

Saxophone penis associated with penoscrotal lymphangiomas

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ABSTRACT

Lymphangiomas are a condition marked by the presence of cysts that result from an increase both in the size and number of thin-walled lymphatic channels that are abnormally interconnected and dilated. It can affect skin and other organs in the body. Genital lymphangiomas may arise from primary or secondary lymphatic abnormalities. It affects both sexes, and frequently misdiagnosed as genital warts. In this concise report, we present an adult male with diffuse penoscrotal lymphangiomas associated with a peculiar penile deformity, known as “Saxophone penis”.

Key words: Genital swelling; Lymphangiomas; Sexually Transmitted Diseases (STDs); Warts

INTRODUCTION

Presence of multiple lymphangiomas in the genital area, otherwise known as genital lymphangiomas, can be primary (due to lymphatic malformation) or secondary to disorders that block or interfere with the drainage of lymph in this area [1-13].

The primary genital lymphangiomas do not usually present at birth (congenital), and often, present later in life.

Here, we present an adult male with diffuse penoscrotal lymphangiomas associated with a peculiar penile deformity, known as “Saxophone penis”.

CASE REPORT

A 60-year-old Afghani male patient, not known to have any chronic medical problems before, who presented

with persistent and progressive, multiple papules on the penis and scrotum for more than 15 years duration. There is mild itching but no pain.

The penis is swollen and deformed (Fig. 1a), giving rise to the so-called ‘ramrod’, “ram horn penis,” or ‘saxophone penis’. Simulated to the saxophone, which is a widely-known musical instrument (Fig. 1b).

He denied any urethral symptoms, genital ulcers, or other systemic symptoms. There was no history of unprotected sexual intercourse, trauma, operations or anything to suggest a disorder of secondary genital lymphedema.

He is diagnosed several times before as genital warts.

All routine tests including Mantoux test, chest X-ray, ultrasound of abdomen, serum Venereal Disease

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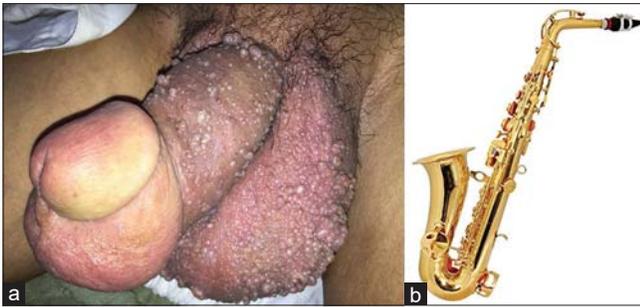


Figure 1: (a) Saxophone penis associated with penoscrotal lymphangiomatosis. (b) The saxophone.

Research Laboratory (VDRL) test were within normal limits. The patient was seronegative for HIV-1 and 2.

Punch skin biopsy confirmed the diagnosis of lymphangiomatosis and the patient is referred for urologist for further evaluation.

DISCUSSION

Cutaneous genital lymphangiomatosis is not a Common condition [1-13].

Lymphangiomatosis of primary cause, like other lymphatic malformations, is thought to be the result of congenital errors of lymphatic development occurring prior to the 20th week of gestation.

The lymphangiomatosis and penile deformity in our case is most likely due to primary isolated lymphadema, which is consequence of genital lymphatic malformation.

Disorders which may precipitate secondary genital lymphedema include filariasis and sexually transmitted diseases (STDs) like lymphogranuloma venereum (LGV), surgical removal of inguinal lymph nodes [7].

Although lymphatic malformations and lymphangiomatosis are technically benign, these deranged lymphatics tend to invade surrounding tissues and cause problems due to invasion and/or compression of adjacent structures. As seen in our case, it precipitate anatomical deformity.

Scrotal lymphatic malformation with absent corpora cavernosa has been also reported [13].

“Saxophone penis” refers to swelling and deformity of the penile shaft secondary to multiple causes. It is a term used when the penis gets twisted along its long axis, giving it the appearance resembling a saxophone.

The latter, referred to colloquially as the sax, is a widely-known musical instrument. It is type of single-reed woodwind instrument with a conical body, usually made of brass.

The cause for this shape is not well understood; however, it is likely that the peculiar penile anatomy makes any fluid or vascular overload in the soft tissue of the penis to render the penis to this shape [6].

The different conditions causing a saxophone penis, reported in the literature, are primary lymphedema [4], penile tuberculosis [5], lymphogranuloma venereum in its tertiary stage [7], topical imiquimod use [8], entomophthoromycosis (basidiobolomycosis) [9], paraffin injections [12], and hereditary angioedema (HAE) [10].

In case of acute male genital edema, it is essential to distinguish if the swelling is painful, which could be a potential emergency, or if it is asymptomatic where HAE could be the rare cause [10].

It is difficult to achieve a complete cure for lymphatic malformations with different therapeutic modalities, including surgery, and its management remain to be a challenge.

Physicians in general and dermatologists, venereologists and STDs specialists in particular, need to keep lymphangiomatosis in their differentials for genital lesions in order not to misdiagnose it as warts [1].

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 gave their 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.

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