Sir,

Epidermodysplasia verruciformis (EV) is a rare, inherited disorder that predisposes patients to chronic widespread human papillomavirus (HPV) infection. Typical lesions are notably variable with pityriasis versicolor-like macules, wart-like flat papules, psoriasiform red papules, pigmented keratotic lesions resembling seborrheic keratosis. Wart-like lesions or atypical warts caused by HPV are especially important, because skin cancers may develop on sun-exposed areas such as the back of hands, neck and the legs. The genital areas are rarely involved and HPV lesions on this area have been previously reported only twice.[2,3] As far as we know, concurrent perianal HPV infection has not been reported to date.

A 37-year-old man presented with a verrucous mass on the perianal region that has enlarged for over 4 years. He had been operated twice for this mass during these 4 years, but the recurrence of painful lesions had occurred after each operation. On admission, it was also apparent that the patient had numerous red and brown maculopapular lesions on his face and dorsum of hands. These lesions existed for about 25 years and he did not complain about them. Examination of all systems was normal. His medical history was unremarkable except for smoking for 20 years. Homosexual history was not obtained. The patient did not have any medications and denied a family history of any skin diseases. Dermatological examination revealed multiple tan brown-colored maculopapular lesions on the face [Figure 1a] and neck, verrucous-hyperkeratotic papules and plaques on the hands and forearms, verrucous lesions on the perianal region [Figure 1b]. Some of the finger and toe nails were also hyperkeratotic. Routine laboratory tests including hematological and biochemical analyses of blood and urine were within normal limits. VDRL, TPHA and HIV serology results were all negative. HPV phenotyping was performed by polymerase chain reaction from perianal region and HPV type 53 with high oncogenic potential was detected. Genetic study was not done in our case. Two punch biopsies were performed from forearm and perianal regions. Histology of the forearm lesions revealed hyperkeratosis, papillomatosis, hypergranulosis, koilocytic changes in upper spinous layer and perivascular mononuclear cell infiltration in papillary dermis [Figure 2a]. Perianal biopsy showed dyskeratosis in epidermis, papillomatosis, loss of polarity, hyperchromatic nuclei, pleomorphism, koilocytic atypical cells which were consistent with condyloma acumina tum with high-grade squamous cell carcinoma (SCC) [Figure 2b]. Surgical excision was planned for perianal mass. Post-operative pathological
examination confirmed previous diagnosis of SCC. Although total excision provided a cure for cancer, post-operative radiotherapy was additionally planned for regional control of the cancer. The patient is now on follow-up.

EV was first described by Lewandowsky and Lutz in 1922 as an epidermal nevus. It is a rare, autosomal recessive genodermatosis that predisposes patients to widespread HPV infection that do not regress due to unique susceptibility to specific HPV types. Lesions typically have either the appearance of flat warts or flat, scaly, red-brown macules that resemble lesions of pityriasis versicolor. The localizations of the lesions are predominantly sun-exposed areas such as the face, neck and dorsa of the hands. However, genital warts involving scrotum, penis may also rarely occur. There are only two reports of genital localizations of wart-like lesions.

Malignant transformation as non-melanoma skin cancers occurs in approximately 30-50% of patients, predominantly on sun-exposed areas and rarely on non-sun-exposed skin. The high transformation rate in sun-exposed areas could be explained by the chronic ultraviolet B exposure and the probable presence of oncogenic EV-HPV in hair follicles. The exact molecular pathway between EV-HPV and development of non-melanoma skin cancer, however, remains unclear. Development of SCC in non-sun-exposed area in our case can be explained by oncogenic potential of HPV type 53.

Malignancies are commonly associated with HPV types 5 and 8 in around 90% of cases, and much less frequently with types 9, 12, 14, 17, 20 and 47, as non-melanoma skin cancers which are usually multiple, locally invasive and rarely metastatic. There is a recent case report in which HPV 22 b was found to be accountable for the development of an aggressive SCC. Our case is interesting in two ways. First is the development of perianal warts which is unusual localization for warts in EV and second is the role of HPV type 53 in the occurrence of SCC. As far as we know, HPV type 53 has never been previously reported in EV. Therefore, one should also pay attention to the warts on non-sun-exposed areas for possible malignant transformation in EV.

Ilknur Kivanc-Altunay, Hilal Kaya Erdogan, Semra Kayaoglu

Department of Dermatology, Sisli Etfal Training and Research Hospital, *Family Medicine Clinic, Sisli State Hospital, Istanbul, Turkey

Address for correspondence: Dr. Hilal Kaya Erdogan, Department of Dermatology, Sisli Etfal Training and Research Hospital Istanbul, 34377 Turkey. E-mail: hilalkayaaa@yahoo.com

Access this article online

Quick Response Code: www.ijdvl.com

DOI: 10.4103/0378-6323.75000

REFERENCES


