SKIN

BRIEF ARTICLE

Perianal Warty Dyskeratomas: Case Report of an Unusual Presentation

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ABSTRACT

Warty dyskeratoma are benign adnexal neoplasms that typically present as isolated lesions on the head and neck. They have distinctive features microscopically, including a cup-shaped invagination with suprabasiliar acantholytic dyskeratosis, corp ronds and grains, and central keratotic plug. A patient presented with multiple, small perianal papules that were initially diagnosed as anal glands, then genital warts. When a biopsy was performed, the classic features of a warty dyskeratoma were seen. The patient was treated with topical calcipotriene which was helpful in reducing the size of the lesions. This case raises the possibility that perianal warty dyskeratomas may be more common than previously thought, which has significant implications for treatment and counseling of patients.

INTRODUCTION

Warty dyskeratoma (WD) is a benign, self-limiting epidermal neoplasm that most commonly presents as a well-circumscribed, singular, umbilicated papule with a keratotic plug.[1] WD most often manifests on the scalp, face or neck of middle aged or elderly individuals. Rare cases of multiple lesions and involvement of oral mucosa and genitalia have been reported.[2] The pathophysiology of WD is largely unknown, though proposed etiologies include viral infection, cigarette use, or autoimmune processes that lead to acantholytic dyskeratosis of a pilosebaceous unit.[1]

Distinguishing WD from other types of focal acantholytic dyskeratosis (FAD) can be a challenge and requires clinicopathologic correlation. WD occur in three main histologic patterns: the predominant cup-shaped formation, cystic, and nodular formations.[3] Other clinical presentations of WD include pruritic papules, brownish verrucous nodules, and grouped verrucous papules on the scalp.[4,5] While clinically varied, a characteristic pattern on histopathology confirms the diagnosis of WD: cup-shaped epidermal invaginations filled with keratin debris, corps ronds and grains, and villi formation with suprabasal acantholysis.[6]

CASE REPORT

A 28-year-old woman had a 3-year history of asymptomatic, small growths in the perianal region that had been slowly increasing in number. She initially presented to a women's health clinic and was referred to gastroenterology for further workup. The gastroenterologist reassured the patient that they were normal anal glands and no treatment was rendered. The patient then sought diagnosis and treatment with gynecology. At that time, the growths appeared as small, soft, verrucous papules in the perianal region, clinically consistent with condyloma acuminata. The patient had no history of sexually transmitted infection and her Pap smear was negative for Human Papilloma Virus (HPV). Screening for HIV, Hepatitis C, chlamydia, gonorrhea, and rapid plasma reagin were negative. Prior to destructive treatment modalities, a biopsy was performed for diagnosis confirmation. The histology showed cup-shaped invagination with a superficial keratotic core (**Figure 1)** including grains and suprabasilar clefting with dyskeratosis and acantholysis (**Figure 2**), consistent with warty dyskeratoma. HPV in situ hybridization analysis for genotype 6/11 was negative. The patient then presented to dermatology. Skin exam did not reveal any other areas of verrucous papules or plaques, ruling out Darier disease, Grover disease, and Hailey-Hailey disease. Though there is no standard treatment for warty dyskeratoma, options including cryotherapy and topicals were discussed. Based on published success in a similar case, the patient opted to try calcipotriene ointment.[1] After several months of treatment with calcipotriene ointment twice daily, the patient noted a reduction in the size and number of lesions. She reported mild irritation from the treatment which improved with less frequent application and is pleased with the outcome.

DISCUSSION

This case highlights the unexpected finding WD in a young female patient and her journey to final diagnosis. The patient presented with small, skin colored, soft verrucous papules in the perianal region. Though WD can cause pruritus, these lesions were asymptomatic. Histopathology was most consistent with the diagnosis of WD, revealing a cup-shaped invagination with a superficial keratotic core, and suprabasilar clefting with dyskeratosis and acantholysis. To our knowledge, this is the second reported case of perianal WD.[7] It may be underdiagnosed.

WDs belong to the family of acantholytic dyskeratoses, which demonstrate a distinct histological pattern of suprabasilar clefts surrounding preserved papillae, acantholytic and dyskeratotic epidermal cells, hyperkeratosis, and parakeratosis.[3] This histologic pattern can be seen in multiple types of acantholytic dyskeratosis, including Darier disease, papular acantholytic dyskeratosis (PAD), squamous cell carcinoma acantholytic type and Grover disease, all of which must be distinguished from WD.[6]

WD was first named in 1954 as an "isolated Darier disease". Darier disease is an inherited autosomal dominant mutation in the ATP2A2 gene that, unlike typical WD, presents with multiple hyperkeratotic papules and plaques in seborrheic regions.[6,8] Furthermore, Darier disease often demonstrates secondary sites of involvement including cobblestoning of the palate, V-shaped nicking of the nails, and longitudinal erythronychia, none of which were observed in our patient.[6]

