

Primary Adenoid Cystic Carcinoma of the Skin

Eduardo Henrique Jorge Lago, MD; Juan Piñeiro-Maceira, MD, PhD; Marcia Ramos-e-Silva, MD, PhD; Sueli Carneiro, MD, PhD

We report a case of primary adenoid cystic carcinoma of the skin in a 44-year-old Brazilian man that was present for 6 years with no metastasis. It is a rare malignant neoplasm of the eccrine gland. Metastases have been found in a few cases, usually to the lungs or lymph nodes. This neoplasm microscopically presents with large nests of cells with adenoid or cribriform features and many epithelial small solid islands. Histopathologic analysis is necessary for diagnosis.

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Primary adenoid cystic carcinoma of the skin was first described by Boggio¹ in 1975. It is an eccrine gland carcinoma characterized by indolent and progressive growth, firm consistency, poorly defined limits, and intradermal or subcutaneous location.² It occurs most often on the scalp and affects middle-aged patients with a female predilection. Its size varies from 0.5 to 8 cm. Local recurrence occurs in nearly 50% of cases.² Microscopic features are identical to salivary gland neoplasms. Metastasis to the lungs or lymph nodes has been reported in approximately 20% of cases of primary adenoid cystic carcinoma of the skin.²⁻⁹

From Hospital Universitário Clementino Fraga Filho, School of Medicine, Federal University of Rio de Janeiro, Brazil. Drs. Lago, Ramos-e-Silva, and Carneiro are from the Sector of Dermatology and Post-Graduation Course in Dermatology. Dr. Piñeiro-Maceira is from the Sector of Pathology and Post-Graduation Course in Dermatology. Dr. Carneiro also is from Hospital Universitário Pedro Ernesto, School of Medicine, State University of Rio de Janeiro. The authors report no conflict of interest.

Correspondence: Marcia Ramos-e-Silva, MD, PhD, Rua Dona Mariana 143 / C-32, 22280-020 Rio de Janeiro, Brazil (ramos.e.silva@dermato.med.br).

Case Report

A 44-year-old Brazilian man presented with a nodular lesion on the left thigh. The lesion started as a papule but slowly and progressively enlarged. Over 6 years it became a large tuberous nodular lesion on the proximal and internal region of the left thigh measuring 12×10 cm with ulcerative and vegetating



Figure 1. A tuberous nodular lesion on the posterior aspect of the left thigh.

areas (Figure 1). He described local pain and bleeding following small traumas. He did not report fever, chills, dyspnea, or weight loss. Peripheral lymph nodes were not palpable. Examination of the salivary gland revealed no abnormalities. The erythrocyte sedimentation rate was within reference range. Chest radiograph, abdominal and pelvic ultrasonography, and computerized tomography of his left thigh and groin showed no alterations.

Biopsy results revealed a cribriform pattern with nests and sheets of epithelial cells in the reticular dermis with multiple cystic spaces of several sizes. Epidermal hyperplasia was observed. The papillary dermis showed no involvement (Figures 2 and 3). The nuclei were large, vesicular, and piled up in some areas. Periodic acid–Schiff stain delineated the cystic spaces and secretory material. Immunohistochemical analysis supported the diagnosis. Carcinoembryonic antigen marked the cystic cavities and secretory material (Figure 4). Immunoperoxidase staining for S-100 protein marked almost the totality of cells, favoring the eccrine origin though not specific. High-molecular-weight cytokeratin diffusely marked most epithelial cells, sharply delimiting the ductal structures.

The clinical differential diagnosis included Buschke–Löwenstein tumor, and the histologic differential diagnosis included adenoid basal cell carcinoma. The tumor was excised with 1-cm margins of normal-appearing skin. The patient was consulted every 8 months without any recurrence in the last 3 years.

