A Case of Multiple Basal Cell Epitheliomas in the Nevus Sebaceus of Jadassohn

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Nevus sebaceus of Jadassohn is a hamartoma of the skin with the potential to develop benign and malignant neoplasms.

This case was characterized by multiple basal cell epitheliomas, clinically one reddish nodule and multiple pigmented papules, arising in the nevus sebaceus. Histologically, epithelial papillomatous hyperplasia and high-positioned hyperplastic sebaceous glands were found, and tumor nests consisting of basaloid cells with peripheral palisading arrangements were mainly situated in the upper dermis without significant infiltrative growth.

We report a rare case of nevus sebaceus with multiple basal cell epitheliomas in the right cheek of a 49-year-old woman. (Ann Dermatol 9:(3) 208~210, 1997).

Key Words : Multiple basal cell epitheliomas, Nevus sebaceus of Jadassohn

Nevus sebaceus of Jadassohn (NSJ), which may be derived from the primary epithelial germ[1], is well known that it develops other appendage tumors secondarily within it[1-3]. NSJ has a three-stage evolution in its life history; infancy, pubertal stimulation, and later histological degeneration[2]. In the third histological degeneration, benign and malignant neoplasms may originate from epidermal or its adnexal structures. Basal cell epithelioma and syringocystadenoma papilliferum are the most common secondary tumors[1,2].

This case is an unusual case of multiple basal cell epitheliomas arising in NSJ.

REPORT OF A CASE

A 49-year old woman had a 5 × 2.5 cm sized, yellowish, slightly elevated, verrucous plaque on the right cheek. The lesion had developed since birth, and become conspicuous at puberty. Physical examination revealed multiple pigmented papules and one reddish nodule on the verrucous lesion (Fig. 1). Although the pigmented papules were asymptomatic and had developed at a prepubertal age, the erythematous nodule was painful and had occurred 1 year before. Each of them were separately located on the congenital nevus. There were no associated systemic abnormalies.

The verrucous plaque was totally excised. The histological examination of the yellowish plaque showed papillomatosis and acanthosis in the epidermis, hyperplasia of sebaceous glands in the upper dermis, apocrine glands and immature hair follicles in the lower dermis (Fig. 2). Histological findings of the pigmented papules showed tumor nests consisting of basaloid cells with peripheral palisading arrangements and melanin pigments (Fig. 2, 3). The reddish nodule had similar histological findings with surrounding dense inflammatory cell infiltration. All these well-circumscribed tumor nests were mainly located at the upper dermis without evidence of invasion.

No recurrence was noted 3 months after surgical

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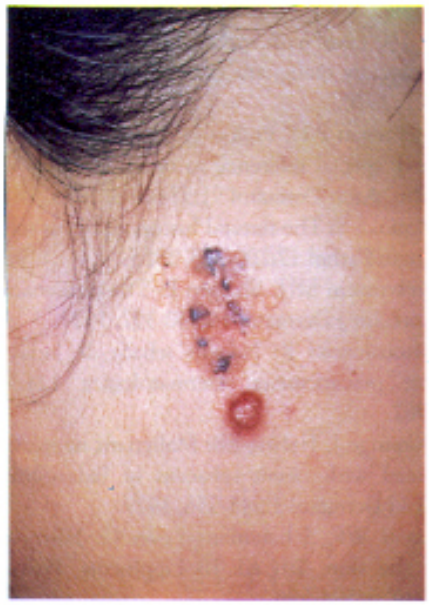

Fig. 1. Multiple pigmented papules and one reddish nodule on the yellowish verrucous plaque.

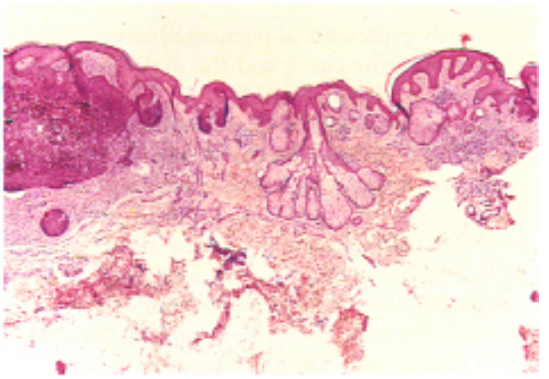

Fig. 2. Pigmented basal cell epithelioma on the left and nevus sebaceus showing papillomatosis, acanthosis, hyperplasia of sebaceous gland on the right (×20).

