A Case of Multiple Appendage Tumors in Nevus Sebaceus

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Nevus sebaceus is a common hamartoma that has a variety of appendage tumors. We report a case of nevus sebaceus with multiple appendage tumors on the scalp of a 43-year-old male. The exudative inflamed patches were admixed with darkly pigmented small nodules which had developed secondarily within a 4.0 × 1.5cm lesion 2 years previously. Histologically, on serial sectioning, there were various types of basal cell carcinoma, syringocystadenoma papilliferum, tubular apocrine adenoma and sebaceous adenoma. Some parts of a biopsy specimen, showed a tumor of the follicular infundibulum-like epidermal changes. There were also calcium depositions in the stroma and apocrine tumors. There was no relapse at 1-year follow-up after surgical treatment. (Ann Dermatol 10:(2) 72~76, 1998).

Key Words : Multiple appendage tumors, Nevus sebaceus

Nevus sebaceus(NS) is present at birth as a hamartoma and a variety of appendage tumors may arise with variable frequency[1-4]. Of these, syringocystadenoma papilliferum has been found in 8 to 19% and basal cell carcinoma(BCC) in 5 to 7% of the lesions of NS[2,3,5]. Less commonly found tumors include nodular hidradenoma, syringoma, sebaceous epithelioma, chondroid syringoma, trichilemmoma, proliferating trichilemmal cysts and apocrine neoplasms. The frequent association with NS and various types of appendage tumors is derived from the pluripotential primary epithelial germ to differentiate along the particular pathways.

We report herein a case of NS with multiple appendage tumors of 4 different types such as BCC, syringocystadenoma papilliferum, tubular apocrine adenoma and sebaceous adenoma. Additionally, there were findings of a tumor of the follicular infundibulum(TFI)-like epidermal changes and as well as calcium depositions in the stroma and apocrine tumors.

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CASE REPORT

A 43-year-old male visited the dermatology clinic for evaluation of newly developed exudative inflammatory lesions within a large brown hairless verrucous plaque of NS on his scalp. The shiny hairless yellowish plaque had developed at birth and became verrucous at puberty. He had felt no discomfort or problems since then. During the preceding 2 years, asymptomatic darkly pigmented nodules and erythematous nodules developed in the 4.0 × 1.5cm verrucous plaque. Recently the erythematous nodules had become erosive and had a bleeding tendency to minor trauma. Thereafter exudative inflammation admixed with darkly pigmented nodules(Fig. 1) developed gradually.

On physical examination, he was otherwise in good health with nonpalpable regional lymph nodes. A family history, past history and systemic review were non-contributory. The results of the

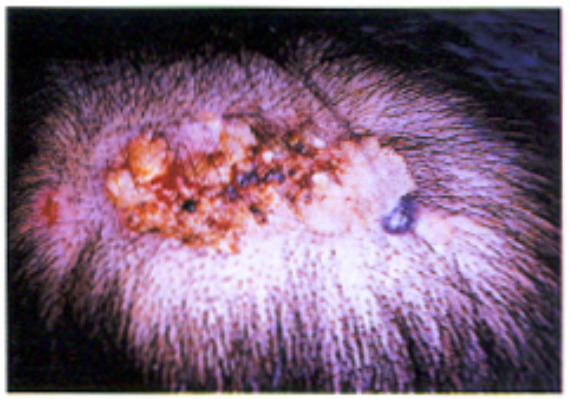

Fig. 1. Exudative inflammation admixed with darkly pigmented nodules on 4.0× 1.5cm, yellowish brown verrucous plaque.

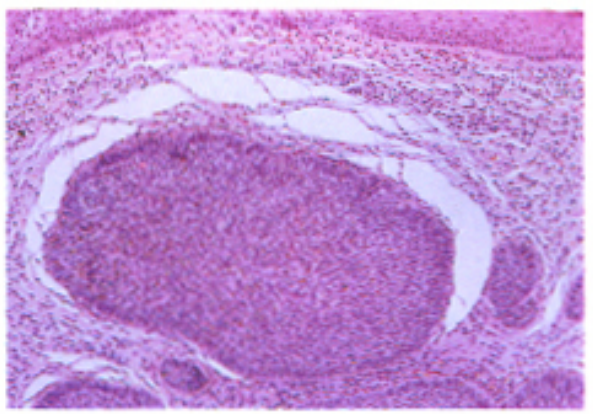

Fig. 3. Basal cell carcinoma(solid type). The mass contains large aggregates of basaloid tumor cells with a peripheral palisade arrangement(H&E, ×100).

