Malignant Degeneration of Spiradenoma of the Eyebrow

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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).

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

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