

Syringocystadenoma Papilliferum, Basal Cell Carcinoma and Trichilemmoma Arising from Nevus Sebaceus of Jadassohn

Sir,

Nevus sebaceus of Jadassohn has been referred to as a organoid nevus that classically evolves through several stages and may be associated with a range of skin tumors (1), and of these, syringocystadenoma papilliferum and basal cell carcinoma have commonly been reported to occur, sometimes simultaneously, within the lesion of nevus sebaceus (1-3).

We here report a rare case of nevus sebaceus associated with syringocystadenoma papilliferum, basal cell carcinoma, and trichilemmoma, exhibiting clinically a warty appearance and histologically poor development of the sebaceous glands. This case suggests that secondary tumors arise more frequently from immature nevus sebaceus than from a mature one.

CASE REPORT

A 27-year-old Korean woman had an asymptomatic large fungating mass on the vertex of her scalp. Examination revealed two tumors arising from the same yellowish plaque, which were immediately adjacent to each other (Fig. 1). The larger one, which had developed insidiously for 10 years, had cauliflower-like papillary projections on the surface of 2×3×3 cm, and the smaller one, which had been growing over the past 2 years, had the smooth and globoid appearance of a papule, measuring 0.7 cm in diameter. In the patient's history, a pea-sized hairless patch on the vertex of the scalp had appeared at birth and had been slightly elevated during puberty.

On the shave biopsy specimens, the large mass showed marked papillomatosis and acanthosis in the epidermis and clear and pale tumor islands in the dermis connected with broad bands anastomosing to the epidermis. Multiple ductal and glandular structures were also seen in the dermal stroma. The tumor consisted of ductal-like structures, showing the characteristic findings of syringocystadenoma papilliferum, which had papillary projections covered by two layers of epithelial cells with numerous stromal plasma cells. A large pale tumor component extended downward into the dermis by the broad bands, anastomosing with the overlying epidermis, and the tumor cells were mainly composed of clear cells strongly reactive to periodic acid-Schiff stain (figure not shown), which was consistent with the findings of trichilemmoma. However, the adjacent globoid papule was a basal cell carcinoma which was made up of multiple lobules of basaloid cells arranged in solid areas. The palisading arrangement of basaloid



Fig. 1. Large fungating mass and small globoid papule (arrowheads) on the vertex of the scalp.

cells in the periphery of the tumor and peritumoral lacuna was also detected in this section (Fig. 2).

The tumor mass and base were treated with electrodesiccation and curettage following the shave biopsy, and no recurrence was observed for 6 months.

DISCUSSION

Although nevus sebaceus shows the characteristic features of increased numbers of mature sebaceous glands and hair follicles on histological examination, Mehregan & Pinkus (1) and Wilson Jones & Heyl (2) reported cases with the atypical lesions of warty surfaces and with the immature lesions of diminished sebaceous glands histopathologically, and they suggested the possibility of variations in the potential for secondary tumor development between typical and warty nevi, or histologically mature and immature nevus sebaceus (3). Our case with multiple secondary tumors also exhibited a clinically warty appearance and histologically poor development of sebaceous glands.

Nevus sebaceus has been considered a premalignant lesion, with an incidence of development of carcinomas of 10 to 30%. Most have been basal cell carcinomas, but squamous cell carcinoma and sebaceous or apocrine carcinoma have been occasionally reported (3, 4). Domingo & Helwig (4) reported that 9 cases of nevus sebaceus had malignant neoplasms, including apocrine carcinoma, adnexal carcinoma, and squamous carcinoma, and 4 of them showed trichilemmal proliferation in the histologic examination.

Our rare case supports the concept of nevus sebaceus as organoid nevus with diverse differentiations of apocrine, primordial epithelium, and outer root sheath of hair, respectively.

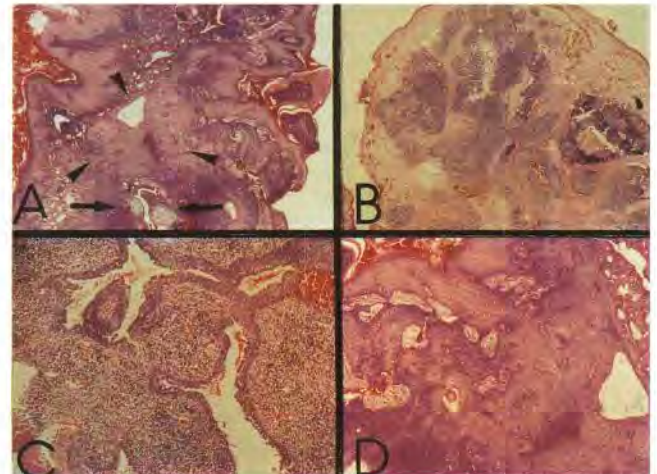


Fig. 2. (A) The large tumor shows marked papillomatosis and acanthosis in the epidermis and the portions of syringocystadenoma papilliferum (arrows) and trichilemmoma (arrowheads). (B) The histopathologic findings of basal cell carcinoma from the specimen of the small tumor (arrowheads in Fig. 1). (C, D) Characteristic findings of syringocystadenoma papilliferum and trichilemmoma are seen in serial sections, respectively (hematoxylin-eosin stain: A and B; ×20, C; ×100, D; ×40).

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