Basal cell carcinoma within nevus sebaceous of the trunk

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ABSTRACT

We report the first case of an 82-year-old woman with five basal cell carcinomas within a large nevus sebaceous of the trunk. Nevus sebaceous of Jadassohn is a congenital cutaneous hamartoma affecting 0.3% of newborns. Clinical diagnosis is based on its distinctive hairless, orange-brown appearance. Up to 51% of nevus sebaceous may exhibit secondary neoplasia, and basal cell carcinoma is thought to occur in approximately 1% of lesions. Our patient presented with an asymptomatic, pigmented verrucous plaque on the upper right back that had been present since birth. Histopathology of concerning papules within the nevus sebaceous were consistent with five basal cell carcinomas and two follicular adnexal neoplasms. The patient was treated with local excisions, topical imiquimod, and close monitoring.

KEYWORDS Basal cell carcinoma; Jadassohn; nevus; sebaceous

Nevus sebaceous (NS) of Jadassohn is a congenital hamartoma of follicular origin affecting approximately 0.3% of newborns.^{1,2} NS typically presents as a waxy, hairless, orange-brown plaque most commonly involving the scalp (62.5%), face (24.5%), trunk (4.7%), or neck (4.2%).^{1,2} The pathogenesis of NS remains unknown, although *RAS* family mutations may be associated.³ More than 40 types of tumors have been documented within NS, with up to 51% of lesions exhibiting secondary neoplasia.²

CASE DESCRIPTION

An 82-year-old woman presented to the dermatology outpatient clinic for full-body skin examination. An irregularly shaped, approximately 18 cm × 8 cm, light brown verrucous plaque in a Blaschkoid pattern was identified on the right upper back (*Figure 1*). The patient stated that the lesion had been present since birth and denied symptoms. Upon dermoscopic evaluation, several pearly papules were appreciated within the large plaque. Shave biopsy of a suspicious papule within the plaque revealed nodular basal cell carcinoma (BCC) on histopathology (*Figure 2a*). Background NS was also noted on histopathology (*Figure 2b*). Additional biopsies taken within the lesion

further demonstrated nodular and superficial BCCs, trichilemmoma, and tumor of the follicular infundibulum with background NS. In total, five BCCs were diagnosed within this patient's NS, which were treated with elliptical excision and topical imiquimod. She did not demonstrate additional features of Schimmelpenning syndrome or phakomatosis pigmentokeratotic.

DISCUSSION

Trichoblastoma (34.7%), syringocystadenoma papilliferum (24.7%), apocrine adenoma (10%), and trichilemmoma (5.3%) are the most common secondary neoplasms found within NS.² The incidence of BCC in NS is about 1%, making it the most common secondary malignancy.² In one report of 65 NS tissue samples, 95% of lesions displayed *HRAS* gene mutations and 5% had a *KRAS* mutation. These oncogenic mutations may explain a predisposition to the development of secondary tumors because additional genetic “hits” occur later in life.³

Multiple tumor types may coexist in a single NS, although having more than two tumors within a single NS is uncommon. In one review of 150 patients with NS, only four patients developed three separate secondary tumors, and one patient developed four secondary tumors.⁴ Definitive

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Figure 1. Clinical photograph of nevus sebaceus.

treatment of NS is local excision to reduce the risk of cancer and for cosmesis.² Treatment guidelines are lacking, and some literature suggests close monitoring, whereas other authors encourage prophylactic excision.^{5,6} Our patient's presentation demonstrates the need for surveillance of NS due to the risk of associated secondary neoplasms.

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Figure 2. (a) Basal cell carcinoma and (b) background nevus sebaceus.

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