Epidermodysplasia verruciformis with plane warts over lower abdomen and genitals

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Abstract

Epidermodysplasia verruciformis (EDV) may clinically vary from pityriasis versicolor-like macules to wart-like flat papules, psoriasiform red papules, or pigmented keratotic lesions resembling seborrheic keratosis. Sun-exposed areas are commonly affected with genital areas rarely involved. It is associated with more than 30 human papillomaviruses (HPVs). In 90% cases of squamous cell carcinomas, HPV5 and HPV8 is isolated. A case of EDV with plane warts involving the genital area in a 35-year-old male is reported here.

Key words: Epidermodysplasia verruciformis, genitals, human papillomavirus, warts

INTRODUCTION

Epidermodysplasia verruciformis (EDV) is a rare inherited disorder that predisposes the patients to widespread human papillomavirus (HPV) infection. Cutaneous lesions may vary from pityriasis versicolor-like macules to wart-like flat papules, psoriasiform red papules, and pigmented keratotic lesions resembling seborrheic keratosis. EDV is associated with more than 30 HPVs. HPV5 and HPV 8 are associated with Squamous cell carcinoma in 90% cases. The lesions mainly occur on the sun-exposed areas, and genital areas are rarely involved. Genital warts in patients with EDV are rare, with only four cases reported till date.[1-4] We describe here an unusual case of a EDV presenting with pigmented seborrheic keratosis-like lesions and plane warts, both together over genital areas.

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CASE REPORT

A 35-year-old married male presented to the dermatology department with a 4-year history of multiple lesions over the abdomen, left side of the penis, and scrotum, which were asymptomatic. The lesions were nonprogressive in nature. There was a history of the same type of lesions present in the wife. There were no complaints of ulcer, any discharge, urinary tract infection, abdominal pain, or fever. There was a history of infertility present in the couple. On examination multiple hyperpigmented dome shaped papules over lower abdomen. [Figure 1a] and some warty papules present over penis and scrotum.

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[Figure 1b] There were no hypopigmented macules over any part of body. Systemic examination showed no abnormalities. Routine laboratory tests including hematological and biochemical analyses of blood and urine were within normal ranges, and serology for HIV and syphilis was negative. Biopsy from dome-shaped plane papule over the lower abdomen was taken keeping bowenoid papulosis, seborrheic keratosis, and warts as differential. Histopathology showed hyperkeratosis, acanthosis, and vacuolated cells in the upper stratum malpighi and granular layer. Keratinocytes have basophilic cytoplasm containing keratohyalin granules, suggestive of EDV [Figure 2].

Biopsy from warty lesion over the scrotum revealed hyperkeratosis and acanthosis. The stratum malphigii showed papillomatosis. The rete ridges were elongated and bent inward at both margins, and large vacuolated cells were seen in the upper stratum malpighisuggestive of warts. Dermis showed mild perivascular lymphoplasmacytic infiltration [Figure 3]. The patient was advised for cryotherapy which was given weekly for 12 weeks and all the lesions subsided. The patient was not advised oral retinoids as there was a history of infertility, and the couple was taking treatment for the same.

DISCUSSION

EDV was first described by Lewandowsky and Lutz in 1922 as an epidermal nevus.[5] It is a rare, generalized, persistent condition with a defective cell-mediated immune response to HPV infection and may be inherited or acquired. The inherited form usually presents in infancy or early childhood and is usually autosomal recessive, though autosomal dominant and X-linked dominant forms have also been reported. Two loci on chromosomes 17q25 and 2p21–p24 are associated with the disease in some families. Mutations in two genes EVER 1 and EVER 2 are linked with the disease in many but not all cases.[6] The acquired type may be secondary in immunocompromised individuals, including those with T-cell lymphoma and HIV. In our patients, the lesions developed at the age of 35 years, so it is likely to be acquired type.

There are at least 30 HPV types with a characteristic of EDV being HPVs 5, 8, 9, 12, 14, 15, 17, and 19–25. The types found more rarely include HPVs 28, 36, 37, 38, 47, 49, 50, 75, 76, 77, 93, and 96. The lesions mainly occur on the sun-exposed area but may be generalized all over the body. It is a rare, autosomal recessive genodermatosis that predisposes patients to widespread HPV infections that do not regress due to unique susceptibility to specific HPV types. Lesions typically have either the appearance of flat warts or scaly, red-brown macules that resemble lesions of pityriasis versicolor.[7]

The development of genital warts in patients with EDV is rare, with only four cases reported till date [Table 1]

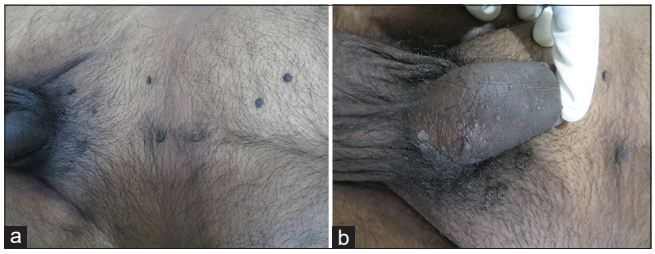

Figure 1: (a) Multiple hyperpigmented dome-shaped papules over the lower abdomen, (b) multiple flat wart-like lesions over the penis and scrotum