PAD morphologically differs from WD as it commonly presents with multiple grayish-

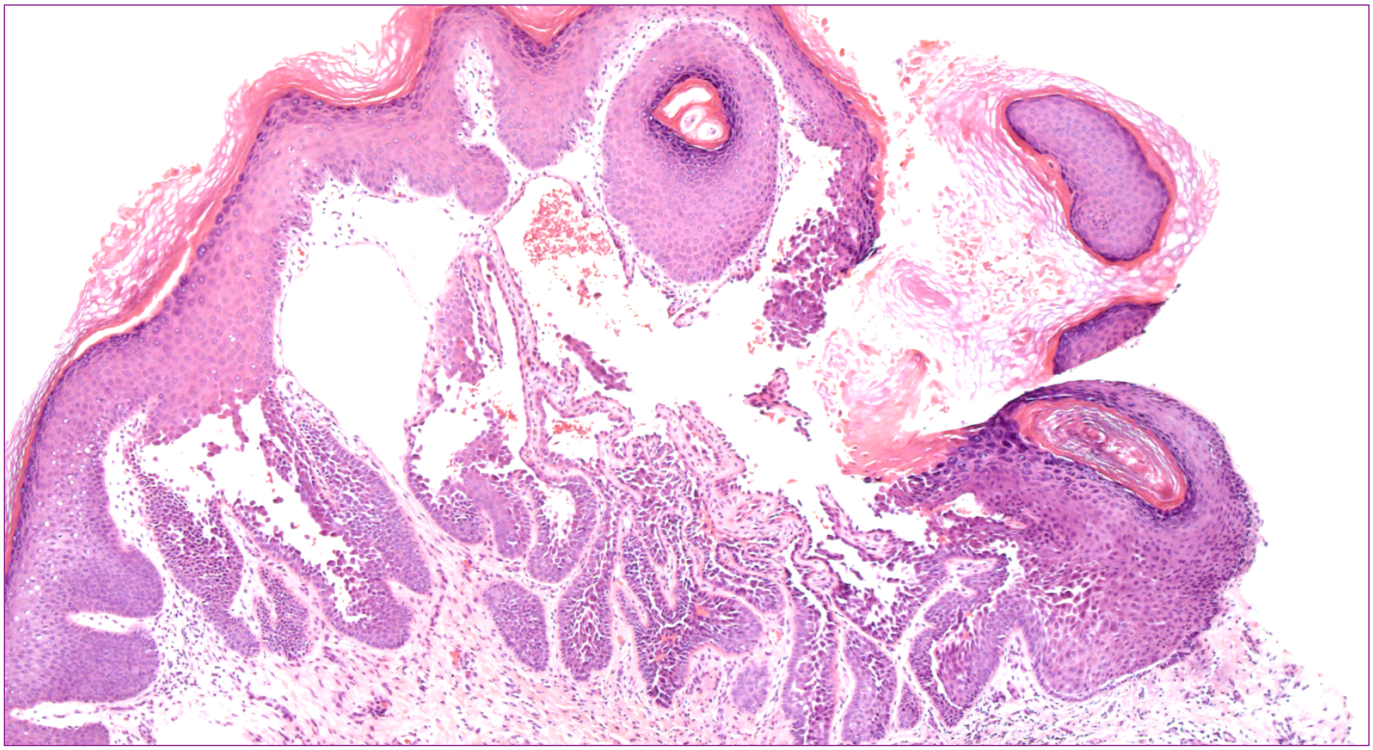

Figure 1: (H&E, 40x) Cup-shaped invagination with intraepidermal clefting and central keratotic core. Exaggerated separation within the epidermis may occur during processing.

