

Title: A Case of Nevus Sebaceous of Jadassohn with Secondary Cystic Papillary Hidradenoma

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Introduction: Nevus sebaceous of Jadassohn (NS) is a congenital cutaneous hamartoma that typically presents at birth as a benign plaque, most commonly on the scalp, face, or neck. Although NS is associated with a risk of neoplastic transformation, the majority of secondary tumors that develop are benign. Papillary hidradenoma, also known as hidradenoma papilliferum (HP), is a benign growth that originates from the secretory portion of apocrine glands or anogenital mammary glands, almost exclusively in the perineal region. Ectopic HP can develop outside of this region, though this has rarely been reported within NS. In this report, we present an unusual case of NS with a secondary cystic papillary hidradenoma on the scalp.

Case Description: A 63-year-old male presented to clinic for evaluation of an enlarging nodule on the right side of his scalp. He reported having a thin, raised plaque in the area since childhood and noted recent development of a nodule within the plaque. He denied pain, pruritus, drainage, or other associated symptoms. Physical examination revealed a 1 cm, skin-colored, smooth nodule on the right temporoparietal region overlying a 2.1 cm x 1.7 cm hyperpigmented, verrucous, alopecic plaque. A shave biopsy was performed to remove the nodule. Following shave removal, a tunnel-like opening remained extending from the dermis into the subcutaneous tissue. Biopsy of the lesion was notable for papillomatosis, hyperkeratosis, and increased epidermal melanin pigmentation. There were increased sebaceous glands, ectopic apocrine glands, and anomalous duct sweat gland hyperplasia. These findings were consistent with Nevus Sebaceous of Jadassohn. Features of eccrine, apocrine, and papillary hidradenoma were all present, giving the lesion a hybrid appearance. Clinical and histological evaluation aligned with a diagnosis of Nevus Sebaceous of Jadassohn with cystic papillary hidradenoma. Total excision of the lesion was recommended. However, after discussion of the risks and benefits, the patient decided to proceed with monitoring.

Discussion: Nevus sebaceous of Jadassohn is a congenital cutaneous hamartoma of the pilosebaceous follicular unit, epidermis, and other adnexal structures. The lesion usually begins as a well-defined plaque, often with overlying alopecia. During puberty, benign enlargement occurs due to hormone-driven hyperplasia. This enlargement results in a more verrucous surface, with the epidermis becoming increasingly papillated and acanthotic. In contrast, growth of NS in adulthood raises concerns for the development of a secondary neoplasm. Hidradenoma papilliferum (HP) is a benign adnexal tumor of apocrine glands that is primarily found in the anogenital region, but rarely occurs in ectopic areas with eccrine and apocrine gland activity. Men are significantly more likely than women to have HP in an ectopic region, with most cases presenting on the head and neck. Histologically, these tumors exhibit cystic eosinophilic spaces with papillary and adenomatous structures lined by a double layer of epithelium. Malignant transformation of HP is rare, but complete excision is recommended to mitigate the risk of progression or recurrence. This case highlights a rare presentation of papillary hidradenoma, both within NS and on the scalp, a region with relatively low apocrine gland density.