IMAGES IN HEMATOLOGY

DOI: 10.4274/tjh.galenos.2022.2022.0165 Turk J Hematol 2023;40:64-65

Sickle Cell Leg Ulcer Extending to the Achilles Tendon

Orak Hücreli Anemi İlişkili Bacak Ülseri

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Figure 1. Appearance of leg ulcers from the left ankle. (A) Posterior view of the skin showing a wide tissue defect extending to the Achilles tendon. (B) Perimalleolar wide ulcer from right ankle.



Figure 2. Appearance of the healing ulcer 5 months after initiation of the erythrocyte exchange protocol. (A) Development of granulation tissue covering the tendon and completely filling the deep cavity. (B) Epithelization of the large perimalleolar ulcer with almost complete disappearance.

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Turkish Journal of Hematology, Published by Galenos Publishing House



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Received/Geliş tarihi: April 14, 2022 Accepted/Kabul tarihi: December 1, 2022 A 45-year-old female patient with sickle cell disease [homozygous hemoglobin (Hgb) SS disease] was referred with a 1-year history of a deep leg ulcer located in both perimalleolar regions and at the level of the Achilles tendon in the left foot. Initially, lesions had appeared as a bruise-like macule, which enlarged progressively, became wide, and turned into a destructive wound in the Achilles region, leading to skin and subcutaneous defects to such an extent that the tendon was visible (Figure 1). Steady-state Hgb was measured as 4.7 g/dL, mean corpuscular volume was 137 fL, white blood cell count was 5x10⁹/L, platelet count was 315x10⁹/L, erythrocyte sedimentation rate was 140 mm/h, and folic acid was 2.5 mg/dL. Hgb electrophoresis revealed Hgb S of 70% and Hgb F of 30% (Hgb S 88.8%, Hgb F 7%, Hgb A2 3.2% when not using hydroxyurea). These measurements supported the conclusion that the patient's continuous intake of hydroxyurea was at an adequate dose. Blood chemistry and coagulation test results were unremarkable. All autoantibodies in the autoimmune panel were negative. No abnormal patterns were found in bilateral lower extremity arterial and venous Doppler ultrasonography examinations. Deep anemia and chronic hypoxia contributed to the worsening of the wound.

The patient was scheduled to undergo an exchange protocol at 3-month intervals with target Hgb of 9 g/dL and Hgb S of <30%. While the patient was undergoing transfusion therapy, hydroxyurea was interrupted so as not to impair tissue repair [1,2,3,4]. In addition, local wound care and oral zinc therapy were applied. After three red cell exchange procedures, the giant ulcer began to heal rapidly and a large amount of granulation tissue formed at the tissue defect site (Figure 2). Leg ulcers are not considered among the absolute indications for red cell exchange in the relevant guidelines [4]. This patient is a good example of how effective red cell exchange procedures may be for sickle cell wound healing.

Keywords: Sickle cell disease, Leg ulcer, Red cell exchange

Anahtar Sözcükler: Orak hücre hastalığı, Bacak ülseri, Kırmızı hücre değişimi

Ethics

Informed Consent: Written and verbal informed consent was obtained from the patient for publication of this paper.

Authorship Contributions

Surgical and Medical Practices: S.A., M.K., B.B.; Concept: S.A., M.K.; Design: S.A. B.B; Data Collection or Processing: S.A.; Analysis or Interpretation: S.A., B.B.; Literature Search: S.A., B.B.; Writing: B.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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