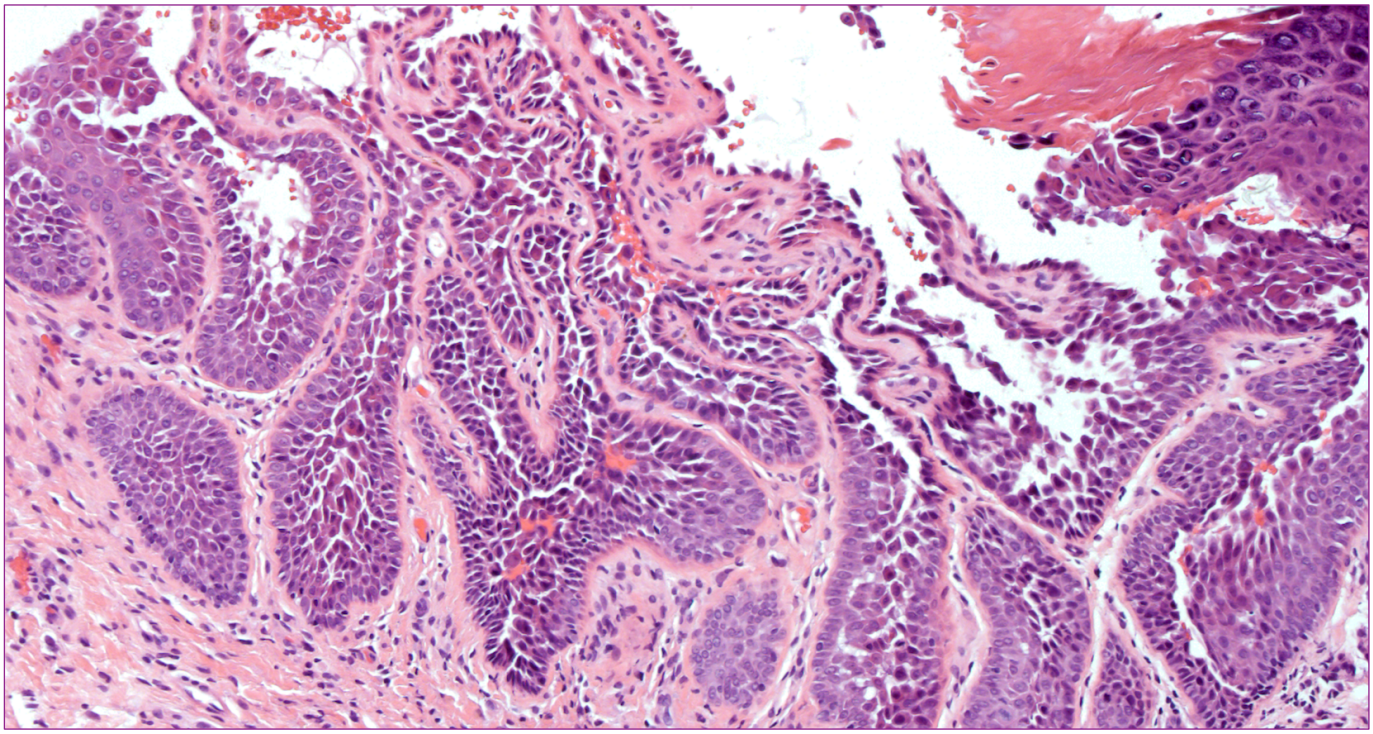

Figure 2: (H&E, 100x) Suprabasilar acantholysis and dyskeratosis with villous architecture, dilated vessels within dermal papillae, few grains are present within keratotic plug.

white keratotic papules demonstrating prominent dyskeratosis and acantholysis, though it does commonly occur in warm moist areas such as the perianal region.[9] Most cases of PAD appear to be sporadic, however, it has been diagnosed in patients with a family history of Hailey-Hailey disease, for which PAD is thought to be a possible variant.[9] Another PAD entity designated in Weedon's Skin Pathology as *acantholytic dermatosis of the genitocrural/perineal region* has been proposed but is not found to be in common use.[10] The histopathological findings in this case were more consistent with WD, however in the future there may be more distinctive diagnostic criteria for dyskeratotic lesions in the anogenital region, which would help guide diagnosis and treatment.

Neoplasms such as squamous cell carcinoma, actinic keratosis and basal cell carcinoma may also present with acantholysis and dyskeratosis, but lack of cellular atypia, mitotic figures and the absence of cyst-like configuration does not support the diagnosis of malignant neoplasm in our patient.[1]

While there is no standard treatment for WD, successful treatments with topical tazarotenic acid gel and calcipotriene ointment have been reported.[1,11] One previous case saw symptom reduction with 60 days of combined topical calcipotriene, clindamycin–benzoyl peroxide gel, and clotrimazole cream.[6] This patient noted lesion reduction with calcipotriene ointment alone after several months of twice daily application. Other reported destructive treatments including curettage with electrodesiccation, radiation therapy, and cryotherapy were all followed by recurrence.[1,6,11] Surgical excision of bothersome lesions remains the most definitive treatment modality.[1]

CONCLUSION

The patient presented in this case struggled for years with anxiety related to the presumed diagnosis of HPV and the stigma surrounding this chronic, contagious condition. She is hopeful that sharing this case will raise awareness about warty dyskeratoma in the perianal region. A skin biopsy should be considered for perianal and vulvar lesions that do not fit the typical presentation of HPV-induced verruca to confirm diagnosis and ensure adequate counseling and treatment for patients, as the misdiagnosis of HPV can cause significant emotional distress.

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