Acquired Epidermodysplasia Verruciformis in a Renal Transplant Recipient

Epidermodisplasia verruciforme adquirida en una paciente transplantada renal

To the Editor:

Epidermodysplasia verruciformis (EV) is a genetic disorder characterized by persistent infection of the skin by human papillomavirus (HPV). Lesions similar to those of classic EV have recently been described in patients with hereditary and acquired immunodeficiency, and these conditions have been referred to as acquired EV.

A 19-year-old woman presented with a 1-year history of multiple asymptomatic skin lesions. Topical treatment with corticosteroids and antifungal agents led to no improvement. At the age of 8, the patient had undergone a kidney transplant because of renal failure secondary to focal segmental glomerulonephritis that had begun 2 years earlier. For this reason, from age 6 she had been receiving immunosuppressive therapy, initially with prednisone and azathioprine, and from the date of transplantation with prednisone, tacrolimus, and mycophenolate mofetil. She reported no affected family members and no history of consanguinity.

Physical examination revealed multiple flat, oval papules on the upper trunk and the root of the upper limbs. The papules were about 1 cm in diameter, reddish-brown in color, and slightly scaly (Figure 1).

Histopathology of a lesion on the trunk revealed hyperkeratosis with mild parakeratosis. In a slightly acanthotic epidermis, the stratum spinosum was disorganized, with dyskeratosis, mitoses, and cells of different sizes, creating a Bowenoid appearance. In the stratum granulosum and upper stratum spinosum, cells with pale cytoplasm and a clear perinuclear halo typical of planar warts were observed (Figures 2 and 3).

Polymerase chain reaction amplification and subsequent automatic sequencing revealed the presence of HPV type 23.

EV is a rare genetic disorder that leads to increased susceptibility of the skin to infection by HPV. The first clinical signs of the disease appear in childhood as tinea versicolor–like, hypo- or hyperpigmented scaly plaques or flat papules that mimic planar warts. Up to 50% of patients develop squamous cell carcinomas in about the fourth decade of life. The HPV types most frequently associated with EV (EV-HPV) are types 5, 8, 17, and 20. Types 5 and 8 have been associated with a risk of malignant transformation and are found in more than 90% of cancers in these patients. Types 14d, 17, 20, and 47 have also been associated with malignant transformation. Although the role of HPV in oncogenesis has not been fully defined, it has been postulated that the virus may be involved in the initial stages of this process, acting synergistically with other carcinogens such as ultraviolet radiation.

In recent years there have been case reports of EV-like dermatosis in several types of immunosuppression, including human immunodeficiency virus infection, renal transplant, graft-versus-host disease, and systemic lupus erythematosus. These conditions have been called EV-like syndromes or acquired EV.

EV-HPV are detected in about 80% of nonmelanoma skin cancers and precancerous lesions in immunocompromised patients. However, the appearance of EV-like lesions in relation to HPV infection is very rare, suggesting that patients with this clinical presentation may have genetic susceptibility. Haplotype HLA-DQB10301, which is associated with classic EV, was also found in 3 patients with acquired EV and may represent a susceptibility allele or a marker of genetic predisposition to the disease.

The clinical and histopathological signs of acquired EV are similar to those of classic EV. The characteristic histopathological finding is mild to moderate hyperkeratosis, acanthosis due to the presence of cells with abundant, pale, basophilic cytoplasm and occasional conspicuous perinuclear halos. This typical presentation is rare and was found in only 3 of 18 patients with classic EV and in 8 of 19 patients with acquired EV. In the remaining cases histopathology revealed thickening and disorganization of the stratum granulosum and isolated cells with a conspicuous perinuclear halo. In our case histopathology revealed planar wart–type images, alternating and coexisting in different zones with areas that had a Bowenoid appearance.

In conclusion, we present the case of a renal transplant patient with EV-like lesions showing the presence of HPV type 23. The absence of affected family members or consanguinity and the fact that the condition appeared after a prolonged period of immunosuppression led to the diagnosis of acquired EV.

References


S. Gómez-Bernal, L. Rodríguez-Pazos,
M.M. Pereiro-Ferreirós, J. Toribio*

*Corresponding author. 
E-mail address: jaime.toribio@usc.es (J. Toribio).