Primary Cutaneous Angioplasmocellular Hyperplasia

– Case Report –

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Primary cutaneous angioplasmocellular hyperplasia was first described by Gonzalez in 1995 as an asymptomatic, non-ulcerated, slightly umbilicated and violaceous nodule. To the best of our knowledge, only one additional case has been reported since the first two cases. We describe the first two patients in Taiwan. A 39-year-old man had a painless, erythematous, approximately 1-cm nodule with a dark red halo on the lower back. A 35-year-old woman presented with a friable nodule that bled easily and had peripheral erythema on the upper chest wall. Neither patient had a history of local trauma or previous lesions. Histopathologic examination of the excisional biopsy specimen from both showed hyperkeratosis and acanthosis of the epidermis. In the upper dermis, there was abundant capillary-like proliferation of blood vessels surrounded by abundant plasma cell infiltrates. It is essential to differentiate this entity from pyogenic granuloma, Kaposi’s sarcoma, epithelioid hemangioendothelioma, arthropod bite reactions, and acral pseudolymphomatous angiokeratoma. (Dermatol Sinica 22 : 317-320, 2004)

Key words: Primary cutaneous angioplasmocellular hyperplasia, Pyogenic granuloma, Kaposi’s sarcoma, Epithelioid hemangioendothelioma, Arthropod bite reactions, and Acral pseudolymphomatous angiokeratoma

原發性皮膚血管漿細胞性增生

―病例報告―

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原發性皮膚血管漿細胞性增生於1995年由Gonzalez首次描述，其爲無症狀、無潰瘍，稍微中凹且紫色的結節。據吾人所知，自首兩例報告之後只有一名新增病例。吾人報告台灣首兩名病例。一名39歲男性，在下背有一個無痛性紅色且直徑約一公分的結節，並在其周圍有暗紅色的光暈。另一名35歲女性病患，於前胸有一個脆弱且易流血的結節，並在其周圍有紅斑。此兩名病人於該部位過去並無局部外傷或其它病灶的病史。兩者的皮膚切除組織病理學檢查顯示表皮過度角化及棘層肥厚。在表淺真皮層中有大量的微血管似的血管增生，而其周圍有許多漿細胞浸潤。此疾病必需與化膿性肉芽腫，卡波西氏肉瘤，上皮樣血管內皮瘤，節肢動物叮咬反應，及肢端假淋巴瘤性血管角化瘤做鑑別診斷。（中華皮誌22 : 317-320, 2004）
INTRODUCTION

Primary cutaneous angioplasmocellular hyperplasia is a distinctive, rare form of blood vessel proliferation with a reactive plasma cell infiltrate. It is important to distinguish it from other skin lesions with vascular proliferation. We report the first two cases in Taiwan.

CASE REPORTS

Case 1 is a 39-year-old man, who had noted an asymptomatic, erythematous, approximately 1-cm nodule with a dark red halo on the right side of his lower back for several months (Fig. 1A). He had no history of local trauma or previous lesions but a history of gout and asthma. Case 2 is a 35-year-old woman presented with a friable nodule that bled easily and had peripheral erythema on the upper chest wall for about several months (Fig. 1B). She denied previous lesions in that area or trauma. Her general examination was unremarkable and laboratory tests were within normal limits. A complete excision of the two skin lesions was performed. Histopathologic examination of both the excisional biopsy specimens demonstrated similar findings. There was hyperkeratosis and acanthosis of the epidermis (Fig. 2). In the dermis, there was capillary-like proliferation of blood vessels characterized by focally plump and partly vacuolated endothelial linings surrounded by

Fig. 1
(A) An asymptomatic, erythematous, approximately 1-cm nodule with a dark red halo was on the right lower back in patient 1. (B) A friable nodule with peripheral erythema on the upper chest wall in patient 2.

