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## Squamoid eccrine ductal carcinoma: series of five cases of a rare tumor<sup>☆</sup>



Dear Editor,

Squamoid eccrine ductal carcinoma (SEDC) is a rare malignant cutaneous neoplasm that is biphasic on histopathology, with both squamous (tumor surface) and ductal eccrine (tumor depth) differentiation, often being confused with squamous cell carcinoma (SCC), especially in superficial biopsies. SEDC has significant clinical relevance due to its potential for metastasis and local aggressiveness.<sup>1,2</sup> Therefore, the objective of this case series is to demonstrate the

SEDC aiming to improve knowledge and management of this rare neoplasm.

Table 1 summarizes the main information of the five SEDC cases. The disease affected exclusively elderly male adults, averaged 68 years old. All cases occurred in the head and neck region, with a predilection for the face (three of five cases). There was a history of immunosuppression due to organ transplantation in three patients and previous local radiotherapy in one case. On histopathology (Fig. 1), the tumors showed an infiltrative growth pattern in the dermis, occasionally invading the subcutaneous and muscular tissue. In the most superficial regions of the tumors, squamous differentiation was observed, similar to well-differentiated SCC, while in the deeper regions, there were different

**Table 1** Summary of cases of squamoid eccrine ductal carcinoma.

Gender	Age (years)	Site of the tumor	Immunosuppression	Presence of perineural and angiolymphatic invasion	Treatment	Time of follow-up (months)	Outcome
1 M	72	Eyebrow	Kidney transplant recipient using tacrolimus, azathioprine and prednisone	Perineural invasion	Not applicable	Does not apply	Lost to follow-up
2 M	60	Temporal region	Kidney transplant recipient using tacrolimus and everolimus	Perineural and angiolymphatic invasion	Surgical excision with intraoperative margin control by frozen sections	35	No signs of local recurrence or metastasis
3 M	73	Forehead	No	No	Conventional surgical excision	21	No signs of local recurrence or metastasis

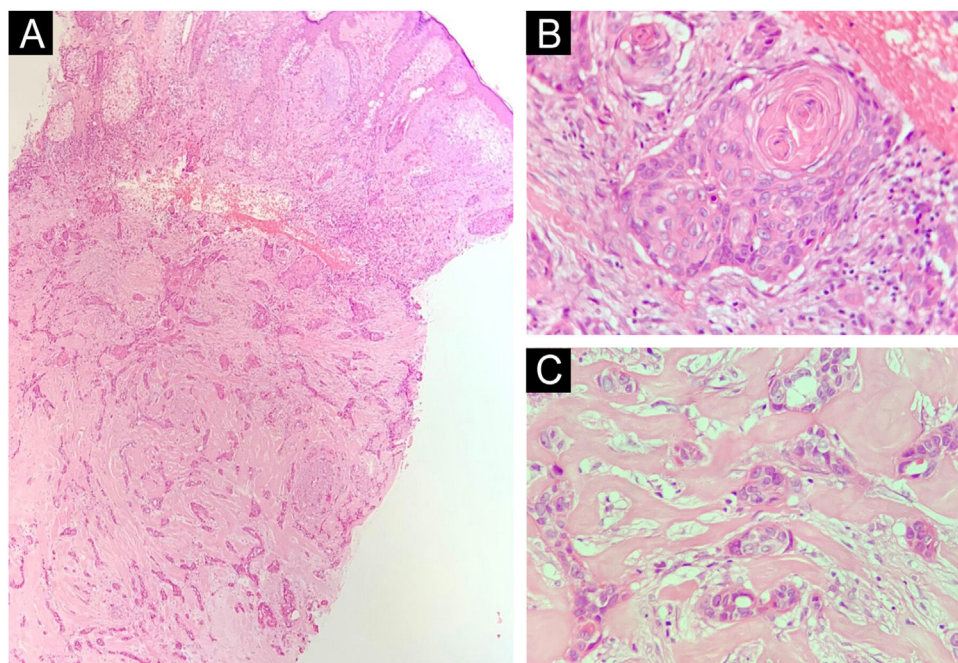
clinical and histopathological presentation of five cases of

<sup>☆</sup> Study conducted at the Faculty of Medicine, Hospital das Clínicas, Universidade de São Paulo, São Paulo, São Paulo, Brazil.

