Marjolin Ulcer: A Report of 2 Cases of Squamous Cell Carcinoma Arising From Posttraumatic Soft-Tissue Scarring☆

Úlcera de majorlin: 2 casos de carcinoma escamoso sobre cicatriz por trauma en tejidos blandos

To the Editor:

Marjolin ulcer is an invasive neoplasm that arises from chronic wounds of any etiology, including traumatic wounds, chronic ulcers, and partial or total skin grafts.[1,2] Squamous cell carcinoma, although rare, is the most frequently produced neoplasm.[3] Here, we present 2 representative cases.

Case Descriptions

Case 1

The patient was a 56-year-old man with a history of soft-tissue trauma caused by crushing of the left leg, for which he had undergone allograft reconstruction 38 years earlier. He was seen for a progressively enlarging verrucous, ulcerated lesion on the left leg that had appeared 1 year earlier (Fig. 1A). A biopsy was performed on suspicion of malignancy and revealed an infiltrating squamous cell carcinoma (Fig. 1B and C). Imaging studies showed bone infiltration, which necessitated amputation of the affected limb.

Case 2

The patient was a 51-year-old man with a history of soft-tissue trauma with circumferential compromise and tissue

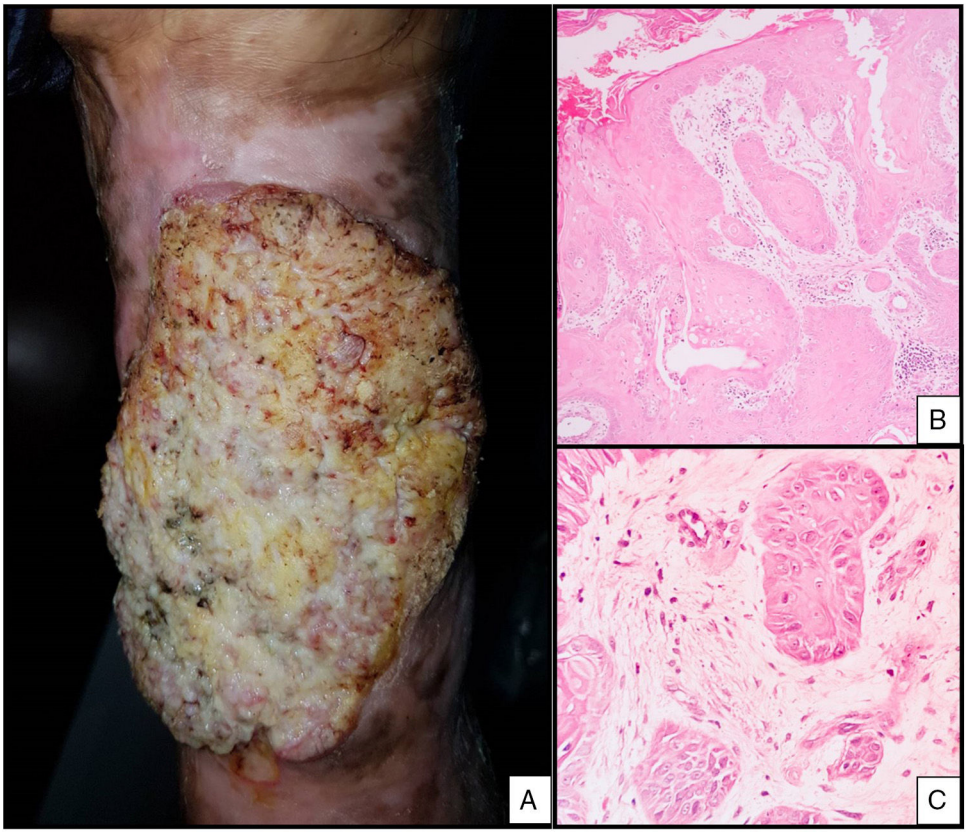

Figure 1 A, Ulcerated verrucous tumor on the left leg. B, Nests of squamous cells infiltrating the dermis (hematoxylin-eosin, original magnification ×10). C, Nuclear pleomorphism (hematoxylin-eosin, original magnification ×40).

