A Case of Basal Cell Epithelioma Arising in a Nevus Sebaceus during Childhood

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The potential for sebaceous nevi to evolve into secondary benign or malignant tumors is well recognized. In general, malignant transformation does not occur until postpubertal life.

We experienced a case of basal cell epithelioma arising in a nevus sebaceous on a 12-year-old boy's parietal scalp.

We recommended that surgical excision should be undertaken as early as possible to detect the malignant transformation of nevus sebaceous.


Key Words: Basal cell epithelioma (BCE), Childhood, Nevus sebaceous.

It has long been known that various benign and malignant neoplasms may develop in association with nevus sebaceous. The most common benign neoplasm is reported syringocystadenoma papilliferum: the commonest of the malignant is said to be basal cell epithelioma. The incidence of BCE complicating nevus sebaceous varies greatly from 6.5% to 50%. But there have been only a few published cases of malignant adenexal tumors arising within sebaceous nevus during childhood.

We report a case of basal cell epithelioma arising in a nevus sebaceous during childhood.

REPORT OF A CASE

A 12-year-old boy was presented with his right parietal scalp lesion. The initial lesion that had been existed at birth was a waxy yellowish, hairless coin sized patch which had showly grown verrucous surfaced, 1.5 × 3.0 cm sized plaque particularly during the last 3 years. Recently the verrucous surface became more irregular, and a small nodule was palpated on the periphery of the lesion since 9 years old (Fig. 1). His past and family history was non-contributory, and there was no evidence of convulsion history and mental retardation.

Routine laboratory findings including skull X-ray were all negative or within normal limits. The skin lesion was totally removed by staged excision, and specimens were examined from the nodular site and verrucous lesion respectively. In the verrucous lesion, there were hyperkeratosis and papillomatosis in the epidermis with proliferation of sebaceous gland in the dermis that allowed us to diagnose the nevus sebaceous (Fig. 2). In the low power view of the nodular lesion, there was a tumor mass with parallel arranged connective tissue stroma bundles without connection to the epidermis. The peripheral layer of tumor mass showed a palisade arrangement and variable sized cysts were present in the center of the tumor mass (Fig. 3). Tumor cells were composed of proliferated atypical basal cell with scanty cytoplasm and round to oval nuclei (Fig. 4). Also, we could see the peritumoral cleft-like separation between the tumor mass and the connective tissue stroma. On PAS stain, the tumor mass showed a mild positive reaction.

In our case, the transformation of sebaceous nevus into BCE apparently began early in life. The nodular lesion on the scalp, which turned out to be
DISCUSSION

Nevus sebaceus was first described by Jadassohn in 1895. Later this nevi is composed of not only sebaceous glands but also all other cells and appendages of the skin, which Pinkus designated as “Organoid Nevis”®. In general, sebaceous nevi occur as circumscribed solitary lesions found in the head and neck, in contrast to verrucous epidermal nevi, which tend to be multiple and extensive in the trunk and extremities7. Sebaceous nevi can be found in about 0.3% of all neonates8. The association of extensive sebaceous nevi with other developmental defects mainly of the CNS, skeletal system and eyes, is well recognized9. Although sebaceous
nevi occur sporadically, there have been occasional reports of familial cases\textsuperscript{10,11}. Our patient was healthy except for the scalp lesion and had no family history.

Microscopically, sebaceous nevi have been described as three stages of development\textsuperscript{7}. The first or early stage in infancy characterized by papillomatous epithelial hyperplasia and underdevelopment of hairs, the second stage at puberty shows a mature lesion composed of massive hyperplastic sebaceous glands, epidermal verrucous hyperplasia and maturation of apocrine glands. In the third stage, the lesion has a tendency to develop secondary benign or malignant tumors, usually in late adulthood.

The most common malignant tumor arising from a sebaceous nevus is a basal cell carcinoma, and the incidence of this varies from 6.5\%\textsuperscript{2} to 50\%\textsuperscript{1}. But the true incidence is difficult to determine because different criteria have been used to gather cases in each of the large series\textsuperscript{7}. In addition to this difficulty, some areas of basaloid proliferation in these lesions were misinterpreted as true BCE\textsuperscript{2}. According to Lever\textsuperscript{11}, BCE develops in a sebaceous nevus not as a result of malignant degeneration, but from primary epithelial germ cells already present at birth that eventually show a decrease in the degree of differentiation, coupled with an increase in the rate of proliferation. Moroioka\textsuperscript{13} commented that basal cell epithelioma developing in nevus sebaceous had characteristics different from other cases of basal cell epithelioma: 1) Not infrequently, cases without any clinical change were revealed to be BCE histologically. 2) Many of the cases complicated with BCE also were complicated with other tumors. 3) BCE is composed of Sudan positive granule as a indication of sebaceous differentiation or glycogen positive tumor cells on PAS staining as a indication of pilar differentiation. 4) The onset of BCE associated with nevus sebaceous was found to be at a markedly younger age than that of banal BCE.

A histopathological study of the nodular lesion of our case revealed tumor cells composed of proliferated atypical basal cell with scanty cytoplasm and round to oval nuclei, and peritumoral cleft-like separation were detected, so we could rule out basaloid proliferation.

There was also a mild positive reaction on PAS stain to the tumor mass that allowed us to suspect possible pilar differentiation.

Other reported complicating malignant tumors include baso-sebaceous carcinoma, sebaceous carcinoma, basosquamous carcinoma, keratoacanthoma, proliferating trichilemmal cyst, and squamous cell carcinoma\textsuperscript{7}. On the other hand, the association of nevus sebaceous with benign epithelial tumors has been widely reported and include syringocystadenoma papilliferum most commonly; syringoma, apocrine cystadenoma, nodular hidradenoma, sebaceous epithelioma, infundibuloma, and trichilemmoma less commonly\textsuperscript{7}.

The risk of malignancy is difficult to establish with any precision, but rapid, circumscribed enlargement, ulceration or development of an exophytic nodule should arouse suspicion of malignant transformation\textsuperscript{12-14}. In spite of occasionally aggressive histopathologic features, most of these tumors are of low-grade malignancy\textsuperscript{7}. However, recurrence after local excision, metastasis and a lethal outcome have all been reported\textsuperscript{4}. Although it usually occurs in middle age, it can undoubtedly occur in adolescence, or even in childhood\textsuperscript{4}. In Korean literature, Park et al\textsuperscript{13} reported a case of nevus sebaceous associated with sebaceous epithelioma and BCE in a 48 years old male patient. Otherwise, our case in a 12-year-old boy almost fulfills the Moroioka's comment on the characteristics of BCE arising in the nevus sebaceous except there was no complication with other tumors.

We experienced a case of BCE arising in a nevus sebaceous during childhood and concluded that surgical excision is recommended for the early detection of malignant transformation in nevus sebaceous, even in childhood.

REFERENCES

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