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

Sun-Wook Hwang, M.D., Hyun-Jong Kim*, M.D., Chang-Soo Park**, M.D.

Department of Dermatology, Seonam University Hospital, Kwangju, Korea,
Departments of General Surgery* and Pathology**, Chonnam University Medical School,
Kwangju, Korea

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 sebaceous

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 × 4 cm 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-
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, ×10).

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

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

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

Fig. 5. Cells of well-differentiated apocrine adenocarcinoma showing abundant eosinophilic cytoplasm and focal area of decapitation secretion (arrowhead) (H&E stain, ×400).
cence of decapitation secretion (Fig. 5). The im-
mediately adjacent skin showed low grade epidermal
hyperplasia with a sparse density of somewhat un-
derdeveloped hair follicles.

Four years after the initial excision, the patient re-
visited 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 pos-
teroanterior 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-dif-
ferentiated 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.
It arises either de novo or secondary to the preex-
isting benign skin tumors such as organoid tumor and
cylindroma. The most reliable histopathologic cri-
teria for identifying apocrine skin carcinoma ap-
pear to be decapitation secretion, PAS positive di-
astase resistant material in the cells and lumen,
and immunoreactivity with gross cystic disease fluid
protein 15

. The architectural pattern of adeno-
carcinoma, cells exhibiting abundant eosinophilic cy-
toplasm, and luminal borders with evidence of de-
capitation secretion seen in the deep dermis of
this case made the diagnosis of apocrine adenocar-
cinoma 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 doc-
umented 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 tu-
mors within the area of nevus malformation, being
alogous to the frequent association of the walls of
ovarian dermoid cyst with various skin tumors.

Those are syringocystadenoma papilliferum, basal
cell epithelioma, syringoma, infundibuloma, os-
teoma, 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 ob-
served in 5% to 19%.

, and 5% to 7%
respectively of the cases of organoid nevus. True malignant
lesions such as squamous cell carcinoma, apocrine
adenocarcinoma, porocarcinoma, sebaceous carcino-
ma and undifferentiated adnexal carcinoma may also occur on rare occasions. The apocrine
adenocarcinomas seem to be the most frequent
among them. The number of its associated sec-
ondary tumors within each hamartoma ranged
from one to four. The current case is a simultane-
ous occurrence of apocrine adenocarcinoma, syringo-
cystadenoma papilliferum, syringoma, and eccrine
hydrocystoma which is a very rare occasion.

The terms nevus sebaceous of Jadassohn and
organoid nevus have been used interchangeably.
However, the term organoid nevus has been pref-
erred to nevus sebaceous by Mehregan and Pinkus,
and many other dermatopathologists because the
name nevus sebaceous 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 conclusions.

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 cases.

. 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(>2cm). In such cases, prompt complete
excision and careful follow-up are required.

. The complete excision, if possible, before the third
stage, is preferable to an excision during the third
stage.
REFERENCES