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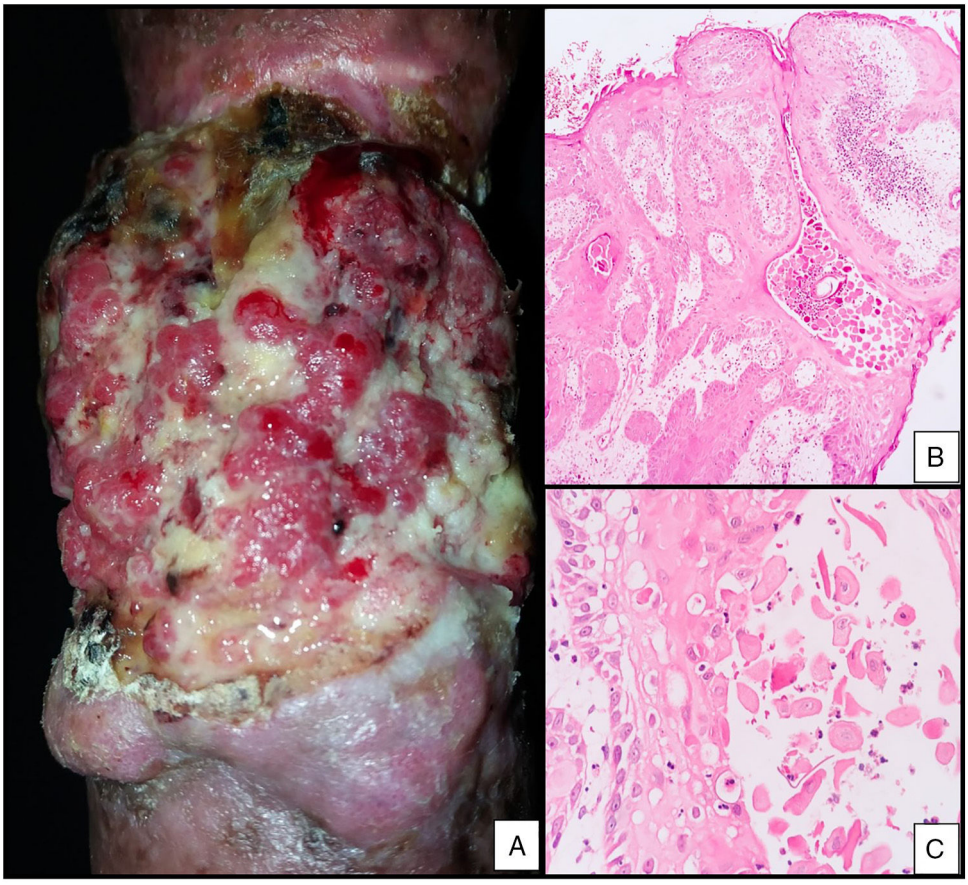

Figure 2 A, Ulcerated verrucous tumor on the right forearm. B, Well-differentiated infiltrating squamous cell carcinoma (hematoxylin-eosin, original magnification ×10). C, Mitosis, nuclear pleomorphism (hematoxylin-eosin, original magnification ×40).

avulsion caused by a crush injury received in a traffic accident 30 years earlier. He was seen for a progressively enlarging verrucous, ulcerated lesion, located on the scar on his right forearm that had appeared 1 year earlier (Fig. 2A). A biopsy was performed on suspicion of malignancy and revealed an infiltrating squamous cell carcinoma (Fig. 2B and C). The patient underwent chemotherapy and radiation therapy, after which tumor recurrence necessitated amputation of the affected limb.

Discussion

Squamous cell carcinoma is the second most common cutaneous neoplasm after basal cell carcinoma. This tumor type can occur de novo or can arise from precancerous lesions or from lesions caused by prior trauma.[4] Tumors that arise from chronic wounds, including chronic ulcers, are known as Marjolin ulcers, and have an estimated incidence of 1.3%–2.2%, which increases with the chronicity of the preexisting lesion.[1] Development of Marjolin ulcer has been reported up to 65 years after the appearance of the initial lesion, although the mean latency is approximately 40 years.[5] In our 2 cases the mean latency to lesion appearance was 34 years after the initial trauma. The inflammatory environment of ulcers (large amounts of cytotoxic products derived from macrophage activity within the wound, cells with a high mitotic capacity, an ischemic environment, and poor lymphatic drainage) favors carcinogenesis and the formation of tumors such as Marjolin ulcer.[1,4,5] In cases of clinical suspicion of this tumor type histology should be performed to confirm the diagnosis. Histology reveals findings compatible with squamous cell carcinoma. The degree of tumor differentiation is the primary long-term prognostic factor; greater degrees of differentiation, ulceration, and dermal infiltration by tumor cells are associated with a higher rate of recurrence.[4] Tumor staging should be performed to evaluate lymph node involvement distant from the tumor and to determine the appropriate therapeutic approach. Surgery is the treatment of choice, with reported 2-year survival rates of 66%–80%.[5] Adjuvant treatments such as radiotherapy or chemotherapy may be useful in patients with inoperable tumors or those who refuse surgery.

