

Sparing phenomenon of cutaneous small-vessel vasculitis around solar lentigines and peripheral halo.

Thanisorn Sukakul MD,
Supenya Varothai MD.

ABSTRACT :

SUKAKUL T, VAROTHAI S. SPARING PHENOMENON OF CUTANEOUS SMALL-VESSEL VASCULITIS AROUND SOLAR LENTIGINES AND PERIPHERAL HALO. THAI J DERMATOL 2017; 33: 292-296.

DEPARTMENT OF DERMATOLOGY, FACULTY OF MEDICINE SIRIRAJ HOSPITAL, MAHIDOL UNIVERSITY, BANGKOK, THAILAND.

This is a case presented with biopsy-confirmed diagnosis of cutaneous small-vessel vasculitis sparing solar lentigines and peripheral normal skin called "halo effect". A 69-year-old healthy Thai male presented with asymptomatic, slowly progressed solar or senile lentigines for more than 10 years with new emerging rashes on legs which were diagnosed as cutaneous vasculitis by clinical and skin biopsy, appeared around halo area adjacent to solar lentigines. Recently, many types of sparing phenomena in dermatology have been reported, but there is no report of this unusual presentation of sparing phenomenon of cutaneous vasculitis in lentigines. However, the pathogenesis still remains unknown, probably related to immunological mechanisms between these 2 conditions which need further studies to identify the cause.

Key words: cutaneous vasculitis, solar lentigines, sparing phenomenon, halo effect

From: Department of Dermatology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand

Corresponding author: Thanisorn Sukakul MD, e-mail: kimthanisornsu@gmail.com

บทคัดย่อ :

ธนิศร สุขะกุล สุเพ็ญญา วโรทัย ปรากฏการณ์บริเวณเว้นว่างของผื่นหลอดเลือดขนาดเล็กอักเสบที่ผิวหนังรอบกระแดดและผื่นหนังปกติข้างเคียง วารสารโรคผิวหนัง 2560; 33: 292-296.

ภาควิชาตจวิทยา คณะแพทยศาสตร์ศิริราชพยาบาล มหาวิทยาลัยมหิดล

รายงานผู้ป่วย ได้รับการวินิจฉัยผื่นหลอดเลือดขนาดเล็กอักเสบที่ผิวหนัง ซึ่งมีบริเวณเว้นว่างของผื่นรอบกระแดด และผื่นหนังปกติข้างเคียง ผู้ป่วยชายไทยอายุ 69 ปี มาตรวจด้วยเรื่องกระแดดที่ไม่มีอาการ ค่อยๆขยายขึ้นในช่วง 10 ปี และมีผื่นหลอดเลือดผิวหนังอักเสบขึ้นใหม่ ซึ่งวินิจฉัยโดยอาการแสดง และการตัดชิ้นเนื้อตรวจทางพยาธิวิทยาเกิด-บริเวณรอบกระแดดและผื่นหนังปกติข้างเคียง ในปัจจุบันมีรายงานปรากฏการณ์บริเวณเว้นว่างของผื่น ในโรคผิวหนังหลายชนิด แต่ยังไม่เคยมีรายงานในผู้ป่วยหลอดเลือดอักเสบร่วมกับกระแดด

โดยสาเหตุการเกิดโรคยังไม่ทราบแน่ชัด อาจเกี่ยวข้องกับกลไกด้านภูมิคุ้มกันระหว่าง 2 ภาวะนี้ ซึ่งต้องศึกษาเพื่อหาสาเหตุต่อไป

คำสำคัญ: หลอดเลือดอักเสบที่ผิวหนัง, กระแดด, ปรากฏการณ์บริเวณเว้นว่าง (sparing phenomenon), ปรากฏการณ์แอมัลซี (halo effect)

Case report

A 69-year-old healthy Thai male had multiple discrete dark brown macules and well-defined round patches on trunk and extremities which slowly progressed in size and number for more than 10 years. The lesions were all asymptomatic. Patient did not concern about these lesions, so they had not been treated. His complaint was recent, mild itchy rashes developed for 3 days around spared area of the pre-existing pigmented lesions. The patient had never initiated any treatment and denied history of trauma. There were neither fever nor systemic symptoms. Skin examination showed ill-defined non-blanchable erythematous to violaceous macules confluent into patches on both thighs, spared the area of previous persistent

pigmented lesions leading to 1-4 centimeters of normal skin around these lesions. (Fig.1) Physical examination of other systems was normal. The pigmented lesions were clinically diagnosed as solar or senile lentigines (SLs), while new emerging rashes were provisional diagnosed as cutaneous vasculitis, so skin biopsy was done to confirm the diagnosis. Laboratory investigations relative to vasculitis including complete blood count, liver function test, renal function test, urinalysis and hepatitis profiles were found to be normal.

Skin tissue was sent for histopathological (H&E) and direct immunofluorescent studies. Epidermis revealed normal. In dermis, superficial blood vessels showed perivascular infiltration of neutrophils and lymphocytes, fibrinoid necrosis

with red blood cells extravasation and nuclear dusts. Blood vessels in deep dermis and subcutaneous fat were unremarkable. Direct immunofluorescence showed positive complement 3 (C3) 1+ (moderate) at superficial blood vessels, positive immunoglobulin-M (IgM) 1+ (moderate) and immunoglobulin-A (IgA) 1+ (few) at colloid body. These findings can be seen in vasculitis. Patient was finally diagnosed as primary cutaneous small-vessel vasculitis (CSVV) and treated with oral colchicine (0.6 mg) twice daily plus topical betamethasone dipropionate 0.05% cream. The symptom was dramatically improved; the vasculitis lesions were totally disappeared within 2 weeks without regression of SLs.

