Syphilis, the “great imitator,” presents with a wide range of mucocutaneous and systemic findings. The primary chancre classically occurs in the genital region, however up to 6.33% can be extragenital. Among the extragenital chancres reported in the literature, very few occurred on the breast, and of these cases only 5% occurred in men. A 43-year-old healthy man visited our clinic complaining of drainage from the right nipple for one month. Exam was notable for a poorly defined, scaly erythematous plaque on the areola with a superficial erosion of the nipple. The rapid plasma reagin (RPR) titer was found to be 1:32, with positive anti-treponemal antibodies. Histopathological examination of the biopsy specimen revealed variable acanthosis of epidermis with dense underlying superficial, deep perivascular and interstitial infiltrate surrounding benign bundles of smooth muscle. The infiltrate was composed of abundant lymphocytes, plasma cells, and rare eosinophils. Immunohistochemical staining for Treponema pallidum revealed numerous spirochetes scattered within the inflammatory cell infiltrate throughout the dermis. Based on these findings, the patient was diagnosed with an extragenital chancre of primary syphilis on the nipple. With a resurgence in the incidence of syphilis, it is important to remind practitioners of the more unusual presentations of this disease.

Introduction

Syphilis is known for its ability to mimic innumerable conditions. This condition is often misdiagnosed due to the wide variety and transient nature of cutaneous, mucocutaneous, and systemic manifestations which vary greatly depending on the stage of presentation. The World Health Organization estimates that each year there are over 11 million new cases of syphilis. With the incidence continuing to rise in the United States at an alarming rate, the US Preventive Task Force updated screening recommendations in 2016 to include asymptomatic, nonpregnant adults and adolescents at increased risk for infection.1,2,3

Primary syphilis refers to inoculation with T. pallidum and manifests as a localized cutaneous chancre at the site of contact, classically occurring in the genital region. However up to 6.33% of primary chancres can be extragenital. Among the extragenital chancres studied, 5.1% occurred on the breast and only 5% of those on the breast occurring in men.4,5

We present one of the first reported cases of a male with primary syphilis on the nipple in the United States.

Abstract

Case Description

• Past medical history: Gout
• Family medical history: Breast cancer (mother)
• Physical exam: Erythematous, ulcerated, plaque with serosanguinous drainage and crusting at the 12 o’clock position of nipple. Tenderness with palpation noted. No palpable axillary or supraclavicular lymphadenopathy noted. No penile ulceration was found.
• Differential diagnosis: Nipple eczema, erosive adenomatosis of the nipple, mammary Paget disease or primary breast carcinoma.
• Pathology: Punch biopsy demonstrated hyperkeratosis of stratum corneum. The epidermis is variably acanthotic with mild spongiosis focal exocytosis of few lymphocytes within spongiosis. A dense superficial & deep perivascular and interstitial infiltrate surrounding benign bundles of smooth muscle consisting of lymphocytes, plasma cells, and rare eosinophils (Figure 2) was also noted.
• Staining: Modified Steiner and immunohistochemical staining for T. pallidum revealed spirochetes (Figure 3). Periodic Acid-Schiff with diastase (PAS-D) negative for fungal elements. Acid cytokeatin (CAM5.2) negative.

Discussion

• According to the CDC, there has been a dramatic increase in the incidence of primary and secondary syphilis in the U.S.
• In 2016, a total of 27,814 cases reported 8.7 cases per 100,000 population spanning equally across all regions of the country.
• An increase of 17.6% compared to 2015
• An increase of 74.0% compared to 2012

Initially, increase in incidence was associated with men who have sex with men. However, most recent data reveals an increased incidence in women as well.7

Extragenital chancres can occur at any site of inoculation including oral (lips, tongue, palate), perianal, breasts/nipple, conjunctiva, neck, abdomen, intrascapular region, arms, palms, fingers or thighs.5,6

Extragenital primary syphilitic chancre of the nipple is an exceedingly rare manifestation of primary syphilis.

Historically, primary syphilitic chancre of the nipple was associated with wet nurses resulting from mucous patches in the mouths of congenitally syphilitic infants.4 Fewer than 10 cases have been reported in modern literature. Most cases are associated with sexual encounter involving oral contact, especially biting, of the nipple.8,9,10

Conclusion

• Patients who present to a dermatologist with syphilis are more likely to demonstrate unusual or advanced forms of this condition.11

Practitioners must be aware of unusual presentations and maintain a high index of suspicion of syphilis in order to make this diagnosis.

• Early recognition and a low threshold for testing is important to limit disease spread.12

References