Fig. 2
Pathology of biopsy specimen from patient 1 showed hyperkeratosis and acanthosis of the epidermis and dermal proliferation of blood vessels. (H & E, X20)

Fig. 3
A higher magnification from patient 2 showed diffuse plasma cell infiltration and proliferation of thin-walled and thick-walled blood vessels. (H & E, X100)

Fig. 4
Abundant infiltration of mature plasma cells with occasional Russell bodies were identified. (H & E, X400)
an abundant infiltrate of mature plasma cells and a few lymphocytes and histiocytes (Fig. 3). Neutrophils, eosino-phils and germinal centers were not evident. Russell bodies were occasionally identified (Fig. 4). No amorphous eosinophilic hyaline globules were observed. Periodic acid-Schiff, Giemsa and Warthin-Starry stains were applied to the tissue sections, and there were no microorganisms noted. Mouse anti-human κ and λ light-chain antibodies (DAKO antibody, code number: M7077) were applied, and the plasma cells showed immunoreactivity for both types of chain in a polyclonal staining pattern (Fig. 5). The ratio of κ-expressing to λ-expressing cells was approximately 1:1.5.

Neither patient had evidence of recurrence of the lesion after biopsy.

**DISCUSSION**

Since Gonzalez first described primary cutaneous angioplasmocellular hyperplasia in 1995, only three cases have been reported. Clinically, primary cutaneous angioplasmocellular hyperplasia is an asymptomatic, violaceous, non-ulcerated, slightly umbilicated nodule. The etiopathogenesis is unknown. One hypothesis has been proposed in an oral mucosal lesion with similar histological features. The author thought a dense plasma cell infiltrate in the upper submucosa may alter the local blood flow that causes vasodilation and proliferation. However, it is difficult to apply this pathogenesis in a cutaneous lesion, that is usually absent of plasma cells. Another possible mechanism is a dense reactive plasma cell infiltration within a hemangioma. Therefore, this unique lesion may also be considered a variant of cutaneous pseudolymphoma and is reactive in nature.

The diagnosis of primary cutaneous angioplasmocellular hyperplasia is made histopathologically in the presence of proliferation of blood vessels and infiltration of dense, mature, polyclonal plasma cells. There were several findings mentioned in a previous ultrastructural study, including abundant thin cytoplasmic filaments within the prominent endothelium of blood vessels, increased multivesicular bodies and micropinocytosis. Among duplicated basal lamina, pericytes were also noted.

The differential diagnosis includes pyogenic granuloma, in which proliferating capillaries appear within an edematous stroma and are surrounded by a dense neutrophilic infiltrate. Although a pyogenic granuloma with chronic inflammation may have a prominent plasma cell infiltrate, the architecture of epidermal collarette and lobular capillary proliferation is helpful in differentiation from angioplasmocellular hyperplasia.

The presence of blood vessels intermingled with plasma cells may also suggest Kaposi’s sarcoma. However, the latter also contains atypical spindle cells and poorly defined slit-like vascular spaces. In addition, Kaposi’s sarcoma may have a promontory sign and irregular proliferation of vascular channels dissecting the collagen bundles.

Epithelioid hemangioendothelioma has an infiltrative growth pattern of ovoid, cuboidal, or short spindle cells with a prominent eosinophilic
Many tumor cells demonstrate hyperchromatic nuclei and intracytoplasmic lumina with erythrocytes. Skin involvement is rare and usually associated with an underlying soft tissue or bone lesion or with multicentric disease. The focally prominent endothelial lining is similar to that found in angioplasmocellular hyperplasia, but the conspicuous plasma cells and bland appearance of the vessels in our patients’ biopsy specimens were inconsistent with the diagnosis of epithelioid hemangioendothelioma.

A chronic arthropod bite reaction may also be included in the differential diagnosis. However, such lesions usually include spongiosis or a wedge-shaped inflammatory cell infiltrate.

Acral pseudolymphomatous angiokeratoma is characterized by epidermal acanthosis, a dense nodular lymphoid infiltrate in the upper dermis, and abundant dilated thickened capillaries lined by plump endothelial cells. However, the infiltrate in this lesion includes a mixed population of lymphocytes, plasma cells, histiocytes, and a few eosinophils and multinucleated giant cells, as opposed to primarily plasma cells.

Plasma cells occur in neoplasms and related disorders of lymphoid cells, but they can also be found in a wide spectrum of common inflammatory dermatoses and infections. Plasma cell granuloma of the oral mucosa with angiokeratomatous features may be the counterpart of primary cutaneous angioplasmocellular hyperplasia. A fuller understanding of this disorder will likely require the finding of more cases.

REFERENCES