Discussion

Cutaneous small-vessel vasculitis (CSVV) is a neutrophilic inflammation disorder of superficial cutaneous postcapillary venules which can be classified into idiopathic which is known as "primary CSVV" and secondary CSVV which may have underlying conditions such as infection, medications, autoimmune diseases, or malignancy. Most of CSVV usually affect only in the skin, but can be associated with other organs involvement. The clinical presentation usually shows multiple symmetrical non blanchable erythematous to violaceous macules or papules of the lower extremities and other dependent areas of the body. Pain or pruritus symptoms

may be found. The diagnosis is usually confirmed by skin biopsy for histological and direct immunofluorescent studies.¹



Figure 1 Clinical presentation: ill-defined non-blanchable erythematous to violaceous macules confluent into patches, spared the area of lentigines and peripheral halo.

This patient was diagnosed as solar lentigines (SLs) and cutaneous vasculitis. On one hand, the former has no doubt with the clinical presentation to make a diagnosis. On the other hand, the later was clinically suspected and had been confirmed by skin histology and direct immunofluorescent study. The physical examination and laboratory results revealed

normal which were compatible with primary CSW. Interestingly, the clinical feature of CSW in this patient was not typical because they were appeared around the halo area of SLs.

In dermatology, many have reported sparing phenomena in both anecdotal case reports and typically diagnostic significance such as anatomic phenomenon, atopic dermatitis spares the napkin area, rosacea spares circum-oral and periorbital area, and tinea cruris spare scrotum.² A case of isomorphic non response or reverse Koebner phenomenon has been found in cutaneous vasculitis, described the sparing area of skin on left leg previously covered with bandage. However, the pathogenesis is still not understood.³ Moreover, there are only few cases of the purpuric rash appeared sparing a tattoo. The proposed etiology of these reports was the use of Hamamelis virginiana in tattoo ink that has anti-reactive oxygen species and anti-tumor necrosis which suggests immunological response may play a role.⁴⁻⁵

Solar or senile lentigines (SLs) is an epidermal pigmentary disorder presents with macules or well-defined patches of hyperpigmentation usually on sun-exposed area of the skin of the elderly. The etiology of SLs is not completely defined, probably related to ultraviolet exposure causes inflammation and dysregulation of pro-melanogenic factors. Not only the abnormalities found to be occurred in epidermis caused by

keratocytes and melanocytes, but in vitro models showed photoaged fibroblast in dermis also plays a significant role.⁶⁻⁷ Moreover, an up-modulation of vascular endothelial growth factor can cause an increased number of blood vessels in pigmented lesions.⁸ Halo phenomenon has been described in melanocytic tumors of the skin, especially in nevi which represents benign regression, mostly involved by T lymphocytes.⁹ Thus, SLs lesions are shown to have immunological mechanisms which may interfere the pathogenesis of CSW. Pseudo-halo nevi and melanoma also have been reported due to use of sunscreen on affected skin, resulted in area of halo adjacent to pigmented lesions.¹⁰ Thus, this patient denied history of sunscreen, corticosteroids or other topical substances application on halo area before rash appeared, so the feature is unlikely to be pseudo-halo phenomenon or effects of any topical substances.

Recently, there was no report of sparing phenomenon of cutaneous small-vessel vasculitis around solar lentigines with peripheral halo. Altogether, the etiology of this phenomenon probably related to structural changes and immunologic response of epidermis, underlying and adjacent area of dermis of SLs which may able to inhibit neutrophils aggregation normally occurs in CSW. However, the exact pathophysiology of this

sparing phenomenon is still under-discovered due to limitation in number of cases.

References

1. Goeser MR, Laniosz V, Wetter DA. A practical approach to the diagnosis, evaluation, and management of cutaneous small-vessel vasculitis. *Am J Clin Dermatol* 2014; 15: 299-306.
2. Pakran J. Sparing phenomena in dermatology. *Indian J Dermatol Venereol Leprol* 2013; 79: 545-50.
3. Yadav S, De D, Kanwar A. Reverse koebner phenomenon in leukocytoclastic vasculitis. *Indian Journal of Dermatology* 2011; 56: 598-9.
4. Pinal-Fernandez I, Solans-Laqué R. The 'sparing phenomenon' of purpuric rash over tattooed skin. *Dermatology*. 2014; 228: 27-30.
5. Chen CA, Mikailov A, Faulkner-Jones B, Wu PA. Leukocytoclastic vasculitis sparing a tattoo with halo effect. *JAAD Case Rep* 2015; 1: 269-71.
6. Bastonini E, Kovacs D, Picardo M. Skin Pigmentation and Pigmentary Disorders: Focus on Epidermal/Dermal Cross-Talk. *Ann Dermatol* 2016; 28: 279-89.
7. Goorochurn R, Viennet C, Granger C, et al. Biological processes in solar lentigo: insights brought by experimental models. *Exp Dermatol* 2016; 25: 174-7.
8. Hasegawa K, Fujiwara R, Sato K, et al. Increased blood flow and vasculature in solar lentigo. *J Dermatol* 2016; 43: 1209-13.
9. Munding GS. Images in clinical medicine. Halo phenomenon. *N Engl J Med* 2014; 370: 262.
10. Suzuki N, Dalapicola M, Argenziano G, et al. Halo and pseudo-halo melanoma. *J Am Acad Dermatol* 2016; 74: e59-61.