Comment

Primary cutaneous adenoid cystic carcinoma of the eccrine gland is an uncommon neoplasm, but metastasis of salivary gland adenoid cystic carcinomas are relatively frequent in the skin.⁹ The diagnosis of this cutaneous adnexal tumor cannot be done on a clinical basis; histopathologic study is mandatory. The typical histologic presentation consists of nests of epithelial cells in the reticular dermis with cribriform features, which renders the diagnosis feasible by experienced pathologists on hematoxylin and eosin–stained sections. This tumor should be distinguished from the adenoid basal cell carcinoma; as a result, immunohistochemical analysis is useful to prove the eccrine origin of the tumor. Perineural invasion, which is related to an increase in recurrence rate,^{1,4,5,7,8} was not observed in our patient.

Primary adenoid cystic carcinoma of the skin is microscopically composed of large nests of cells with adenoid or cribriform features and many epithelial small solid islands. Cribriform nests present with 2 kinds of cystic spaces: pseudocysts and true glandular

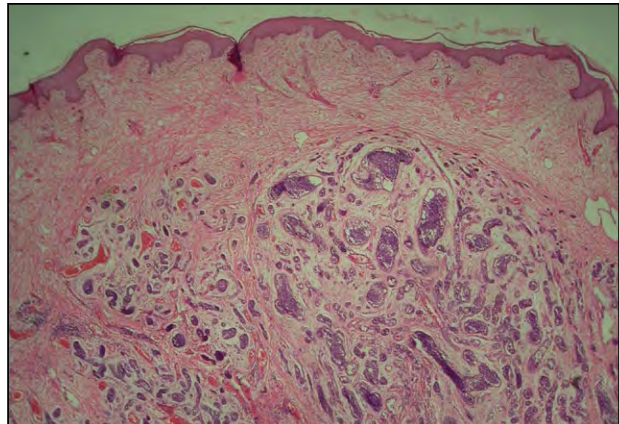


Figure 2. Numerous nests of epithelial cells with multiple cystic spaces characterizing a cribriform morphology. There was no involvement of the epidermis and papillary dermis (H&E, original magnification $\times 40$).

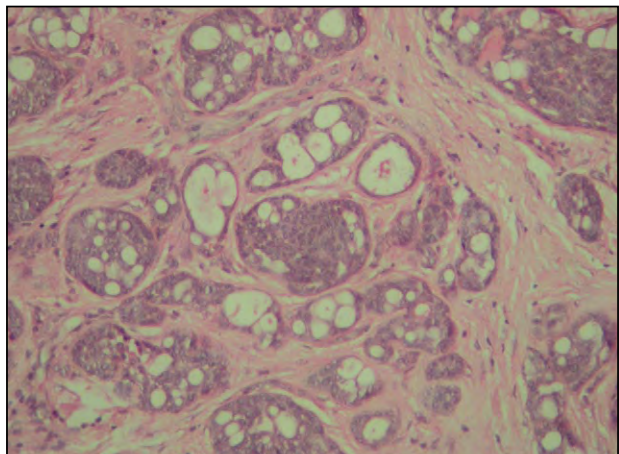


Figure 3. A cribriform pattern consisting of groups of cuboid cells with multiple foci and cystic differentiation surrounded by hyalinized dermis (H&E, original magnification $\times 100$).

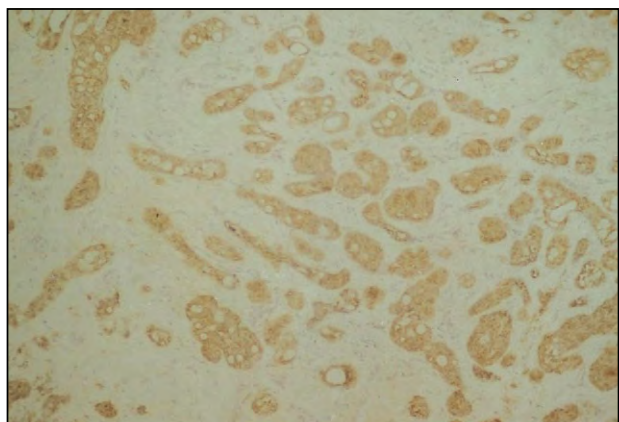


Figure 4. Cystic cavities and secretory material marked by carcinoembryonic antigen (immunoperoxidase technique)(original magnification $\times 40$).

lumens.¹⁰ Perineural invasion was observed in several described cases.¹¹ The adenoid and cystic spaces contain pale-staining mucin, which in some cases is hyaluronidase sensitive.¹² Wick and Swanson¹³ reported that microcyst content reacted with the Alcian blue staining (pH 0.5 and 2.5) and was resistant to hyaluronidase digestion.

