

CASE REPORT

An enlarging keratotic verrucous papule on the vulva

Lamiaa Hamie, MD, Mazen Kurban, MD, Ossama Abbas, MD, Nelly Rubeiz, MD

Department of Dermatology, American University of Beirut Medical Center, Beirut, Lebanon

ABSTRACT

Trichilemmoma as a benign neoplasm with differentiation toward pilosebaceous follicular epithelium, or outer root sheath. Although benign in nature, its importance lies in its association with Cowden syndrome. In which it is found to be associated with multiple types of tumors. Also, it needs to be differentiated from other more aggressive cutaneous tumors, such as trichilemmal carcinoma. Clinically, they present as well-defined, smooth, asymptomatic papules or verrucous papules. Which may occur either as a solitary lesion or in some cases as multiple lesions. They are commonly located in the head and neck region. We are herewith presenting a case of solitary trichilemmoma occurring in a rather unusual location.

KEYWORDS: Trichilemmoma, adnexal tumor, vulva

INTRODUCTION

Headington and French in 1962, first described trichilemmoma as a benign neoplasm with differentiation toward pilosebaceous follicle. Although it is benign in nature, its significance lies in its association with Cowden syndrome. In which, it is found to be associated with multiple other types of tumors. Also, it needs to be differentiated from other more aggressive cutaneous tumors, such as trichilemmal carcinoma. Clinically, it may present as a solitary or multiple skin-colored papules distributed typically over the central face. Desmoplastic variant of trichilemmoma is known to develop in the genital area and can also present as a secondary neoplasm in nevus sebaceous. The clinical differentials of trichilemmomas includes verruca vulgaris and other hair follicle or epidermal tumors. The diagnosis can only be

confirmed by histopathological examination. Herein, we are presenting a case of biopsy confirmed solitary trichilemmoma occurring in a rather unusual location.

CASE REPORT

A 72-year-old woman presented to the dermatology clinic with a verrucous papule on the right vulva of 2 weeks duration. She was known to have lichen sclerosus of the perineum, controlled with topical clobetasol and barrier creams. Medical history was pertinent for diabetes, hypertension, and dyslipidemia. Physical examination revealed a 4 mm keratotic verrucous papule over the right vulvar area (Fig. 1). No other lesions were identified. Biopsy specimen revealed a folliculo-centric lobular tumor composed of monomorphous

Correspondence: Dr. Ossama Abbas, Professor, Department of Dermatology, American University of Beirut Medical Center, Riad El Solh/ Beirut 1107 2020, Beirut, Lebanon, P.O.Box 11-0236, Email : ossamaabbas2003@yahoo.com, Tel.: +961-1-350000, ext. 7915, Fax : +961-1-745320

epithelial cells with clear cytoplasm. Peripheral palisading of compact basal keratinocytes and a PAS-positive thickened basement membrane were noted. The overlying epidermis was hyperkeratotic. No signs of dysplasia were present (Fig. 2). These findings were consistent with the diagnosis of trichilemmoma.



Fig. 1 Clinical photograph of keratotic verrucous papule on right vulva.