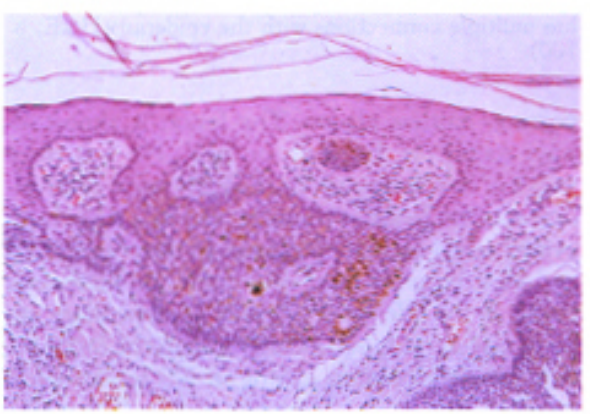

Fig. 5. Basal cell carcinoma(superficial type). Tumor tissue attached to the undersurface of the epidermis(H&E, ×100).

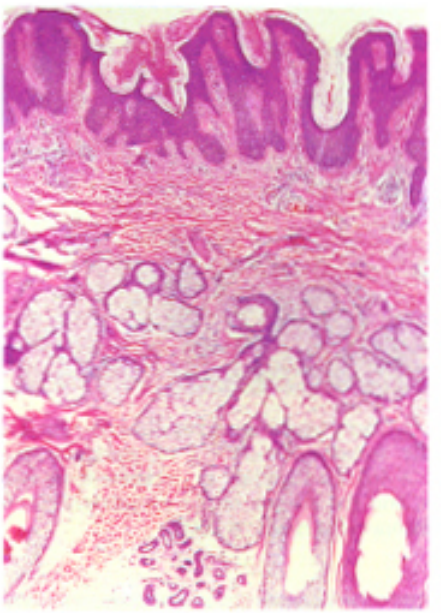

Fig. 2. Epidermal hyperplasia and mature sebaceous glands in the dermis(H&E, ×40).

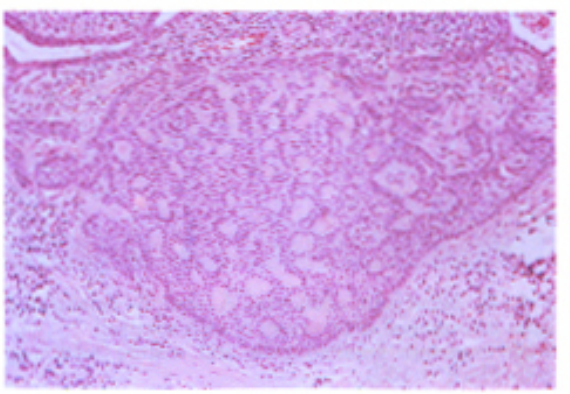

Fig. 4. Basal cell carcinoma(adenoid type). The tumor with a lacelike pattern shows gland-like structures (H&E, ×100).

following routine studies were within normal limits or non-specific: a blood cell count, urinalysis, liver function test, chest X-ray and skull X-ray.

Histopathological findings of the excised lesion on serial sectioning, showed features of NS(Fig. 2) associated with multiple appendage tumors.

Darkly pigmented nodules revealed various types of BCC, such as, solid(Fig. 3), adenoid(Fig. 4) and superficial types(Fig. 5). Erythematous erosive nodules revealed syringocystadenoma papilliferum(Fig. 6A, 6B). In the dermis, there were