Table 1 (Continued)

Gender	Age (years)	Site of the tumor	Immunosuppression	Presence of perineural and angiolymphatic invasion	Treatment	Time of follow-up (months)	Outcome
4 M	73	Cervical region	No	No	Conventional surgical excision	36	Local recurrence
5 M	62	Scalp	Kidney transplant recipient using prednisone	Perineural invasion	Surgical excision with intraoperative margin control by frozen sections	22	Local recurrence and lung and lymph node metastasis, resulting in death

M: Male.

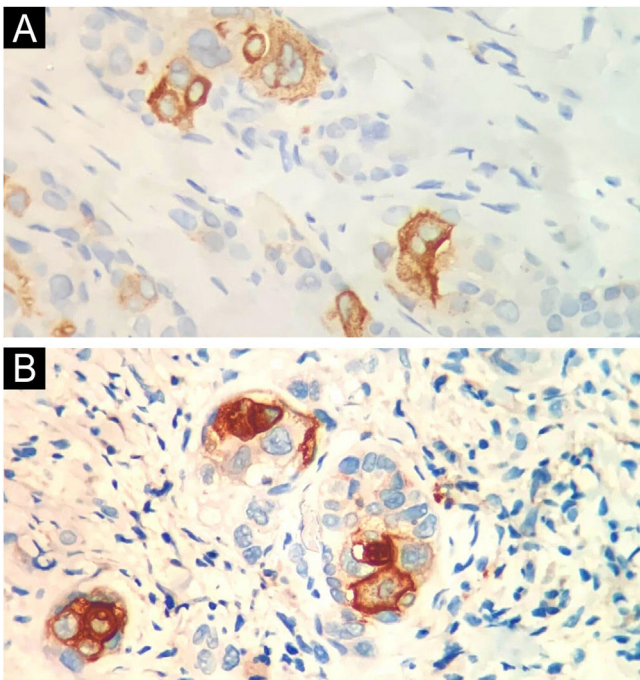


**Figure 1** Histopathology. Histological section showing tumor with infiltrative growth invading the deep dermis (A: Hematoxylin & eosin,  $\times 4$ ), with squamous differentiation in the tumor surface (B: Hematoxylin & eosin,  $\times 20$ ) and eccrine ductal differentiation in the tumor depth (C: Hematoxylin & eosin,  $\times 10$ ).

degrees of ductal differentiation and nests and cords of epithelial cells with moderate to severe atypia, surrounded by desmoplastic stroma. The presence of perineural invasion was observed in three cases, and angiolymphatic invasion in one case. Ductal differentiation was confirmed by immunohistochemistry positivity for epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA) in all tumors (Fig. 2). Information on treatment and evolution is available for four of the five patients, as one of them was lost to follow-up after incisional biopsy of the tumor. All four cases underwent surgical treatment with free surgical margins after the procedure. The surgical margins were evaluated intraoperatively by frozen sections in two of the four

cases, while in the other two cases, margin analysis occurred after surgery. The mean follow-up time after surgery was 28.5 months. Evidence of local recurrence was observed in two patients: one had undergone conventional surgery with margin analysis, while the other had intraoperative margin analysis performed using frozen sections. Moreover, in addition to developing local recurrence, one of the patients also showed lymph node and lung metastases, resulting in death.