Conclusion

Lesion chronicity, clinical findings, and clinical-pathological correlation are key to the diagnosis of Marjolin ulcer. A high index of suspicion is necessary to establish an early diagnosis and thereby provide patients with timely treatment.

Conflict of Interest

The authors declare that they have no conflicts of interest.

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V.L. Dávila Osorio,[a,*] L. Méndez Gaviria,[a] J.R. Amador Patarroyo,[a] M.I. González[b]

[a] *Departamento de Dermatología, Hospital Militar Central, Universidad Militar Nueva Granada, Bogotá, Colombia*
[b] *Departamento de Patología, Hospital Militar Central, Bogotá, Colombia*

* Corresponding author.
E-mail address: dra.daviladermatologa@gmail.com (V.L. Dávila Osorio).

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Classic Ehlers-Danlos Syndrome: Clinical and Ultrasound Findings☆

Síndrome de Ehlers-Danlos clásico: hallazgos clínicos y ecográficos

To the Editor:

Ehlers-Danlos syndrome (EDS) is a heterogeneous group of congenital connective tissue diseases caused by mutations in genes involved in the synthesis or processing of collagen fibers.[1] The phenotypic manifestations of EDS vary greatly, and mild cases can go unnoticed until late in life. We present the case of a girl who was diagnosed with classic EDS in our hospital based on clinical and ultrasound findings.

An 8-year-old girl was referred from the emergency department for evaluation of a painful lesion on the left leg that had appeared several weeks earlier after a fall from a ladder. Physical examination revealed a subcutaneous bulge of about 4 cm in diameter in the left pretibial region with yellowish-purpuric overlying skin and a strikingly gummy consistency. The patient had reticulated erythematous-violaceous lesions on the right leg and dehiscent and atrophic scars on the right leg and left knee (Fig. 1). High-frequency ultrasound (18 MHz) of the left pretibial lesion revealed an anechogenic collection delimited by a thin pseudocapsule, compatible with an organized hematoma (Fig. 2A). Septal edema compatible with traumatic panniculitis was evident in the surrounding hyperechogenic subcutaneous tissue. Doppler signal was absent. In the directed anamnesis the patient's mother reported that the girl had been born preterm due to premature rupture of membranes and had muscular hypotonia during the neonatal period. The patient was undergoing tests in the endocrinology department of another hospital for short stature and disproportion between the trunk and limbs. The family history provided by the mother included joint hyperlaxity, abnormal scarring, and early osteoarthritis. The patient also presented with skin hyperextensibility, joint hypermobility, and Gorlin sign (ability to reach the nose with the tip of the tongue) (Fig. 3). Based on these data a suspected diagnosis of classic EDS was established. A cardiological examination, including electrocardiogram and echocardiography, revealed no findings of interest, and the

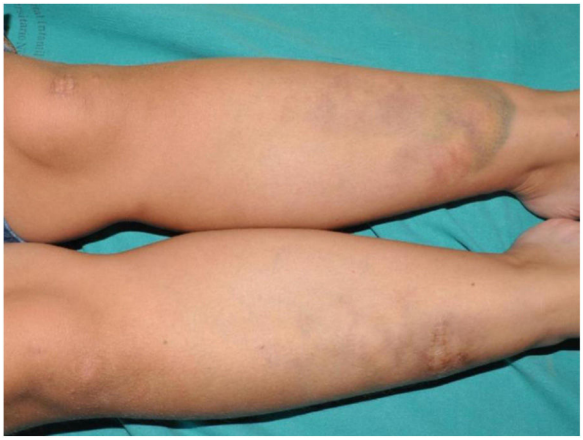

Figure 1 Extensive purpuric lesion on the anterior aspect of the middle and distal third of the left leg, and reticulated erythematous-violaceous lesions on the contralateral leg. Note the presence of 2 atrophic scars on the distal third of the right leg and on the left knee.

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