Occurrence of 2 Primary Rare Tumors on the Scalp of a 43-Year-Old Woman

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Question

A woman in her early forties (43-year-old) presented to the Head and Neck Surgery Department of the Hospital Santa Casa in Porto Alegre, in 2010, with a normochromic papule on the scalp that she had noticed 6 months earlier. An incisional biopsy was performed. Excision for margin expansion was chosen as treatment. The histopathologic exam demonstrated margins free of the lesion. After 5 years, the patient returned with a nodule in the cervical region. An excisional cervical lymph node biopsy was performed, with positive immunohistochemistry for carcinoembryonic antigen (CEA)-M, epithelial membrane antigen (EMA), and cytokeratin AE1 + AE2 (CKM),

which confirmed metastasis of the primary tumor. The patient underwent cervical lymph node dissection and radiotherapy on the right side of the cervical region. In 2018, the patient returned with a new retroauricular lesion on the right side, which was an erythematous plaque with central erosion; at dermoscopy, there were clods and atypical and arborizing vessels (Fig. 1, 2). The histopathology of a new biopsy revealed tubulocystic structures lined by basaloid cells with syringoma-like tadpole morphology (Fig. 3, 4). The patient underwent a new surgery for margin expansion and then was referred for dermatological evaluation. Currently, she is on follow-up at the Dermatology Department of the Hospital Santa Casa in Porto Alegre.

What is your diagnosis?

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Fig. 1. Retroauricular lesion: erythematous plaque with central erosion.

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Fig. 2. Dermoscopy: erythematous plaque with central erosion, clods, and atypical and arborizing vessels.

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Fig. 3. Syringoid eccrine carcinoma extending to the reticular dermis (HE, ×10).

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Fig. 4. The tumor is composed of tubulocystic structures lined by basaloid cells with syringoma-like tadpole morphology (HE, ×40).

Answer

The histopathology of the case was compatible with syringoid eccrine carcinoma (SEC). SEC is a rare tumor, accounting for less than 0.01% of all skin cancers [1, 2]. The classification is very complex since several different terms are used to describe the same tumor. As a broad histologic spectrum is often seen, the exact origin of these lesions is unknown [3–6].

It is believed that the origin is from the eccrine glands. First described as a basal cell tumor with eccrine differentiation, in 1969, it already had several terminologies, including malignant syringoma and the original term eccrine epithelioma (basal cell tumor with eccrine differentiation) [7–11]. It usually manifests in the fifth and sixth decades of life as a nodule or slow-growing plaque on the head and neck region and, less commonly, on the trunk. The clinical presentation can be nonspecific, and the tumor usually has an indolent course [12]. The involvement of the extremities is uncommon. These tumors are highly invasive, can be locally aggressive, and often metastasize (more than 60%). Histologically, SEC exhibit syringoma-like tadpole morphology composed of basaloid cells with ductal differentiation within a fibrocollagenous matrix. Deep invasion is commonly seen. Immunohistochemically, SEC can show expression of cytokeratins, EMA, and CEA [13]. The choice of treatment is usually surgical, with margin enlargement in localized lesions. Chemotherapy and radiotherapy are performed when there is local metastasis [7–13].

In the case reported, the tumor developed in a young 43-year-old female, first on the scalp, followed by cervical

and lymph node metastases, and the treatment was excision of the entire lesion, with margins, cervical emptying, and radiotherapy. The second lesion located on the retroauricular region was confirmed as a primary lesion of SEC, which is very rare. In conclusion, SEC diagnosis can be a challenge and should be considered as a differential diagnosis in lesions with no specific pattern and dermoscopy, especially when occurring in head and neck areas. A multidisciplinary approach is important. We report the case of a young patient with 2 primary lesions of SEC to increase the awareness of this rare tumor.

Statement of Ethics

The patient was informed and gave her consent to describe the case.

Disclosure Statement

The authors declare no conflicts of interest.

Keywords

Clinicopathologic correlation · Scalp disease · Surgery · Tumor

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