Malignant Degeneration of Spiradenoma of the Eyebrow

Christopher White, DO, Kristin Witfill, DO, Richard Miller, DO
Department of Dermatology | HCA Healthcare/USF Morsani College of Medicine

Introduction

Spiradenocarcinoma is a malignancy of eccrine origin, with only 120 reported cases in the literature.[1] Elderly males and females are equally affected.[2] Benign spiradenomas along with their malignant counterpart are more frequent in patients with Brooke-Spiegler syndrome or familial eccrine spiradenoma, owing to the increased incidence of folliculosebaceous-apocrine tumors seen with these entities.[3]

Case

A 54-year-old female presented for evaluation of a tender, pink-to-violaceous nodule within her right eyebrow (Figure 1). Shave biopsy of the lesion was performed, which was histologically consistent with spiradenocarcinoma arising within a spiradenoma (Figures 2, 3).

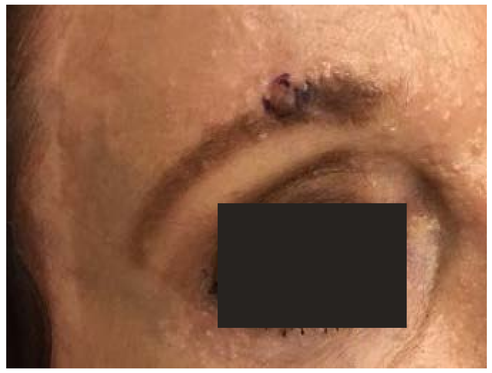


Figure 1: Pink-to-violaceous nodule within right eyebrow

The malignant lesion was excised and the patient was cancer free at her recent 9-month follow-up. With these findings, and subsequent identification of additional spiradenomas, cylindromas, and trichoepitheliomas, the patient was clinically diagnosed with Brooke-Spiegler syndrome and referred to a geneticist for further evaluation and counseling.

Discussion

Spiradenocarcinoma commonly develops within an existing spiradenoma.[4] Mutations in the *CYLD* tumor suppressor gene are known to play a role in hereditary cases, but the pathogenesis of *de novo* tumor development is poorly understood. This malignancy primarily affects the head, trunk, and extremities.[5] Tenderness, ulceration, or rapid evolution herald malignant transformation and mandate further investigation.[4]

Histopathology reveals abrupt transition from benign biphasic to malignant monomorphic cells with nuclear pleomorphism and dilated ductal and vascular structures.[4] Higher rates of recurrence, metastases, and mortality are associated with a high-grade pattern denoted by stark histologic transition from benign to carcinomatous tumor.[6] Overall, spiradenocarcinomas metastasize at a rate as high as 40%, with preferential spread to regional lymph nodes, lung, and bone.[7]

Once diagnosed, thorough imaging via computer tomography (CT), magnetic resonance imaging (MRI), positron emission tomography (PET), or a combination thereof is recommended.[1] While first line treatment has historically been wide local excision with margins >2 cm, recent literature has endorsed Mohs micrographic surgery (MMS) as a viable, and potentially superior, alternative.[7] Local recurrence rates range from 17-57%.[1,5] For unresectable or metastatic disease, multiple chemotherapy and radiation regimens have been utilized with variable success, but no formal recommendation yet exists.[7]

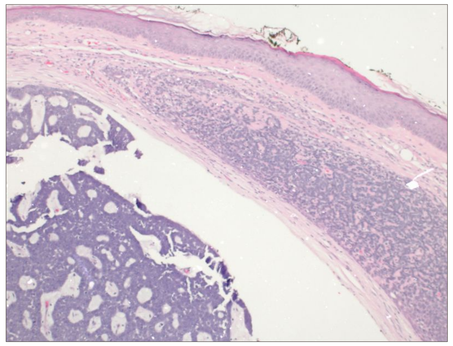
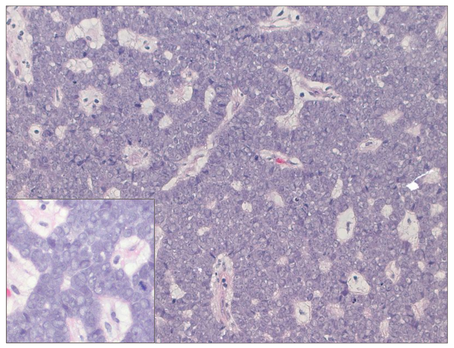


Figure 2: Abrupt transition of benign biphasic cells to compact, monomorphic malignant cells.

Figure 3: Monophasic, hyperchromatic cells with mitotic figures, dilated vascular and ductal structures, and occasional necrosis (inset).



Conclusion

Eccrine spiradenocarcinoma is a rare sweat duct malignancy that may arise via malignant degeneration of a benign tumor or *de* novo formation. Increased incidence in patients with Brooke-Spiegler syndrome or familial eccrine spiradenoma requires consideration of this tumor in the appropriate patient base. It is particularly important to identify this neoplasm early, as spiradenocarcinoma carries a 10-15% mortality rate.[8]

Following diagnosis and extirpation of spiradenocarcinoma via surgical or chemotherapeutic methods, frequent follow-up is recommended. No specific guidelines exist, but it is reasonable to assess these patients equivalently to those with previous melanoma diagnoses. Incorporation of follow-up imaging should also be strongly considered.

References

1. Staiger RD, Hemmchen B, Papet C *et al*. Spiradenocarcinoma: A comprehensive review. *Am J Dematopahtol 2017*;39:715-725.
2. Miedema JR, Burgon E, Burkhart C *et al*. Metastatic spiradenocarcinoma occurring in an 7-year-old boy. *Pediatr Dermatol* 2015;32:122-127.
3. Van Der Host MP, Brenn T. Update on Malignant Sweat Gland Tumors. *Surg Pathol Clin* 2017; 10:383-39.
4. Requena L, Kiryu H, Ackerman AB. Cylindrocarcinoma and spiradenocarcinoma. In *Neoplasms with apocrine differentiation.* Philadelphia, PA: Lippincott-Raven. 1998;733-770.
5. Granter S, Seeger K, Calonje E *et al.* Malignant eccrine spiradenoma (spiradenocarcinoma): A clinicopathologic study of 12 cases. *Am J Dermpath* 2000;22(2):97-103.
6. Ahn CS, Sangueza OP. Malignant Sweat Gland Tumors. *Hematol Oncol North Am* 2019;33:53-71.
7. Andreoli MT, Itani KM. Malignant eccrine spiradenoma: a meta-analysis of reported cases. *Am J Surg* 2011; 201:695-699.
8. Tay J, Tapen E, Solaria P. Lalignant eccrine spiradenoma: Case report and review of the literature. *Am J Clin Oncol* 1997;20(6):552-557.

This research was supported (in whole or in part) by HCA and/or an HCA affiliated entity. The views expressed in this publication represent those of the author(s) do not necessarily represent the official views of HCA or any of its affiliated entities.