



A Rare Case of Verruciform Xanthoma of Vulva

Mrityunjay METGUD¹, Prakash MALUR², Subash MALLYA³

¹Department of Obstetrics & Gynecology, Jawaharlal Nehru Medical College, Belgaum, Karnataka, India

²Department of Pathology, Jawaharlal Nehru Medical College, Belgaum, Karnataka, India

³Department of Obstetrics & Gynecology, Jawaharlal Nehru Medical College, Belgaum, Karnataka, India

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Abstract

Verruciform xanthoma of the vulva is an extremely rare entity, the etiology of which is unknown. The diagnosis is almost always made on histopathological examination. There have been only 7 cases of verruciform xanthoma reported worldwide to date. A lady aged 30 years with 2 children presented with the complaints of copious white discharge per vagina and vulval swelling of 5 year duration. She had received antibiotics and antiviral drugs for the same. On examination three plaque like lesions were seen on both the labia minora each measuring 2x1 cm. Vaginal smear examination, vaginal Swab culture, TPHA and VDRL, HIV, HSV, HPV and Mantoux were negative. Endometrial tissue did not show any growth on culture and was negative for tuberculosis by PCR. A diagnosis of verruciform xanthoma of vulva was made on histopathological examination of the biopsied tissue from the vulval swelling by the presence of foam cells. There was no evidence of tuberculosis, fungal and parasitic infection or malignancy. The lady was advised to undergo surgery (complete excision of the vulval lesions) for the vulval xanthoma. However, she did not follow-up in spite of all communication efforts. Verruciform xanthoma of the vulva is an extremely rare entity which cannot be diagnosed clinically. It needs to be differentiated from other rare vulval lesions resembling it. Hence, all such patients need to be approached meticulously in a centre having the necessary diagnostic modalities and expertise in managing such rare cases.

Keywords: verruciform xanthoma, vulval xanthoma, foam cells

Özet

Nadir Bir Olgu Olarak Vulvanın Verrüköz Ksantoması

Vulvanın verrüköz ksantoması, etiyolojisi tam olarak bilinmeyen oldukça nadir görülen bir durumdur. Teşhis, hemen hemen her zaman histopatolojik tanıya dayanmaktadır. Literatürde dünyada sadece 7 olguda verrüköz ksantoma bildirilmiştir. Sunulan olgu (30 yaşında 2 çocuk annesi kadın) yoğun beyaz akıntı ve yaklaşık olarak 5 yıldır süregelen vulvar şişlikler nedeniyle kliniğimize başvurdu. Anamnezinde, bu nedenle antibiyotik ve antiviral tedavi aldığı ifade eden hastanın muayenesinde her iki labia minorada 3 adet 2x1 cm boyutlarında plak benzeri lezyon görülmekteydi. Vajinal smear, kültür, TPHA, VDRL, HIV, HSV, HPV ve Mantoux testleri negatif idi. Endometriyal dokudan alınan örneğin kültürde üremesi olmadı ve PCR sonucu tüberküloz açısından negatif geldi. Vulvar verrüköz ksantoma tanısı ancak histopatolojik olarak köpüksü hücrelerin görülmesiyle konuldu. Aldığımız örnekte tüberküloz, mantar ve parazitik enfeksiyon ya da malignite bulgusuna rastlanmadı. Hastaya cerrahi eksizyon önerildi. Ancak hasta tüm iletişim çabalarına karşın takibi bıraktı. Verrüköz ksantom, vulvada oldukça nadir görülen ve klinik olarak tanısı zor konulabilen bir durumdur. Lezyonun diğer vulvar lezyonlardan ayrımının yapılması gerekmektedir. Dolayısı ile bu hastaların teşhis ve tedavinin doğru olarak yapılabileceği merkezlerde takibi gerekmektedir.

Anahtar sözcükler: verrüköz ksantom, vulvar ksantom, köpük hücreleri

Corresponding Author: Dr. Mrityunjay Metgud
48/49 Shivakrupa, Near Mahaveer Bhavan,
Hindwadi 590011 Belgaum, India
Phone : +91 831 246 67 46
GSM : +91 944 852 76 67
E-mail : metm@rediffmail.com

Introduction

Verruciform xanthomas are rare, benign lesions predominantly reported in oral cavity but are also known to occur over the genital skin and mucosa of the vulva (1), perianal area (2), the penis (3) and the scrotum (4). The etiology of verruciform xanthoma is not clearly known. Majority of the cases are reported in adults and whites. They usually present as asymptomatic solitary flat plaque or warty lesions up to 2 cm in diameter. The clinical appearance of a verruciform xanthoma is not diagnostic; the diagnosis is almost always made on histological examination. The most striking and characteristic histological feature is the presence of large foam cells in the connective tissue. Verruciform xanthoma of the vulva is an extremely rare entity and there have been only 7 cases reported worldwide to date (5). Here we report one such rare case of verruciform xanthoma of vulva.

Case Report

A lady aged 30 years, with 2 children presented to us with the complaints of copious white discharge per vagina and vulval swellings of 5 year duration. She had received antibiotics and antiviral drugs for the same complaints in the past, with no relief. The swelling over the vulva was associated with pain and itching which increased during and just after her menstrual cycles. She also complained of excessive menstrual flow with severe dysmenorrhea for 3 years. Her built was average and had a normal blood pressure. On examination three plaque like lesions were seen on both the labia minora each measuring 2x1 cm (Figure 1). There was marked tenderness over the vulval swelling and also in the vagina with copious, non foul smelling discharge.

