Acute and dramatic saxophone penis

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ABSTRACT

We present a case of intense genital swelling because of a hereditary angioedema. This rare disease should be included in the differential diagnosis of acute and asymptomatic genital edema, because it may prevent future potentially life-threatening episodes of visceral angioedema.

Key words: Angioedema, C1 inhibitor, genital swelling, hereditary

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We present the case of a 33-year-old white Spanish man with two days history of an asymptomatic fast-growing edema, first on both ankles and later on his penis. He denied any rash, urethral symptoms, or other systemic symptoms. The edema decreased on ankles after 24 h but not on the penis, where it began over the distal foreskin, progressing to the entire shaft and scrotum. was no inguinal lymphadenopathy. Due to extensive edema, the glans could not be visualised and the opening of the urethral meatus was blocked [Figure 1]. There was no history of unprotected sexual intercourse, trauma, insect bites, or previous similar episodes. However, he revealed that his maternal grandfather had recurrent limb edema, therefore, he and his mother were studied and diagnosed with a quantitative C1 inhibitor deficiency a year ago. We treated him with Berinert 500 U intravenous and after 5 h, the edema disappeared and no cutaneous pathology was observed on the glans.

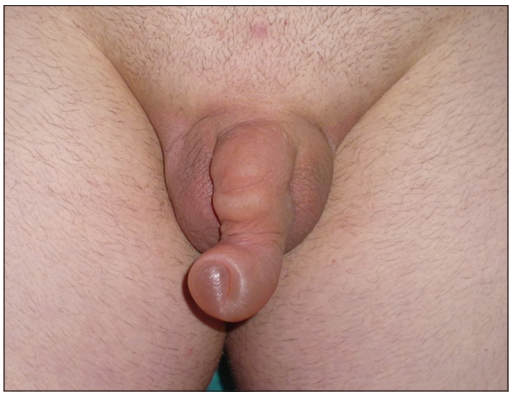

Figure 1: Intense edema of the penis shaft and scrotum with no inguinal lymphadenopathy, erythema or skin lesions

Saxophone penis refers to swelling and deformity of the penile shaft secondary to multiple causes. In case of acute male genital edema, it is essential to distinguish if the swelling is painful, which could be a potential emergency,[1] or if it is asymptomatic where hereditary angioedema (HAE) could be the rare cause. Angioedema may be hereditary, accounting for 5% of cases, or acquired by causes such as allergy, angiotensin-converting enzyme inhibitors, malignancies, autoimmunity, and connective tissue diseases. HAE is caused by a defect in C1 inhibitor, usually a quantitative deficiency, only 20% of it is because of a functionally impaired C1 inhibitor. It has an autosomal dominant inheritance. Although there is often a family history, a quarter of individuals may have a de novo gene mutation.[2] The first presentation of HAE most commonly affects the skin, usually edema of the extremities, face, or genitals. Patients can subsequently develop episodes of visceral edema –gastrointestinal and respiratory systems are the most frequently involved.

HAE is diagnosed by measuring the levels and functionality of serum C1 inhibitor; affected individuals also have low C4 levels. Acute episodes should be treated with intravenous purified C1 inhibitor concentrate or a bradykinin-2 receptor antagonist. For long-term prophylaxis, attenuated androgens or antifibrinolytic agents could be used.[3]

Including HAE in the differential diagnosis of genital swelling may prevent future potentially life-threatening episodes of visceral angioedema.

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