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BRIEF REPORT

Acquired Port-Wine Stain Following Minor Trauma: Fegeler Syndrome

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Dear Editor:

Fegeler syndrome, also known as trauma-related acquired port-wine stains (PWS), is a rare skin disease that occurs after birth. Its morphological characteristics of well-demarcated, bright red to purple colored macules or patches are the same as those observed in congenital PWS. While most cases of congenital PWS involve lesions located in the head and neck region, there is no dominant site where acquired PWS can occur. Hence, acquired PWS can appear anywhere, including the head and neck region. As per the literature, various causes, such as trauma, hormones, thermal damage, and solar damages, may contribute to the occurrence of acquired PWS. On histological examination, acquired PWS also shows features identical to congenital PWS, characterized by multiple dilatations of capillaries in the upper dermis with normal endothelial cells. The differential diagnosis for acquired PWS includes angioma serpiginosum (AS), and unilateral nevoid telangiectasia (UNT) in terms of showing acquired and dilation of capillaries. Dermoscopic findings such as rounded, globular vessels in acquired PWS, demarcated red lagoons in AS, and thin and tortuous linear vessels in UNT are helpful for differential diagnosis[1].

Recently, we observed a 28-year-old Korean female patient presenting with persistent erythematous patches on the right-hand dorsum and right forearm (Fig. 1). She had been fitted with an orthopedic cast due to a sprain on the right hand and right forearm as a 6-year-old. After the cast was removed, she discovered that the skin lesions appeared abruptly. A 3-mm punch biopsy was performed on

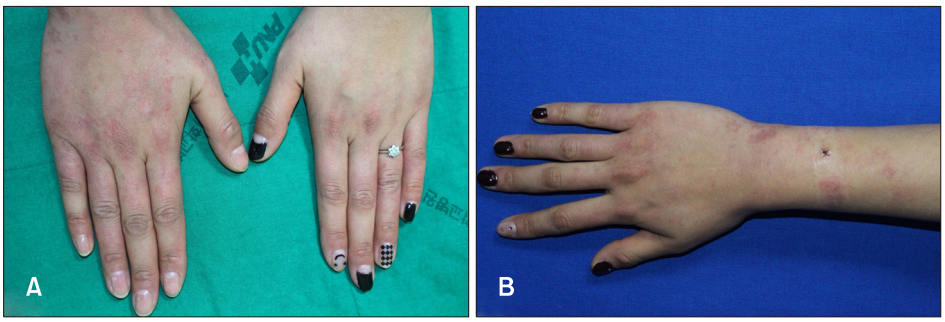

Fig. 1. Erythematous to purpuric patches on the right fingers and the dorsum of the hand (A), and the right forearm (B).

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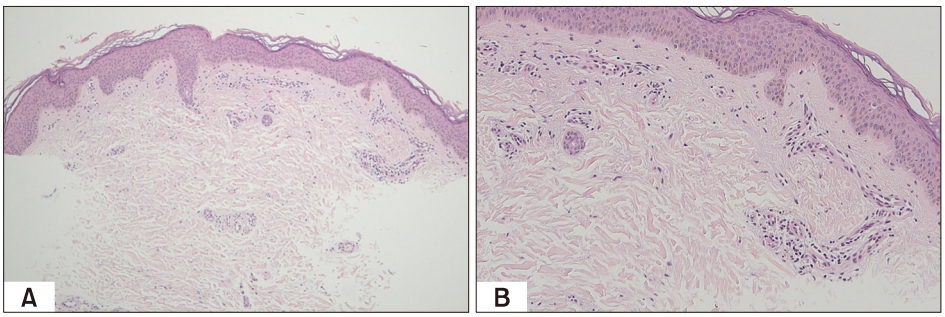

Fig. 2. (A) Histopathologic examination of an erythematous patch on the right wrist revealed multiple ectatic capillaries in the papillary dermis (H&E, ×40). (B) High-magnification view of the papillary dermis (H&E, ×200).

the erythematous patch on the right wrist. A histopathological examination showed multiple ectatic capillaries in the papillary dermis, which is a common finding for PWS (Fig. 2). Based on these, she was diagnosed with trauma-related PWS, also known as Fegeler syndrome, induced by the orthopedic cast, and we recommended pulsed dye laser therapy. However, she did not want to receive treatment, and no change was observed in the lesions over time. We received the patient's consent form about publishing all photographic materials.

Several studies of acquired PWS have been reported; among them, the most frequent cause of acquired PWS is preceding trauma[2,3]. Trauma-related PWS are also called "Fegeler syndrome." The pathogenesis of acquired PWS, resulting from repetitive trauma, has been attributed to a deficit in the perivascular nerve elements. During the post-trauma repair process, an altered neural modulation of sympathetic and parasympathetic systems leads to vascular ectasia that is characteristic of PWS, whereas normal vessels show patent nerve innervation[4]. In addition, Tsuji and Sawabe[5] have shown that impaired vascular contractility during the repair process causes vascular ectasia. In acquired PWS, pulsed dye laser is often the treatment of choice. Acquired PWS is expected to show a better response to laser therapy than is congenital PWS.

Our report suggests that PWS can occur following repeated minor trauma such as the use of an orthopedic cast. In case of acquired PWS following a traumatic event, dermatologists should consider these lesions as "Fegeler syndrome" and should provide explanations for the patient, informing them that if these lesions are not treated, they will persist.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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