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E-CASE REPORT

Eccrine Porocarcinoma: Patient Characteristics, Clinical and Histopathologic Features, and Treatment in 7 Cases☆

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KEYWORDS
Eccrine porocarcinoma;
Histological prognostic factors;
Therapeutic algorithm;
Sentinel lymph node biopsy;
Mortality

Abstract Eccrine porocarcinoma is a rare, malignant cutaneous adnexal tumor that arises from the ducts of sweat glands. Found mainly in patients of advanced age, this tumor has diverse clinical presentations. Histology confirms the diagnosis, detects features relevant to prognosis, and guides treatment. Growth is slow, but the prognosis is poor if the tumor metastasizes to lymph nodes or visceral organs. We report 7 cases of eccrine porocarcinoma, describing patient characteristics, the clinical and histopathologic features of the tumors, and treatments used. Our observations were similar to those of other published case series. Given the lack of therapeutic algorithms or protocols for this carcinoma, we propose a decision-making schema based on our review of the literature and our experience with this case series. The algorithm centers on sentinel lymph node biopsy and histologic features.

PALABRAS CLAVE
Porocarcinoma ecrino;

Porocarcinoma ecrino: aspectos demográficos, clínicos, histopatológicos y terapéuticos en una serie de 7 pacientes

Resumen El porocarcinoma ecrino es un tumor anexial cutáneo maligno poco frecuente, que deriva de la porción ductal de las glándulas sudoríparas. Predomina en pacientes de edad

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Factores pronósticos histológicos;
Algoritmo terapéutico;
Biopsia selectiva ganglio centinela;
Mortalidad

avanzada, pudiendo adoptar diversas presentaciones clínicas. El estudio histológico confirma el diagnóstico y establece factores pronósticos relevantes a la hora de decidir el tratamiento. El curso evolutivo es lento, pero su pronóstico es sombrío cuando aparecen metástasis linfáticas o viscerales. Presentamos 7 pacientes diagnosticados de porocarcinoma ecrino, y describimos los aspectos epidemiológicos, clínicos, histopatológicos y los datos relacionados con el tratamiento de cada uno de ellos. Comparamos los datos obtenidos con las mayores series de casos publicadas, obteniendo similares resultados. Ante la ausencia de algoritmos terapéuticos protocolizados se propone un esquema de tratamiento basado en la literatura revisada y la experiencia personal con nuestra serie, que tiene como ejes centrales los factores pronósticos histológicos y la biopsia selectiva de ganglio centinela.

Introduction

Eccrine porocarcinoma (EP) was described in 1963 by Pinkus and Mehregan,[1] but it was Mishima and Morioka[2] who introduced this name in 1967. EP is a rare neoplasm (0.005% to 0.01% of all malignant skin tumors).[3] It arises most commonly on the lower limbs[4–6] and on the head and neck[7–9] of patients of advanced age,[10] with no difference between the sexes.[5,7] Clinical presentation is variable, and the disease can mimic many other tumors. Histology not only confirms the diagnosis but also identifies prognostic factors that will determine the therapeutic approach.[4] The majority of descriptions in the literature are isolated case reports or small case series, making it difficult to draw up standardized treatment guidelines beyond surgical excision. Prognosis is very poor when metastases are present.[11]

Case Descriptions

We present 7 patients diagnosed with EP, evaluating their demographic and clinical characteristics (Table 1), histopathology findings (Table 2), and data regarding treatment and clinical course (Table 3).

The mean age of the patients in the series was 69 years (range, 39-91 years; median, 75 years). There were 5 (71%) men and 2 (29%) women. The most common site of the tumor was on the lower limbs (3/7; 43%). Tumor diameter was equal to or less than 2 cm (mean, 1.46 cm; median, 1.5 cm). The most frequent clinical pattern was the pink papule (3/7; 43%) (Fig. 1). Recent changes in the lesions were reported by 71% (5/7) of patients. Time since first appearance of the lesion varied between 2 and 60 years (mean, 15 years; median, 4 years). EP was only suspected in 1 case (14%).

Histology confirmed the diagnosis of EP in all cases, observing broad, anastomosing cords, solid columns, and nests of polygonal basaloid cells with hyperchromatic nuclei, prominent nucleoli, and mitotic figures, with spread from the epidermis into the papillary and reticular dermis, invading the stroma asymmetrically; these changes were accompanied by dermal ductal structures (intracytoplasmic lumina or mature ducts [Fig. 2A]). In situ EP was only detected in 1 case (14%). Mean tumor thickness in the other patients was 5.49 mm (range, 3.1-7 mm; median, 5.56 mm). The most common growth pattern was the pushing pattern (4/7; 57%; Fig. 2B), in which the cords of neoplastic cells advance by pushing the dermis, from which they are clearly separated. The infiltrative pattern was observed in 43% (3/7) of cases, with poorly defined tumor nests invading the dermis and even the hypodermis. Intraepidermal spread was observed in 1 patient (14%), the presence of necrosis in 2 (29%) (Fig. 2C), and a benign component (poroma) in 1 case (14%). The number of mitoses per square millimeter varied between 1 and 18 (mean 5.7; median, 3) (Fig. 2D). Only patient 5 presented lymphovascular invasion (1/7, 14%). Immunohistochemistry was performed in 5 patients and was positive for carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA).

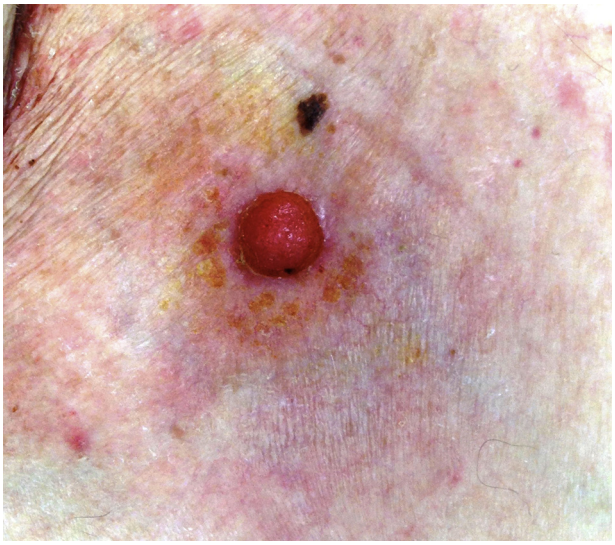

Figure 1 Clinical image of an eccrine porocarcinoma in a patient in the series: a pink papule less than 2 cm in diameter. This is the most common form of presentation of this neoplasm.

Initial treatment was surgical excision of the lesion, with margins of 3 to 5 mm. Selective sentinel lymph node biopsy (SLNB) was considered in 3 patients (43%), based on the histologic characteristics (tumor thickness, mitoses, and vascular and lymphatic invasion). SLNB was positive in patient 2, and regional lymphadenectomy was therefore performed with adjuvant radiotherapy. Patient 5 rejected the procedure and the result was negative in patient 6. Imaging studies (ultrasound [US] and/or computed tomography [CT]) revealed lymph node metastases only in patient 5. Lymphatic spread was present in 29% (2/7) of cases at the time of diagnosis.