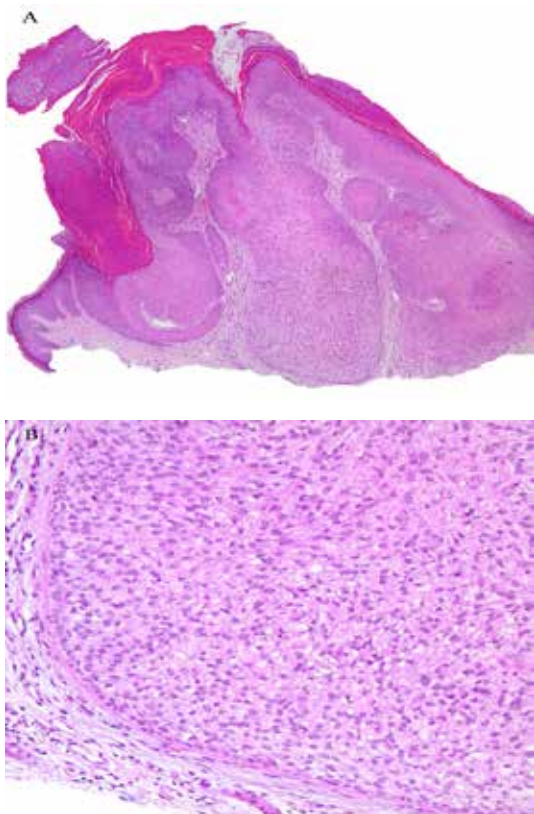


Fig. 2 Histopathology showed folliculo-centric lobular tumor composed of monomorphous clear epithelial cells, peripheral palisading, and thickened basement membrane (hematoxylin-eosin x4, x40).

DISCUSSION

Trichilemmoma is a benign adnexal neoplasm with differentiation towards the follicular outer root sheath.¹ It may present as a solitary or multiple skin-colored papules distributed typically over the central face (eyelids, nose and upper lip) but can uncommonly affect other areas such as the genital skin. Multiple trichilemmomas are highly suggestive of Cowden syndrome; a genetic disorder characterized by the loss of PTEN expression.¹ Patients with Cowden syndrome can develop sclerotic fibromas, acral keratoses and adenocarcinoma of the breast, thyroid and gastrointestinal tract.² Our patient had no other cutaneous features of Cowden syndrome.

The microscopic examination of trichilemmoma usually displays a lobulated proliferation composed of central pale, periodic acid Schiff (PAS) positive, diastase sensitive, glycogen containing cells and peripheral basophilic palisading cells enclosed within a eosinophilic PAS positive basement membrane.³ These cells can be CD34 positive in line with their differentiation towards the outer root sheath. Clinically, the differential diagnosis of trichilemmomas includes verruca vulgaris and other hair follicle or epidermal tumors.¹ Microscopically, trichilemmoma needs to be differentiated from intraepidermal clear cell neoplasms such as clear cell poroma (sharp contrast from adjacent epidermis, absence peripheral palisading, absent hyaline peripheral membrane, ductal differentiation, may have dermal extension); clear cell acanthoma (lower limb localization, psoriasiform scale, well circumscribed epidermal hyperplasia with pale large keratinocytes); and clear cell squamous cell carcinoma in situ (atypia of keratinocytes in

all epidermal layers).¹

In addition, trichilemmomas have a desmoplastic variant that can develop on the genital area and may occur as a secondary neoplasm in nevus sebaceous.² Desmoplastic trichilemmoma is characterized by clusters of the outer sheath cells surrounded by a sclerotic stroma, hence, it can be confused with carcinomas such as squamous cell carcinoma, sclerosing basal cell carcinoma, and trichilemmal carcinoma. However, these lack the distinctive histological features of trichilemmoma.¹

The relationship between trichilemmoma and verruca vulgaris is unclear. Both entities can affect similar areas and share clinical characteristics such as a verrucous or hyperkeratotic surface. Some authors postulate that trichilemmomas are “burnt-out verruca” with trichilemmal differentiation rather than primary cutaneous hamartomas.⁴ However, others oppose the former postulation due to the distinguishing peripheral palisading, thick eosinophilic surrounding membrane, sclerotic stroma between epithelial lobules among other reasons.⁵ Also, studies using in situ hybridization with low- and high-risk human papillomavirus (HPV) probes failed to detect HPV in trichilemmomas.⁴ In one study which analyzed 29 papules suspected of being trichilemmomas in patients with Cowden

syndrome;⁶ only 2 were trichilemmomas, one of which was positive for HPV 80.⁶ Thus, more studies are required to establish a definitive association.

The treatment of trichilemmoma is usually optional and ablative in nature. Multiple therapeutic options can be performed, such as cryotherapy, curettage, shave or surgical excision.² In conclusion, trichilemmoma should be kept in the differential diagnosis of verrucous papules on the genital area and Cowden syndrome should be considered especially in the context of multiple lesions.

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