemia due to sudden loss of plasma protein into the tissues, followed by hypocalcaemia. Oligemia may cause acute and fatal renal tubular necrosis. Staphylococcal infection may complicate the course of the disease. Inflammatory polyarthritis is common. When the disease lasts for a long period, generalized hair loss may occur. Histopathology study reveals typical features of psoriasis accompanied by spongiosis and pooling of numerous neutrophils in the intraepidermal zone. Laboratory findings in acute generalized pustular psoriasis of Von Zumbusch include absolute lymphopenia at the onset of the disease, followed by polymorphonuclear leukocytosis. ESR is usually raised. Plasma albumin, zinc and calcium may be low partially due to malabsorption. GPP has to be
differentiated from subcorneal pustular dermatosis of Sneddon and Wilkinson (a variant of IgA pemphigus) wherein, sterile pustules have predilection for flexures, axillary and inguinal folds and commonly seen in women. Although subcorneal pustules are seen in both conditions, spongiform pustules are seen only in pustular psoriasis (Amanda Robinson et al., 2012). Successful treatment modalities include withdrawal of provocative factors, acitretin, dapsone, methotrexate, cyclosporine, colchicine, biologic agents such as basiliximab (Salim et al., 2000) and phototherapy (PUVA & NB-UVB) (Herbert Honigsmann et al., 2000). Systemic steroids are used only as a lifesaving measure in case of crisis.

CONCLUSION
Acute generalized pustular psoriasis is a rare but severe type of pustular psoriasis distinctly featured by sterile spongiform pustules, generalized distribution, and systemic symptoms. Timely monitoring and sufficient management is warranted. When untreated or inadequately treated, can be fatal.

ACKNOWLEDGEMENTS: None.

CONFLICT OF INTEREST:
The authors declare that they have no conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS
All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

REFERENCES
Sandra Philipp. Pustular psoriasis. Psoriasis Diagnosis and Management, 2014, chap 9