

Leg ulcers and Klinefelter syndrome

Siham Belmourida, Hind Palamino, Mariame Meziane, Nadia Ismaili, Laila Benzekri, Badredine Hassam, Karima Senouci

Department of Dermatology-Venerology, Mohammed V University, Ibn Sina Hospital, Rabat, Morocco

Corresponding author: Dr. Siham Belmourida, E-mail: belmouridasiham2017@gmail.com

Sir,

Klinefelter syndrome (KS) is a rare sex chromosome disorder [1], associated with leg ulcers in 6% of cases. These ulcers are usually recurrent, refractory to treatment, and are a major socioeconomic handicap [2].

We report the case of a patient with a chronic leg ulcer associated with KS.

A 45-year-old patient, who underwent ligation of perforating veins of the left lower limb in 2005, was followed with endocrinology for KS for three years and put on androgens. He consulted with the dermatologist for a leg ulcer.

An examination found an elongated patient with large limbs (Fig. 1) as well as undeveloped sexual characteristics.

A dermatological examination revealed internal retromalleolar ulcers with plaques of ocher dermatitis associated with varicose veins of the lower limbs (Figs. 2 and 3).

A biological examination revealed hyper gonadotropic hypogonadism and osteoporosis. A thrombophilic and an immunological checkup were negative. Venous ultrasonography of the lower limbs showed staged venous thrombosis, which contraindicated any surgical procedure in view of the extent of thrombosis.

Treatment with platelet antiaggregant was initiated with fatty dressings and compression stockings in addition to the androgen treatment.

KS is the most common congenital anomaly causing primary hypogonadism [1,3], occurring in 1/500 to 1/1000 live births. The prevalence of leg ulcers in people with KS is 6%, 30 times higher than in the



Figure 1: Long upper limbs in relation to Klinefelter syndrome.



Figure 2: Ulcers of the lower limbs developed on a site of chronic venous insufficiency.

How to cite this article: Belmourida S, Palamino H, Meziane M, Ismaili N, Benzekri L, Hassam B, Senouci K. Leg ulcers and Klinefelter syndrome. Our Dermatol Online. 2021;12(3):345-346.

Submission: 17.06.2020; **Acceptance:** 13.09.2020

DOI: 10.7241/ourd.20213.32



Figure 3: Perimalleolar ulcers of the lower limbs with associated ocher dermatitis.

general population. The pathogenesis of KS-associated leg ulcers is complicated due to a combination of factors: venous incompetence caused by an increase in venous pressure associated with long lower limbs, hormonal abnormality (inhibition of fibrinolysis caused by low levels of testosterone) [1,3], and abnormality of coagulation and the fibrinolytic system, which promotes thrombosis [3]. All these hemostatic abnormalities may explain ulcers by thrombosis of cutaneous microvessels and/or by venous thrombosis inducing postphlebotic disease, as in our patient. These leg ulcers are often refractory to treatment, require multidisciplinary therapeutic management, and involve the use of androgenic treatment, because improvement

of leg ulcers in people with hypogonadism has been observed when on hormone therapy [2,3].

One must always keep in mind KS in young patients with recurrent venous ulcers refractory to treatment with a notion of sterility associated with the hypogonadism chart.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients have given consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCES

1. Jouin R, Chateaux D, Drapier D, Pham Hoang T. [Towards a rapprochement of Klinefelter syndrome and psychopathy in forensic psychiatry]. *Ann Med Psychol*. 2019;177:902-10.
2. Shanmugam VK, Tsagaris KC, Attinger CE. Leg ulcer associated with Klinefelter's syndrome: a case report and review of literature. *Int Wound J*. 2012;9:10-47.
3. Ines L, Youssef M, Marmouch H, Mohamed H, Klii R, Akkari H, et al. Ulcère de jambe révélant un syndrome de Klinefelter. *Rev Méd Int*. 2015;36S:A76-85.

Copyright by Siham Belmourida, et al. This is an open access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Source of Support: Nil, Conflict of Interest: None declared.