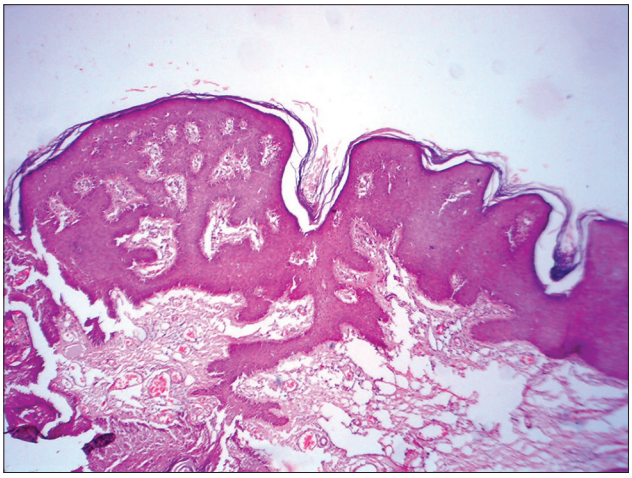

Figure 2: Hyperkeratosis, acanthosis, and vacuolated cells in the upper stratum malpighiand granular layer keratinocytes have basophilic cytoplasm containing keratohyalin granule (H and E, ×4)

Table 1: Reported cases of epidermodysplasia verruciformis and genital wart

Parameter	Age (years)/sex	Duration	Family history	Morphology	Sites	Treatment
Our study	35/male	4 years	Positive	Dome-shaped and warty lesions	Abdomen and genitals	Surgical excision + cryotherapy
Kivanc-Altunay *et al.*[1]	37/male	25 years 4 years	Negative	Pityriasis versicolor-like lesions Hyperkeratotic papules and plaques	Face and hands Perianal region	Surgical excision
Sanclemente *et al.*[2]	25/male	20 years	Negative	Pityriasis versicolor-like lesions, verrucous and moist tumors	Scrotum	Surgical excision and cryotherapy
Zhu *et al.*[4]	30/male	10 years 6 months	Negative	PV, seborrheic keratosis-like lesions Cauliflower-like exophytic masses	Face, upper limb, trunk Scrotum and axillae	Surgical excision + oral retinoids

PV=Papilloma virus

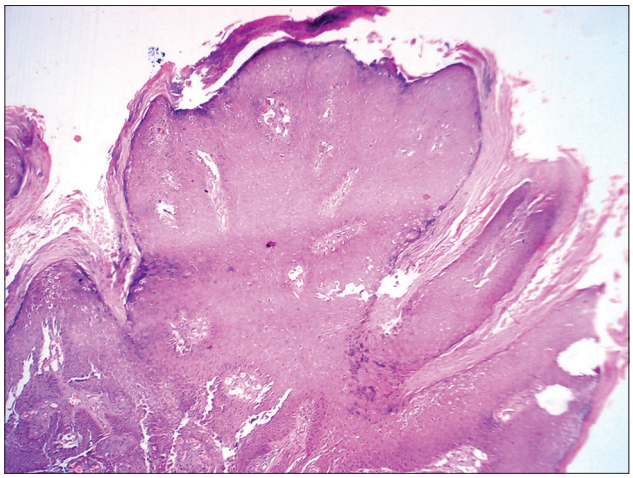

Figure 3: Hyperkeratosis, acanthosis, and papillomatosis. The rete ridges were elongated and bent inward at both margins with large vacuolated cells in the upper stratum malpighi (H and E, ×4)

Rogers *et al*. reported the development of EDV lesions in two HIV-positive immunocompromised patients.[3] Our patient presented with dome-shaped papules mimicking seborrheic keratosis and warts. The localizations of the lesions are predominantly sun-exposed areas such as the face, neck, and dorsa of the hands. However, warty lesions involving the scrotum and penis may also rarely occur[2] as reported in our case. Oral mucosa is spared. Malignant change occurs in 20%–30% of cases mainly on sun-exposed areas, and metastasis is uncommon. Carcinogenic risk is determined by HPV infection, relative immunosuppression, and sunlight exposure.

The histological picture is similar in the different clinical types of lesion of EDV. Skin biopsy of EDV shows swollen keratinocytes with vacuolation that contains basophilic granules, whereas hyperkeratosis, acanthosis, with a characteristic feature of koilocytosis (vacuolation of keratinocytes) of upper keratinocytes, and papillomatosis along with incurving of rete ridges are the main features of warts. Other differentials include seborrheic keratosis which shows basaloid cell proliferation with horn cysts as a characteristic feature in histopathology, whereas bowenoid papulosis shows windblown appearance of large atypical keratinocytes.

There is no effective treatment for EDV. Etretinate 1 mg/kg/day is helpful in some cases, but the lesions recur. Oral isotretinoin is also effective. In our patients, we had used liquid nitrogen to destroy the lesions. Topical imiquimod, calcipotriol, and squaric acid dibutyl ester are the other treatment options that can be used. Sun protection measures are essential to prevent neoplasia if lesions are over sun-exposed areas.

CONCLUSION

EDV can mimic seborrheic keratosis and warts and can involve even genitalia. Therefore, biopsy should be taken to confirm the diagnosis at the earliest in case of any dilemma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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