

Apocrine Adenocarcinoma and Three Different Benign Skin Tumors Probably Arising in an Organoid Nevus

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We herein present a case of simultaneous occurrence of apocrine adenocarcinoma, syringocystadenoma papilliferum, syringoma, and eccrine hydrocystoma arising in an organoid nevus (nevus sebaceus of Jadassohn) which had been present on the right occipitoparietal scalp of a 60-year-old man since birth. (*Ann Dermatol* 12(2) 122~125, 2000).

Key Words : Apocrine adenocarcinoma, Organoid nevus, Nevus sebaceus

Organoid nevus, originally described as nevus sebaceus by Jadassohn¹ in 1895, is a kind of congenital hamartoma which has the potential to develop into various types of benign and malignant skin tumors during adulthood².

The authors herein present a case of organoid nevus associated with four different types of skin appendage tumors including apocrine adenocarcinoma, a combination that is so rare that it is the first documented instance in Korea.

CASE REPORT

A 60-year-old man was seen at Chonnam University Hospital on September 12, 1984 with a polypoid mass with pus discharge and bleeding tendency, located on the right occipitoparietal area of the scalp. The patient described the existence of a darkish red hairless plaque on the right occipitoparietal area of the scalp as a birth mark. It had been stationary until about 3 years prior to the first visit, when a new nodule began to appear within the preexisting scalp lesion. During the

year of the first visit, the new lesion had grown rapidly and become inflamed.

Physical examination did not reveal any specific abnormalities except high blood pressure (160/100) and a scalp lesion. Dermatologic examination revealed an ulcerative, pus containing, protruding mass measuring 3 × 4cm in size, located on the right occipitoparietal area of the scalp. Routine laboratory test results including CBC, liver and lipid profiles, electrolytes, renal profiles, coagulation battery, fasting blood sugar, and urinalysis were all negative or within normal limits. Chest PA and EKG findings were non-specific. Anteroposterior and lateral views of skull X-rays disclosed a soft tissue mass-like density in the occipital region but no evidence of bony abnormality.

Under the clinical impression of squamous cell carcinoma, the tumor mass was removed by total excision followed by a skin graft. The surface of the tumor mass was composed entirely of syringocystadenoma papilliferum under which the areas of the well differentiated to moderately differentiated apocrine adenocarcinoma abutted on the areas of syringoma admixed with eccrine hydrocystoma (Figs. 1-4). The area of the adenocarcinoma consisted of many lobules of glandular tumors with many lumina of varying sizes and central hemorrhagic cysts (Fig. 4). The tumor cells of the well-differentiated adenocarcinoma had abundant eosinophilic cytoplasm (Fig. 5). Some foci of the luminal borders of glandular structures exhibited the evi-

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Fig. 1. Features of syringocystadenoma papilliferum (arrows), apocrine adenocarcinoma (open arrows), eccrine hydrocystoma (arrowheads), and syringoma (double arrowhead) abutting one another in a section (H&E stain, $\times 10$).

Fig. 2. The tumor surface composed entirely of syringocystadenoma papilliferum (H&E stain, $\times 40$).

Fig. 4. Several islands of apocrine adenocarcinoma with central hemorrhagic cyst (H&E stain, $\times 40$).

Fig. 3. Several eccrine hydrocystomas admixed with syringomatous lesions (H&E stain, $\times 40$).

Fig. 5. Cells of well-differentiated apocrine adenocarcinoma showing abundant eosinophilic cytoplasm and focal area of decapitation secretion (arrowhead) (H&E stain, $\times 400$).

dence of decapitation secretion (Fig. 5). The immediately adjacent skin showed low grade epidermal hyperplasia with a sparse density of somewhat underdeveloped hair follicles.

Four years after the initial excision, the patient revisited the hospital because of dyspnea and two palpable masses on the right retroauricular area. Those subcutaneous nodules were said to have begun to be palpated 2 years previously and measured 2.5×2.5 cm and 3×3 cm in size respectively. The dyspnea had begun 20 days prior to the last visit. The posteroanterior and lateral views of the chest suggested the possibility of a metastatic tumor of the left lung. The biopsy specimen taken from the right retroauricular mass showed similar but not exactly identical morphologic features with the well-differentiated areas of the previous scalp lesion 4 years ago. According to his son, the patient died 2 years thereafter.

DISCUSSION

Apocrine adenocarcinoma comprises a group of rare primary cutaneous adenocarcinomas that show features of apocrine differentiation and most frequently arise in regions of high apocrine gland density, particularly axillae, scalp, eyelids and ear^{3,5}. It arises either *de novo* or secondary to the preexisting benign skin tumors such as organoid tumor and cylindroma. The most reliable histopathologic criteria for identifying apocrine skin carcinoma appear to be decapitation secretion, PAS positive diastase resistant material in the cells and lumen, and immunoreactivity with gross cystic disease fluid protein 15⁴. The architectural pattern of adenocarcinoma, cells exhibiting abundant eosinophilic cytoplasm, and luminal borders with evidence of decapitation secretion seen in the deep dermis of this case made the diagnosis of apocrine adenocarcinoma possible. The apocrine adenocarcinoma in association with three other benign skin tumors in the current case is supposed to probably arise in the preexisting organoid nevus because of the documented history of a plaque lesion present since birth.

The third stage of organoid nevus, during adult life, is distinguished by development of a variety of tumors within the area of nevus malformation, being analogous to the frequent association of the walls of ovarian dermoid cyst with various skin tumors⁶.

Those are syringocystadenoma papilliferum, basal cell epithelioma, syringoma, infundibuloma, osteoma, apocrine cystadenoma, keratoacanthoma, nodular hidradenoma, sebaceous epithelioma, and trichilemmoma. Among them, the association rates of syringocystadenoma papilliferum and basal cell epithelioma are so high that they have been observed in 5% to 19%^{2,7}, and 5% to 7%^{7,8} respectively of the cases of organoid nevus. True malignant lesions such as squamous cell carcinoma, apocrine adenocarcinoma, porocarcinoma, sebaceous carcinoma and undifferentiated adnexal carcinoma may also occur on rare occasions. The apocrine adenocarcinomas seem to be the most frequent among them⁹. The number of its associated secondary tumors within each hamartoma ranged from one to four². The current case is a simultaneous occurrence of apocrine adenocarcinoma, syringocystadenoma papilliferum, syringoma, and eccrine hydrocystoma which is a very rare occasion.

The terms nevus sebaceus of Jadassohn and organoid nevus have been used interchangeably. However, the term organoid nevus has been preferred to nevus sebaceus by Mehregan and Pinkus², and many other dermatopathologists because the name nevus sebaceus represents the second stage only of this disease, thus leading to confusion in realizing its true nature, whereas the name organoid nevus may encompass more characteristics of this complicated disease, therefore leading to proper prognostic and therapeutic conclusions¹⁰. The authors agree with this opinion and adopted this term in this report.

Apocrine adenocarcinomas usually are associated with a nonfatal course, but it is known to cause death in rare cases⁴. Even though there is no definite evidence that in the present case, the retroauricular nodules resulted from the regional metastasis of the scalp lesion and that he died of pulmonary metastasis, those possibilities can not be absolutely ruled out. Therefore, the clinician should be alert to the signs of possible malignant changes of so-called premalignant organoid nevus, such as the sudden onset of a nodular growth within the organoid nevus and the large size of the developed nodule (>2 cm). In such cases, prompt complete excision and careful follow-up are required¹¹. The complete excision, if possible, before the third stage, is preferable to an excision during the third stage.

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