

# A case of pityriasis rosea of vidal accompanied by neurofibromatosis type 1

by Ahu Yorulmaz, Ferda Artuz, Sezer Kulacoglu, Elif Sen

## COMMENT

**Ass. Prof. Antonio Chuh MD FRCP FRCPCH<sup>1</sup>, Prof. Vijay Zavar MD DNB DV&D FAAD<sup>2</sup>**

*<sup>1</sup>School of Public Health, The Chinese University of Hong Kong and Prince of Wales Hospital, Shatin, Hong Kong,*

*<sup>2</sup>Department of Dermatology, Godavari Foundation Medical College and Research Center, DUPMCJ, India*

**How to cite this article:** Chuh A, Zavar V. Comment: A case of pityriasis rosea of vidal accompanied by neurofibromatosis type 1. Our Dermatol Online. 2015;6(3):375-377.

**Submission:** 17.06.2015; **Acceptance:** 18.06.2015

# The association of *pityriasis circiné et marginé* and neurofibromatosis type 1

## COMMENTS

We read with pleasure the case report by Yorulmaz A *et al* on the occurrence of *pityriasis circiné et marginé* which is a variant of pityriasis rosea (PR) in a 23-year-old lady with known neurofibromatosis type 1 (NF-1) [1].

We agree to both diagnoses [2-4]. Both diseases are uncommon, but definitely not rare. We have no data on the prevalence of NF-1 in Turkey. In the United Kingdom, the prevalence is around 1:4560 [5], with the prevalence at birth being 1:2699 [5]. The prevalences of NF-1 for six-year-old German children, and 9-11 year-old children in Cuba, are 1:2996 [6] and 1:1141 [7] respectively. The prevalence of PR is around 1:167 [8]. This means that if a clinic follows 200 patients with NF-1, and sees them once every year, around 1.20 patient with PR would be expected to be seen.

While reporting them concomitantly in one individual, we might consider exploring the mechanisms as to whether these two diseases: (1) are merely coincidental; (2) are being innocent bystanders (NF-1); (3) are related to the same confounder(s), and (4) have underlying immunopathogenetic connections, which could be risk factors, precipitating factors, or be genuine causal relationships.

For patient with NF-1, the immunological system is compromised to various extents [9-12]. The processes are not comprehensively known, although it is likely that multiple immunological pathways and cellular mechanisms reduce the antigen-processing and antigen-presenting cells in NF-1 [12].

Moreover, large groups of immune function genes in human Schwann cells are down-regulated in NF-1 [13]. Acute phase reactants such as interleukins are reported to be adversely affected in NF-1 [13]. Other than systematic effects, topical immunological responses could also be compromised [11], which might facilitate the inoculation of viruses at the herald patch, a postulation not yet substantiated.

A simplified immunopathological sequence would be: primary viral infection in childhood, the body then

launches a primary and non-specific immunological response, then clonal expansion of T-cells (*memory cells*), then life-long latent infection of the virus in the peripheral blood mononuclear cells, then physical or psychological stresses together with NF-1 weakening the immunity, then endogenous reactivation of the viruses, then secondary immunological response (mainly by cell-mediated immunity), then the visible PR rash. The immunological basis for the predilected sites of lesions in bilateral axillae and groins in *pityriasis circiné et marginé*, however, is completely unknown.

For other paraviral exanthems, eruptive pseudoangiomatosis was reported to be associated with hospitalisations and treatment for cancers [14]. It was postulated that relative immunocompromisation is the missing link. We have reported the association of Gianotti-Crosti syndrome – another paraviral exanthem – and hyperimmunoglobulinaemia E syndrome (Job's syndrome), which is a congenital immunodeficiency disease [15]. Whether relative immunocompromisation is associated with other paraviral exanthems, such as asymmetric periflexural exanthem (unilateral laterothoracic exanthem) and papular-purpuric gloves and socks syndrome, is yet to be investigated.

Finally, we congratulate Yorulmaz A and his colleagues for such an outstanding piece of work which can be applied to patients immediately. We humbly recommend Yorulmaz A *et al* and other investigators to explore the possible associations between relative immunocompromisation and PR or other paraviral exanthems.

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**Source of Support:** Nil, **Conflict of Interest:** None declared.