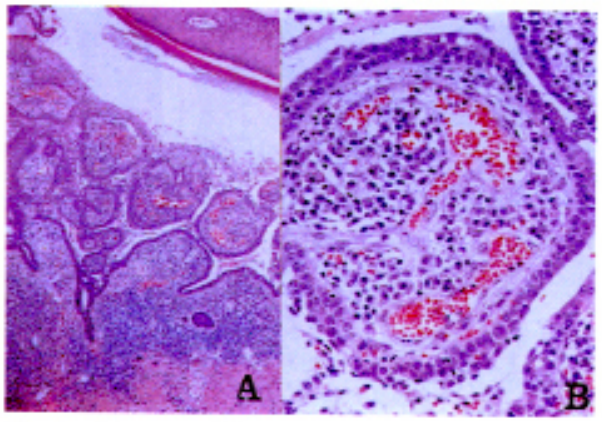

Fig. 6. Syringocystadenoma papilliferum. A : There are markedly increased numbers of glandular structures (H&E, ×40). B: Papillary projections have two layers of cells, and the stroma of the projection is infiltrated with plasma cells(H&E, ×200).

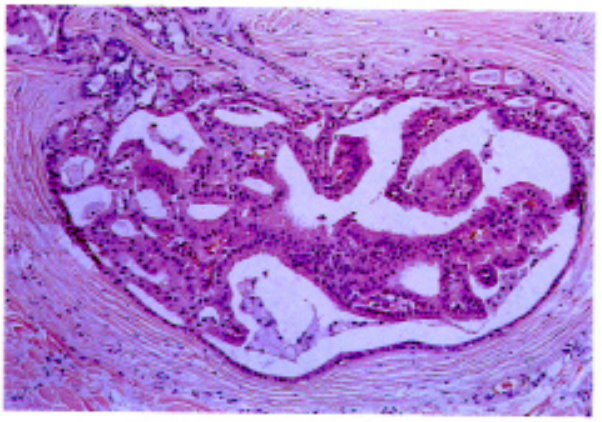

Fig. 7. Tubular apocrine adenoma. Tumor island is surrounded by increased collagen fibers and consists of two or more layers of luminal cells. Decapitation secretions are seen(H&E, ×100).

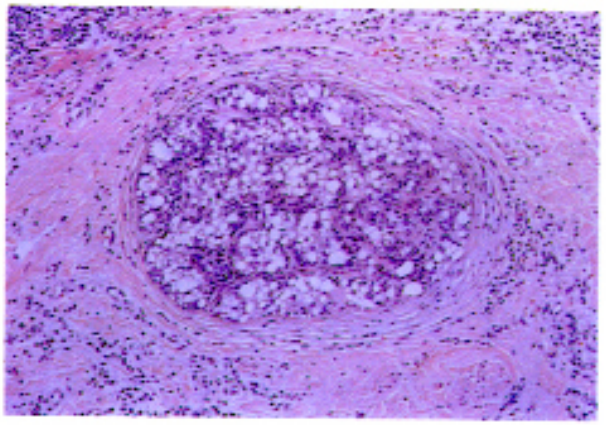

Fig. 8. Sebaceous adenoma. A sharply demarcated mass is composed of two types of cells admixed with immature basaloid cells and mature sebaceous cells(H&E, ×100).

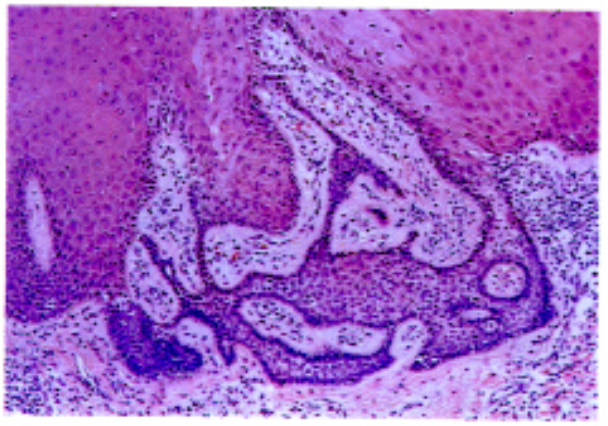

Fig. 9. Tumor of the follicular infundibulum-like epidermal changes. A plate-like growth of pale-staining epithelial cells extends parallel to the epidermis and has multiple connections with the epidermis(H&E, ×100).

histopathological findings of tubular apocrine adenoma(Fig. 7) and sebaceous adenoma(Fig. 8).