In our patient, the tumor slowly grew. Although the average tumor size is approximately 3.2 cm², our patient's tumor reached great dimensions (12×10 cm). A detailed investigation of the oral mucosa was done to exclude primary salivary gland carcinoma. Lung and lymph node metastases were investigated through chest radiograph, abdominal and pelvic ultrasonography, and computerized tomography. The patient underwent extensive resection with 1-cm margins of normal-appearing skin as recommended¹¹ and was observed every 8 months for 3 years without any recurrence.

Conclusion

Primary adenoid cystic carcinoma is an eccrine gland carcinoma that shows slow but usually progressive growth. Lung or lymph node metastasis occurs in approximately 20% of cases and local recurrence in nearly half. It is important to make an early diagnosis and treat these patients precociously to prevent metastasis and avoid extensive resections, thus resulting in a better prognosis for patients.

REFERENCES

1. Boggio R. Letter: adenoid cystic carcinoma of scalp. *Arch Dermatol.* 1975;111:793-794.
2. Kato N, Yasukawa K, Onozuka T. Primary cutaneous adenoid cystic carcinoma with lymph node metastasis. *Am J Dermatopathol.* 1998;20:571-577.
3. Gómez Orbaneja J, Sánchez Yus E, Díaz-Flores L, et al. Adenocarcinoma of the eccrine sweat glands. clinical, histopathological, histoenzymatic and ultrastructural studies of a case [in German]. *Hautarzt.* 1973;24:197-202.
4. Sanderson KV, Batten JC. Adenoid cystic carcinoma of the scalp with pulmonary metastasis. *Proc R Soc Med.* 1975;68:649-650.
5. Seab JA, Graham JH. Primary cutaneous adenoid cystic carcinoma. *J Am Acad Dermatol.* 1987;17:113-118.
6. Ikegawa S, Saida T, Obayashi H, et al. Cisplatin combination chemotherapy in squamous cell carcinoma and adenoid cystic carcinoma of the skin. *J Dermatol.* 1989;16:227-230.
7. Pappo O, Gez E, Craciun I, et al. Growth rate analysis of lung metastases appearing 18 years after resection of cutaneous adenoid cystic carcinoma: case report and review of the literature. *Arch Pathol Lab Med.* 1992;116:76-79.
8. Zimmerman RL, Bibbo M. Fine needle aspiration diagnosis of a pulmonary metastasis from a cutaneous adenoid cystic carcinoma. a case report. *Acta Cytol.* 1998;42:367-370.
9. Chang SE, Ahn SJ, Choi JH, et al. Primary adenoid cystic carcinoma of skin with lung metastasis. *J Am Acad Dermatol.* 1999;40:640-642.
10. Fukai K, Ishii M, Kobayashi H, et al. Primary cutaneous adenoid cystic carcinoma: ultrastructural study and immunolocalization of types I, III, IV, V collagens and laminin. *J Cutan Pathol.* 1990;17:374-380.
11. van der Kwast TH, Vuzevski VD, Ramaekers F, et al. Primary cutaneous adenoid cystic carcinoma: case report, immunohistochemistry, and review of the literature. *Br J Dermatol.* 1988;118:567-577.
12. Headington JT, Teears R, Niederhuber JE, et al. Primary adenoid cystic carcinoma of the skin. *Arch Dermatol.* 1978;114:421-424.
13. Wick MR, Swanson PE. Primary adenoid cystic carcinoma of the skin. a clinical, histological, and immunocytochemical comparison with adenoid cystic carcinoma of salivary glands and adenoid basal cell carcinoma. *Am J Dermatopathol.* 1986;8:2-13.