



Halo formations around senile hemangiomas in diffuse plane normolipemic xanthomatosis associated with monoclonal gammopathy

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Key words: diffuse plane normolipemic xanthomatosis; free light chain; halo; monoclonal gammopathy; monoclonal gammopathy of undetermined significance; paraprotein; senile hemangioma; yellow skin; xanthoma.

INTRODUCTION

Diffuse plane normolipemic xanthomatosis (DPNX) is a rare form of xanthoma characterized by yellow macules or slightly elevated plaques not associated with hyperlipidemia. It is frequently accompanied by various hematologic diseases, particularly monoclonal gammopathy of undetermined significance (MGUS) and multiple myeloma.¹ Here we present a case of DPNX associated with MGUS, which showed unique halo formations around senile hemangiomas. We discuss the relationship between the distribution of skin lesions of DPNX and blood flow.

CASE REPORT

A 54-year-old man presented with asymptomatic widespread yellow macules that had persisted for a year. On examination, the macules were nonelevated, well-circumscribed lesions, distributed symmetrically on the inner aspect of the forearms, axillae, posterior neck, shoulders, back, and thighs but not on the face (Fig 1, A through C). Halo formations around senile hemangiomas were observed (Fig 2). Dermoscopically, the macules showed diffuse yellow pigmentation, but the areas of halos were devoid of yellow pigmentation. The patient's medical history included idiopathic lobular panniculitis, of which systemic symptoms had been repressed by tocilizumab for the previous 4 years.

Abbreviations used:

DPNX: diffuse plane normolipemic xanthomatosis
MGUS: monoclonal gammopathy of undetermined significance

A punch biopsy specimen taken from the forearm found foam cells around the capillary vessels in the papillary and reticular dermis. A laboratory examination found low C4 (<2 mg/dL) and high level of serum λ free light chain (λ 165 mg/L, κ 8.3 mg/L). Serum immunofixation electrophoresis showed monoclonal bands of IgG with λ free light chains. Complete blood cell counts and serum chemistry were normal. Values for C3, C1 inhibitor, cryoglobulins, low-density lipoprotein, high-density lipoprotein and triglycerides were within normal ranges. Bone marrow aspiration demonstrated a normocellular marrow, with 3.4% consisting of plasma cells. Flow cytometric analysis found that CD38⁺ cells in bone marrow exhibited λ free light chain restriction. No bone lesions were found on computed tomography. From these results, DPNX accompanied by MGUS was diagnosed.

DISCUSSION

The pathogenesis of DPNX remains uncertain. It has been attributed to paraprotein-lipoprotein

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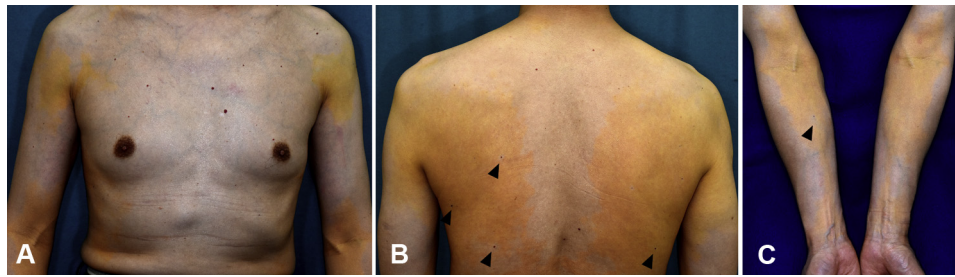


Fig 1. Diffuse plane normolipemic xanthomatosis. Nonelevated well-circumscribed yellow macules were distributed symmetrically on the axillae (A), back (B) and inner aspect of the forearms (C). Arrowheads indicate the position of senile hemangiomas.

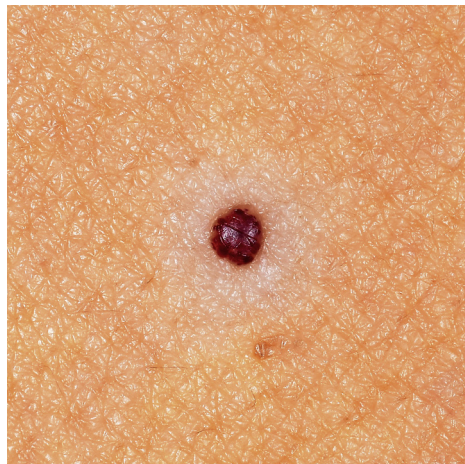


Fig 2. Halo formations around senile hemangiomas. Normal skin color was observed around senile hemangiomas in the xanthomatous lesions.

immune complexes that may activate complement and induce the consumption of serum C4 and C1 inhibitor. The complexes are subsequently phagocytosed by macrophages, leading to cholesterol accumulation.^{1,2} The distribution of the xanthomatous lesions in DPNX is unique. Common sites are the inner canthi and eyelids, axillae, upper back, and cubital fossae, although the distribution varies among cases.¹⁻⁵ The mechanism of the distribution is unclear, but the halo formations around the senile hemangiomas could be a clue toward elucidating the pathogenesis. Lower blood flow might be associated with the avoidance of lipid deposition because an anemic halo is seen in some cases of senile hemangiomas.⁶ In our case, anemic halos were also observed around senile hemangiomas in nonxanthomatous areas (data not shown). The result of dermoscopy suggested that the halo formation was due to the absence of yellow discoloration, and not merely to a reduced capillary blood flow.

Data of body surface temperatures might support the hypothesis concerning blood flow. Areas with a

high body surface temperature in healthy individuals measured by thermography correspond well to the common sites of DPNX.⁷⁻¹⁰ For example, the inner canthi, which are the warmest sites of the face,^{9,10} are also the most common sites of DPNX. Additionally, the anterior aspect of the arms, also a common site for DPNX, is warmer than the posterior aspect.^{7,8} Xanthomatous lesions avoid areas near the nipples/areolae,^{2,3} which are cooler than other parts of the trunk.⁷ These examples suggest that a high body surface temperature, which is related to high blood flow in the capillaries of the skin, might enhance xanthoma formations.

In our case, there were no skin lesions in the face or the upper back, which was inconsistent with the data of body surface temperatures. Variety in distribution among cases may rely on other physiologic factors, such as age and perspiration, and environmental factors, such as climate, clothing, and daily lifestyles. Further physiologic, histopathologic and chemical analyses of lesional/nonlesional skin may elucidate the pathophysiology of DPNX.

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