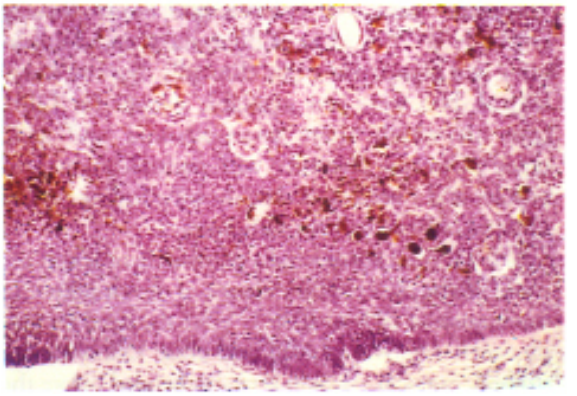

Fig. 3. Basaloid tumor cell mass with a peripheral palisade arrangement and melanin pigments (×200).

treatment.

DISCUSSION

Nevus sebaceus of Jadassohn (NSJ) is a congenital skin lesion with excesses or deficiencies of one or more of the normal mature constituents of the skin, such as hairs, glands, epidermis, or connective tissue. It is usually present at birth or early childhood and is rarely found later in life.

The lesion is most frequently found on the scalp, face, and neck and rarely on the trunk or other areas. NSJ appears clinically as a waxy, circumscribed, hairless, yellow-to-red, smooth patch in early life. The lesion becomes verrucous with the age of the patient. Histologically, NSJ includes three stages. The first infancy/childhood stage shows underdeveloped hairs and sebaceous glands. The second puberty stage is characterized by massive development of sebaceous glands with papillomatous epidermal hyperplasia and maturation of apocrine glands. The third stage is marked by complications of benign and malignant nevoid tumors[2]. Epithelial papillomatous hyperplasia, prominent subepithelial hyperplastic sebaceous glands, and apocrine glands and immature hair follicles were histologically revealed in our NSJ case.

The most common tumors arising from NSJ are basal cell epithelioma (BCE) and syringocystadenoma papilliferum. The incidence of BCE complicating nevus sebaceus vary greatly from 5% to 22%[1, 2, 4, 5]. Less commonly associated tumors include nodular hidradenoma, syringoma, sebaceus epithelioma and other types of appendageal tumors, keratoacanthoma, squamous cell carcinoma, apocrine carcinoma and osteoma[1, 2, 6]. Occasionally, the combination of a variety of tumors was found in a lesion of NSJ, which supports the pluripotential primary epithelial germ origin of NSJ.

A few important differences were noted between the BCE complicating NSJ and ordinary BCE. The BCE in NSJ frequently develops in early life[2].

Clinically it presents as pigmented lesions in more than half of the cases, and the major histological types are solid and adenoid[2].

The development of BCE in a nevus sebaceus is regarded as a decrease in the degree of differentiation, not malignant degeneration[7], and such tumors have a very benign and slow evolution. The basal cell epitheliomas in our case also showed benign behavior with a long indolent course and histologically they showed solid types and no deep dermal invasions. However, some reported cases of BCE arising from NSJ have aggressive growth patterns[1, 8]. Although the incidence of aggressive basal cell epithlioma is low, the possibility of malignancy should be considered in rapidly growing tumors.

Multiple basal cell epitheliomas rarely develop on the NSJ[8, 9, 10, 11]. Most of them show noduloulcerative and pigmented types clinically, but the histological data were not available except for Lillis' case, in which there were adenoid and solid types[10]. Our case presented with multiple pigmented papules and 1 reddish nodule showing features of solid types of basal cell epitheliomas.

NSJ has been considered a premalignant lesion because of the accepted incidence of development of carcinomas of 10% to 30%[1, 2, 5, 6]. So, the need for prophylactic removal of NSJ and close follow-up are stressed. Surgical excision is recommended before the sebaceous elements enlarge[12, 13].

We herein report an unusual case of multiple basal cell epitheliomas in NSJ.

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