In some parts of the biosy specimen, there were findings of TFI-like epidermal changes[9](Fig. 9), which resembled TFI morphologically and showed negative staining for elastic fibers. Calcium depositions in the stroma and apocrine hyperplasia were also noted. No recurrence was noted at about 1 year after surgical treatment.

DISCUSSION

NS is a nevoid malformation present at birth or in early childhood as a yellowish brown, waxy-appearing, hairless plaque. The lesion is almost always found on the scalp or face[2]. NS has three stages of development[2]. In the early stage of development, the lesion is characterized by a slightly elevated yellowish hairless plaque. In the postpubertal period, it becomes more exuberant and verrucous. Later in the life, various types of appendage tumors may develop secondarily within the lesions.

A variety of tumors arising in NS lesions show variable frequency. Morioka[4] reported that the most common tumor is trichilemmoma(19.3%). However, as a rule, the most common benign tumor is syringocystadenoma papilliferum in 5 to 19% of

Table 1. Tumors associated with nevus sebaceus in Korean dermatology literature

Case No.	Age	Sex	Site	Associated tumors	Authors(years)
1	47	M	Scalp	SP*	Lee et al(1971)[10]
2	42	M	Scalp	Sebaceous epithelioma	Lee & Byun(1976)[11]
3	31	M	Scalp	SP + BCC**	Ahn & Jung(1977)[12]
4	25	M	Scalp	Sebaceous epithelioma	Park & Jeong(1987)[13]
5	32	M		SP	Choi & Jun(1988)[14]
6	37	F		BCC	Choi & Jun(1988)[14]
7	42	F		SP	Choi & Jun(1988)[14]
8	47	M		SP	Choi & Jun(1988)[14]
9	56	M		SP + BCC	Choi & Jun(1988)[14]
10	63	F		BCC	Choi & Jun(1988)[14]
11	26	M	Scalp	Tubular apocrine adenoma	Jang et al(1990)[15]
12	19	F	Scalp	Trichilemmoma	Lee et al(1990)[16]
13	68	M	Scalp	Sebaceous carcinoma	Hur et al(1991)[17]
14	17	F	Scalp	Cystic BCC	Oh et al(1991)[18]
15	19	M	Face	Sebaceous epithelioma	Min et al(1991)[19]
16	48	M	Scalp	Sebaceous epithelioma + BCC	Park et al(1992)[20]
17	27	F	Scalp	SP + Trichilemmoma + BCC	Jang et al(1996)[21]
18	49	F	Cheek	Basal cell epithelioma	Ha et al(1997)[22]
19	47	M	Scalp	Inverted follicular keratosis	Hong et al(1997)[23]
20	43	M	Scalp	SP + BCC + Tubular apocrine adenoma + Sebaceous adenoma	Author's case

* SP : Syringocystadenoma papilliferum ** BCC : Basal cell carcinoma.

cases[2, 3], whereas BCC is in 6 to 14% with the highest frequent malignancy[2, 3]. According to a study by Mehregan and Pinkus[2], only one type of tumor developed in 20 of 33 tumor-associated cases, two different types in 8 cases , three different types in 4 cases, and 4 different types in 1 case, but case associated with 5 or more types was not recorded. Morioka[4] had reported 5 different types of tumor arising in one NS. In a review of 19 cases(Table 1) reported in the Korean dermatology literature, our present case may be assumed to be the first reported one that developed multiple appendage tumors of 4 different types in a NS lesion.

The mechanism of development of the associated appendage tumors remains speculative. Basically, the congenital potency of NS lesions to develop tumorous proliferation may be implicated[5]. That is, a variety of tumors found in NS lesions is derived from the pluripotential primary epithelial germ in origin[5]. Inflammatory cellular infiltration caused by repeated mechanical and inflammatory stimuli may have additional influence on follicles, sebaceous glands and apocrine glands and develop various undifferentiated-differentiated types of tumors of the skin appendages[4]. In our case, the induction of multiple appendage tumors might be due to mechanical irritation.

Before the sebaceous elements enlarge, surgical excision of the lesions or prophylactic removal is recommended because of the high incidence of development of tumors[7, 8]. As the incomplete removal of the lesion may result in recurrence, close follow-up is needed. In our case, wide total excision was done, with no relapse at follow-up after one-year.

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