Ulcers in congenital anemia

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Introduction

Survival rates in patients with chronic forms of anemia such as sickle cell disease and thalassemia have improved, but the management of comorbidities, such as leg ulcers, remains a challenge. Recalcitrant leg ulcers are a common complication of such conditions and may occur early in life. The incidence of leg ulcers in adults with sickle cell disease is reported to be approximately 30%; ulcers in these patients can last for very long time.¹² The pathogenesis of ulcerations in congenital anemia is multifactorial: hypoxia due to anemia, increase of blood viscosity because of bone marrow stimulation, changes in the coagulation and fibrinolytic system, rheological effects, iron overload, venous stasis causing red blood cell diapedesis, followed by extravascular hemoglobin catabolism. The higher intravenous pressure in the upright position acts as a predisposing factor even in the absence of a venous pathologic factors, by general and local perturbances in the lower extremities.

The common denominator in these ten cases was high venous pressure due to gravity. High intravenous pressure caused by sitting and standing without movement is sufficient to trigger chronic inflammation, leading to skin defects on the legs of patients with additional hematological risk factors associated with several microcirculatory abnormalities, as discussed above. Trauma may play an additional role by stimulating the damage of red blood cells. The prolonged average healing time of 7 months in our ten patients points is due to the chronic, recalcitrant nature of the ulcers, which had persisted for several years before compression treatment was initiated. Only compression treatment with short-stretch bandages and walking exercises made the ulcers heal. The therapeutic success of good compression stockings was prescribed to prevent recurrences.

Case Series

Ten patients affected by congenital dyserythropoietic anemia, sickle cell anemia and Thalassemia intermedia with chronic leg ulcers persisting between 12 and 29 years (mean 16 years) without venous out flow disturbances in the lower extremities. Compression therapy was able to heal the ulcers in 5 to 8 months (average 7 months) after several attempts of purely local treatment had failed. Once the ulcers had healed, compression stockings were prescribed to prevent recurrences.

The common denominator in these ten cases was high venous pressure due to gravity. High intravenous pressure caused by sitting and standing without movement is sufficient to trigger chronic inflammation, leading to skin defects on the legs of patients with additional hematological risk factors associated with several microcirculatory abnormalities, as discussed above. Trauma may play an additional role by stimulating the damage of red blood cells. The prolonged average healing time of 7 months in our ten patients points is due to the chronic, recalcitrant nature of the ulcers, which had persisted for several years before compression treatment was initiated. Only compression treatment with short-stretch bandages and walking exercises made the ulcers heal. The therapeutic success of good compression therapy in our cases shows that this treatment is able not only to act against venous reflux but also to counteract gravity in cases with normal valve function. Compression not only reduces edema, it also releases vasoactive enzymes from the endothelial cells, improving microcirculation, reducing inflammation, and promoting lymph drainage. This is especially true for non-yielding, inelastic bandages, which exert a considerable massage effect during walking.¹²⁻¹⁷ These mechanisms may also explain the beneficial effects of compression treatment in patients with other non-venous lesions on the leg, such as vasculitis, cellulitis, and lesions after trauma or surgery.

In conclusion, compression therapy was successful in the treatment of the reported cases and should be considered as a component of treatment strategies for patients with hematological ulcers. In our cases, this simple, inexpensive approach was the winning strategy for resolving a longstanding problem.

References

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[Veins and Lymphatics 2014; 3:5985]