SEDC is traditionally classified as a subtype of eccrine carcinoma. However, its etiopathogenesis remains uncertain, as it is not clear whether it arises from the eccrine ducts with subsequent squamoid differentiation, whether it is an SCC subtype, or whether it is a truly hybrid tumor. SEDC mainly

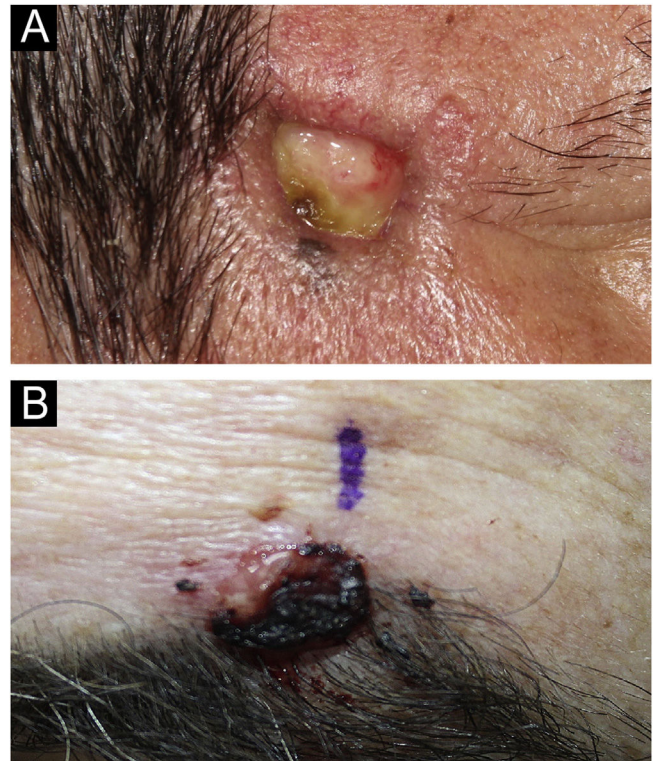


**Figure 2** Immunohistochemistry positivity for EMA (A) and CEA (B), confirming the presence of ductal differentiation.

affects the elderly, generally in the seventh or eighth decade of life, and it is suggested that immunosuppression may be a risk factor. Most cases are observed on sun-damaged skin, especially on the head and neck, with the face being the most common location. Clinically, it appears as small, often ulcerated, nodules and plaques, as shown in Fig. 3.<sup>1-3</sup>

The diagnosis is made through anatomopathological examination, in which the SEDC presents as a biphasic tumor. In the superficial areas of the tumor, squamous differentiation occurs with connection to the epidermis, while in deeper areas there is clear ductal eccrine differentiation. Squamous differentiation is typically absent in deeper regions. Furthermore, infiltrative growth is observed, with the presence of cords of cytologically atypical epithelial cells, as well as a surrounding desmoplastic stromal response.<sup>1-5</sup> SEDC frequently extends to the subcutaneous tissue and may be associated with perineural and angiolymphatic invasion, factors that may explain its high local recurrence rate (25%), even after complete excision, and its potential for metastasis (13%), according to literature data.<sup>1</sup> SEDC demands involves wide local excision and regular clinical follow-up, with Mohs micrographic surgery as a beneficial option.<sup>1</sup>

Therefore, given the rarity and lack of knowledge associated with this neoplasm, together with its potential for unfavorable outcomes, it is essential to conduct additional studies to expand the understanding and management of SEDC. The present findings suggest the need for intensified surveillance of kidney transplant recipients, as three of the five cases occurred in this group of patients. Raising awareness about this neoplasm is essential, as it is probably underdiagnosed, aiming to ensure its early diagnosis and adequate treatment and, consequently, improve clinical outcome.



**Figure 3** Clinical aspect. (A) Normochromic plaque, measuring approximately 2 cm, with irregular vessels at the border and an ulcerated center. (B) Erythematous nodule, measuring approximately 2 cm, presenting a friable surface covered by an hematic crust.

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### Authors contributions

Cecília Mirelle Almeida Honorato: Design and planning of the study; drafting and editing of the manuscript; collection, analysis, and interpretation of data; critical review of the literature.