Vaginal smear examination revealed numerous pus cells and was negative for trichomoniasis and candidiasis. Vaginal Swab culture did not grow any organisms. Treponema Pallidum Haemagglutination (TPHA) test, and Venereal Disease Research Laboratory slide screening (VDRL) for syphilis were negative. ELISA for Human Immunodeficiency Virus (HIV) was non-reactive. Serological tests for Herpes Simplex Virus (HSV -IgM antibody) and Human Papilloma Virus (HPV) were negative. Mantoux test for tuberculosis was negative. Endometrial tissue did not show any growth on culture and was negative for tuberculosis by polymerase chain reaction (PCR).

Histopathological examination of the biopsied tissue from the vulval swelling showed hyperkeratosis with parakeratosis, elongation of rete ridges along with acanthotic spinous cell layer (Figure 2). The dermis was diffusely infiltrated by large foam cells. Numerous Touton's giant cells were present throughout the dermis (Figure 3). There was no evidence of tuberculosis, fungal and parasitic infection or malignancy. A diagnosis of verruciform xanthoma of vulva was made.



Figure 1. Multiple plaque like lesions over the vulva at the time of presentation.

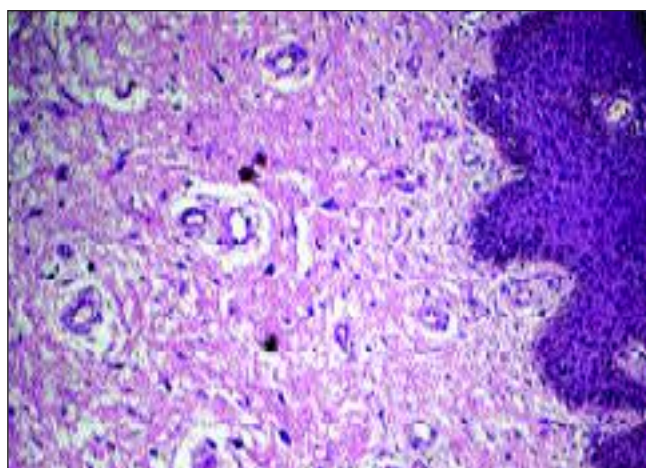


Figure 2. Hematoxilin and eosin stain (x10). Hyperkeratosis with parakeratosis, elongation of rete ridges along with acanthotic spinous cell layer. The dermis is diffusely infiltrated by large foam cells.

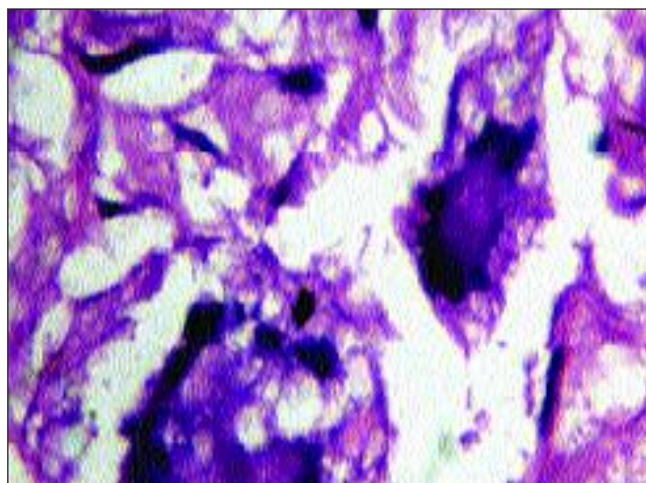


Figure 3. Hematoxilin and eosin stain (x40). Fat-laden foam cells in the connective tissue of the dermis.

The lady was given 2 weeks course of oral antibiotics and local vaginal pessaries for her recurrent vaginal discharge. However, she did not respond to the treatment and later was put on anti-tuberculosis treatment empirically for the same problem. She responded to the treatment after a month and was advised to continue the same for another 5 months. The lady was also advised to undergo surgery (complete excision of the vulval lesions) for the vulval xanthoma. However she was lost for follow-up in spite of all communication efforts.

Discussion

Verruciform xanthoma of the vulva is an extremely rare entity which cannot be diagnosed clinically. It needs to be differentiated from other vulval lesions resembling it. Hence all patients presenting with labial warty growths should have to undergo the investigations including TPHA, HIV, HSV, HPV; Mantoux test for tuberculosis; blood lipid profile and excision biopsy of the lesion.

The etiology of xanthomas is not clearly known. Trauma and inflammation are the two best established trigger factors. As trigger factors like inflammation could be due some other pathology leading to recurrent vaginal discharge, a vaginal smear examination to rule out vaginal candidiasis and trichomoniasis, endometrial tissue culture for organisms and endometrial tissue PCR for tuberculosis would be necessary as was required in our case. Verruciform xanthoma has been reported to occur in association with carcinoma *in situ* or with squamous cell carcinoma (6). Even though it is a very rare phenomenon a meticulous approach is needed for the exact diagnosis and prompt treatment as it has been known

to have malignant transformation. Verruciform xanthoma is unresponsive to steroids, immunosuppressive agents or radiation, and therefore require complete surgical excision.

Tendency to recur following complete excision is very little (7). When a complete excision of the lesion is not done but only a part of it is biopsied, it has to be deep enough, as verruciform xanthoma can be misdiagnosed for verrucous carcinoma if a superficial biopsy is performed. Verrucous carcinoma is of particular importance because, although it is a carcinoma, it shows little or no epidermal cytological atypia. So, the performing gynecologist must always perform a good deep biopsy if a complete excision is not being done. Rarity and complexity of the lesions mandate these patients to be managed in a centre that is familiar with various diagnostic modalities to ensure the best curative treatment option.

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