

Keratoacanthoma in Co-existence with Nevus Sebaceus

Seung Won Ahn, M.D., Tae Jong Chun, M.D.,
Seong Joon Seo, M.D., Chang Kwun Hong, M.D.

*Department of Dermatology, College of Medicine, Chung Ang University,
Seoul, Korea*

Nevus sebaceus of Jadassohn has been referred to as a organoid nevus that classically evolves through three stages and may be associated with a range of skin tumors, and of these, syringocystadenoma papilliferum and basal cell carcinoma have been commonly reported to occur. The occurrence of keratoacanthoma is a very rare event in nevus sebaceus.

Herein we report on a 23-year-old man with nevus sebaceus of the cheek. He represents a keratoacanthoma arising within nevus sebaceus. (*Ann Dermatol* 12(2) 138~140, 2000).

Key Words : Nevus sebaceus, Keratoacanthoma

Nevus sebaceus of Jadassohn is a hamartoma of primary epithelial germ cells. The typical solitary lesion of the head or neck appears as a yellow-tan plaque with a distinctive cobble-stoned surface that becomes most prominent during puberty. In adulthood, organoid nevi may develop secondary tumors, of which the most frequent are syringocystadenoma papilliferum (8 to 19 percent) and basal cell carcinoma (5 to 7 percent)^{1,2}. Keratoacanthomas are not commonly reported in organoid nevi.

Herein we describe a rare case of a keratoacanthoma in co-existence with nevus sebaceus.

CASE REPORT

A 23-year-old man came to our dermatology clinic for examination of a recently changing congenital lesion on his left cheek. Two weeks before his visit a small papule appeared in the center of the birthmark. During the ensuing weeks it enlarged rapidly. He had no history of seizures or other central nervous system abnormalities.

Clinical examination showed a 5.0 × 1.4 cm, poorly demarcated, slightly papillated, flesh-col-

ored plaque on his left cheek. A 6 mm shiny, dome-shaped, pink papule with a central keratin plug protruded from the center of an organoid nevus that had developed a secondary tumor.

The entire lesion was sent for histopathologic examination. Microscopic examination showed a raised, symmetric, cup-shaped tumor with a prominent keratin-filled crater flanked by buttresses of eosinophilic epidermis. Narrow, irregular strands of neoplastic keratinocytes extended from the base of the tumor into the mid-dermis. The tumor cells showed moderate nuclear atypia and focal dyskeratosis, and had abundant glassy cytoplasm. The tumor arose within an organoid nevus of an adult, characterized by mature sebaceous glands in the upper dermis overlying small apocrine glands.

DISCUSSION

Since the initial description of an "organ-nevus" by Jadassohn in 1895, this entity has been reported repeatedly in the literature. The clinical and histologic evolution of organoid nevi has been well established^{1,3}. In Korea, Choi and co-workers reported the clinical and histopathologic observations on nevus sebaceus of Jadassohn⁴. Usually present at birth, the lesions remain quiescent during childhood, become verrucous during puberty, and may develop secondary tumors in adulthood. Alessi and Sala³ explained this evolutionary process by defining "organoid" as "incompletely developed" or "still

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Reprint request to : Seung Won Ahn, M.D., 82-1, Pildong 2nd st. Chung-ku, Seoul.

Tel : (02)2260-2173, Fax : (02)2268-1049

Fig. 1. Dome-shaped nodule arising in a nevus sebaceus on the cheek.

capable of undergoing further development". This suggests that components of organoid nevi maintain the potential for future proliferation, malignant or otherwise.

Various benign and malignant neoplasms develop in organoid nevi. The frequency of malignant tumors reported in a larger series ranged from 6.5% to 22%^{2,5}. Benign tumors include trichoblastoma, syringocystadenoma papilliferum, nodular hidradenoma, apocrine cystadenoma, tumor of the follicular infundibulum, trichilemmoma, leiomyoma, and keratoacanthoma. Malignant neoplasms include basal cell, squamous cell, sebaceous, and apocrine carcinoma^{1,2,6}. Syringocystadenoma papilliferum (8 to 19 percent) and basal cell carcinoma (5 to 7 percent) are the most frequent^{1,2}, and the development of squamous cell carcinoma, apocrine carcinoma, and malignant eccrine poroma have been reported but are rare^{7,8}. We have recently experienced 6 cases of secondary developed neoplasms in organoid nevus, including syringocystadenoma papilliferum, basal cell carcinoma, squamous cell carcinoma, trichilemmoma, and keratoacanthoma⁹.

The patients' ages at the time of secondary tumor development have not been reported consistently, but malignant neoplasms generally arise in adults averaging from 38 to 63 years of age, but

Fig. 2. Hyperkeratosis and papillomatosis are present. Numerous mature sebaceous glands lie in the upper dermis (H&E, $\times 40$).

Fig. 3. Cup-shaped exophytic epithelial tumor flanked by buttresses of the epidermis that extend over the sides of the crater (H&E, $\times 40$).

keratoacanthomas have been reported in childhood^{1,7}.

Keratoacanthomas are not commonly reported in organoid nevi. Oehlschaegel and Rakoski¹⁰ reported a case of keratoacanthoma developed on a nevus sebaceus on the cheek of a 7 year-old girl. Beer et al¹¹ reported a patient had a mixture of a cystadenoma, basal cell carcinoma besides keratoacanthoma developed in nevus sebaceus. Four adult patients with keratoacanthomas were included in a series of 150 organoid nevi¹, and of the two patients with keratoacanthoma-like lesions, one was 19 years old². Three children had organoid nevi complicated by squamous cell carcinoma¹². The lesions were described as resembling verrucae and evolved rapidly, and in retrospect may be keratoacan-

thomas.

In summary, our case represents a keratoacanthoma arising within an organoid nevus in adulthood.

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