Giovanna Gelli Carrascoza: Drafting and editing of the manuscript; collection, analysis, and interpretation of data; critical review of the literature.

Nubia Marrer Abed: Approval of the final version of the manuscript; intellectual participation in the propaedeutic and/or therapeutic conduct of the studied cases; critical review of the manuscript.





Fernanda Gonçalves Moya: Approval of the final version of the manuscript; design and planning of the study; effective participation in research orientation; critical review of the manuscript.

### Conflicts of interest

None declared.

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## Translation, cultural adaptation, and validation of the Family Dermatology Life Quality Index instrument into the Brazilian Portuguese language (FDLQI-BRA)\*



Dear Editor,

Dermatological diseases are very prevalent; however, despite generally showing low mortality, they can have high morbidity and impact on Quality of Life (QoL). Moreover, living with people with dermatoses can reflect anguish, requiring care and financial expenses. Therefore, dermatological diseases can interfere with family dynamics. The impact of the disease on patients QoL is called “primary impact”, and when it involves family members and/or cohabitants, it is called “secondary impact”.<sup>1</sup> Understanding the difficulties faced by family members is essential for creating educational programs and planning support to promote the QoL of those who live with dermatological patients.

The Family Dermatology Life Quality Index (FDLQI) is a generic, self-completed instrument for assessing the secondary impact on people living with patients with dermatological diseases. It consists of ten items with responses graded on a Likert-type scale, ranging from 0 (“not at all”) to 3 (“very much”), considering physical aspects (fatigue and physical overload), psychological and social aspects, as well as those related to personal, work and financial relationships.<sup>2</sup> It showed high internal consistency for several languages such as English (original), Japanese, and Persian.<sup>3,4</sup> It was developed at Cardiff University (Wales), by the same group that developed the Dermatology Life Quality Index (DLQI). There are no other generic instruments for

assessing the QoL of people living with skin diseases. However, there are specific questionnaires available, such as the Psoriasis Family Index (PFI) and the Family Dermatitis Impact (FDI), both translated and validated in Brazil.<sup>5,6</sup>

A methodological study was carried out aiming at translating, culturally adapting, and validating the FDLQI into Brazilian Portuguese (FDLQI-BRA). The project was approved by the institutional ethics committee and consent was obtained from the participants.

After obtaining the authors’ authorization, the translation into Portuguese and back-translation were carried out. For this stage, four experts fluent in Portuguese and English, and one non-expert, generated a consensual translated version, the back-translation of which was approved by the instrument authors.<sup>7</sup>

For the cultural adaptation, ten family members of patients with dermatological diseases answered a questionnaire about the clarity of the language used, the adequacy of the linguistic terms adopted, the applicability of the instrument, and its relevance in dermatological clinical practice. This stage generated the final Brazilian version: FDLQI-BRA (Supplementary Materials 1 and 2).

For content validation, five experienced dermatologists were selected and asked to evaluate the items according to their relevance and pertinence. This assessment was carried out using a Likert scale, graded from 1 to 5 – with 1 being irrelevant/not very pertinent; and 5 very relevant/pertinent.<sup>7</sup> The calculation of the Content Validation Index (CVI) resulted in scores greater than or equal to 0.8 in most items, with the exception of item 6 (“recreation and leisure”), with 0.6.<sup>8</sup>

For the FDLQI-BRA validation phase, 111 participants, aged 18 years or older, who lived in the patient’s home were included. The non-inclusion criteria were family members with complicated or uncontrolled comorbidities, which could interfere with their QoL. The initial sample size was based on the need for around ten participants for each item of the unidimensional instrument being validated, totaling at least 100 family members.<sup>9</sup>

\* Study conducted at the Dermatology Outpatient Clinic, Faculty of Medicine, Universidade Estadual Paulista, Botucatu, SP, Brazil; Private Practice, Sorocaba, SP, Brazil.