

A Case of Hemorrhagic Bullous Morphea

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Dear Editor,

Bullous morphea is a rare form of localized scleroderma (morphea) characterized by bullae on or around an atrophic morphea plaque [2]. The cause of bullae formation in morphea is multifactorial, with lymphatic obstruction from the sclerodermatous process being considered as the most likely cause [3]. Bullous morphea is a rare that represents about 7.5% of all cases of scleroderma [1]. To our knowledge, hemorrhagic bullous morphea have been reported only five cases in the world literatures [1-5]. We here in report a rare case of hemorrhagic bullous morphea.

A 75-year-old female presented with a painful, solitary, 9x6cm-sized, erythematous, sclerotic plaque surrounded by a purpuric bullae on the back for a month (Figure 1). The bullae contained hemorrhagic fluid. The patient refer the lesion had gradually increased in size, and she denied of any trauma history. She had diabete mellitus in her past history. The family history for other autoimmune diseases was negative. The laboratory tests including routine CBC, urine analysis, LFT, RFT and auto antibody tests were within normal limits. We did a biopsy on her back for the diagnosis.

Histopathologic findings of the sclerotic plaque revealed thick collagenous tissue deposition in the superficial dermis, and chronic inflammatory cells infiltration in the dermis. The adnexa appeared reduced in number. (Figure 2). There were no infiltrations of IgG, IgA, IgM, C1q and C3, and minimal focal infiltration of fibrinogen. According to histologic and clinical features, we diagnosed this case as hemorrhagic bullous morphea.

After a month of daily dressing, the bullae spontaneously regressed. There is no recurrence after 6 months.

Peterson et al. proposed that morphea can be classified into five groups: plaque, generalized, bullous, linear, and deep [2]. Among these types, bullous morphea is quite rare, and hemorrhagic bullous morphea are reported only five cases worldwide [1-5].

The pathogenesis of bullous morphea is still unknown, but several mechanisms have been proposed, such as inflammation, lymphangiectases and immune-mediated aggression [1]. The current belief is bullous lesions are more frequently observed on the lower extremities, which suggests that lymphatic



Figure 1: (A) There is a 9x6cm-sized, erythematous, sclerotic plaque surrounded by a purpuric bullae on the back. The center of lesions is pink, firm, sclerotic plaque, and the peripheral lesion is violaceous, soft blisters. (B) The serohemorrhagic fluid in the surrounding tense bullae.

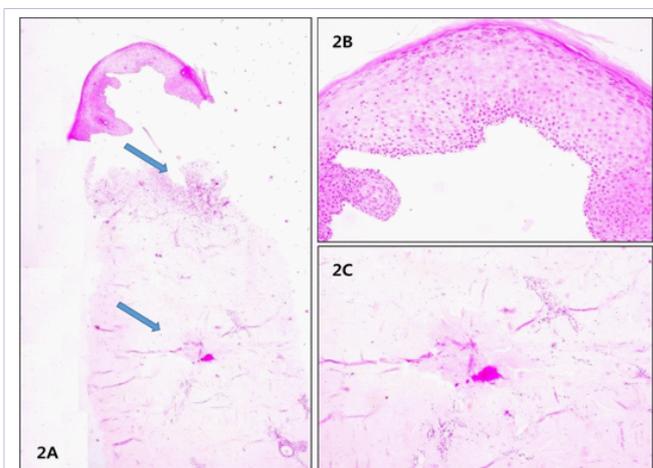


Figure 2: Histopathologic findings of the sclerotic plaque revealed thick collagenous tissue deposition in the superficial dermis, and chronic inflammatory cells infiltration in the dermis. (A: H&Ex40, B, C: H&E x100).

obstruction, combined with increased hydrostatic pressure, leads to bullae formation [1-3]. But Angel reported three cases of bullous morphea that did not show any histopathological feature suggestive of lymphatic blockage, so insisted some signs supporting local trauma as a cause [1]. Pautrier also suggested that vascular changes like arterities and phlebosclerosis play role in bullae formation, so that hemorrhagic bullae can occur in bullous morphea, although rare. Daoud found that major basic protein is responsible for blister formation in at least some cases of morphea [3]. Skin biopsy should be done for diagnosis. Numerous agents, such as corticosteroids, anti malarial drugs, colchicine, retinoids, salazopyrin, pentoxifilline, etc. have been reported to be effective for treating bullous morphea. Despite these large numbers of reported treatments, no consistent recommendations exist for treatment. Prognosis of bullous morphea is rarely life-threatening like other type of morphea, but significant joint deformities are usually common in linear- and

Profunda-type bullous morphea [1]. It maybe self-limited, but has a remitting relapsing or chronic course producing significant disorder burden over time.

As stated above, bullous morphea is very rare disease. And

to our knowledge, hemorrhagic bullous morphea is much less, which have only been previously reported in the literature by Hewitt in 1959, by Antonella in 1990 [5] and by Angel 2015 [1]. This case was described in order to provide a report as a very rare case of hemorrhagic bullous morphea.

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