Extraocular Sebaceous Carcinoma
—A Case report—

Sang Yoon Kim, Je G. Chi, and Yoo Shin Lee*

Departments of Pathology and Dermatology*, College of Medicine, Seoul National University

Abstract—A case of sebaceous carcinoma in a 40 years old male occurring in frontal scalp is described. This tumor was very unique in gross appearance showing surface protruding yellow mass associated with ulceration and was histologically characterized by basal cells, squamous cells and cells with clear cytoplasm that showed strong ORO positivity and maturation gradient found in the center of each lobule of the tumor.

Key Words: Sebaceous carcinoma, Extraocular sebaceous carcinoma, Skin adnexal tumor

Extraocular sebaceous carcinoma is a very rare tumor and is morphologically less well defined when compared to ocular sebaceous carcinoma. The correct diagnosis is very important because histologically this tumor can mimic many different types of adnexal or epidermal tumors. Recently we had a chance to examine a tumor of the scalp in an adult, which showed histologically very unique features that could best be attributed to sebaceous carcinoma.

REPORT OF CASE

This 40 years old male was admitted to the Seoul National University Hospital because of a large mass on the right frontal scalp. The parents recalled a small alopecic patch in this area at the time of birth, which subsequently developed into a small verrucous papule and remained so for 10 years. Recently it seemed to grow rapidly, being accompanied by pruritus and pricking pain on touch. On examination it was a well-defined lobulated, walnut-sized, protruding mass which bled easily and had surface ulceration. All laboratory findings were within normal limits. The skull X-ray films showed no bony erosion or periosteal reaction under this lesion. No palpable lymph node was noted in the cervical area. A punch biopsy was done initially at the margin of the lesion, which revealed two components. One was irregular small nests of large polygonal cells which had plump eosinophilic cytoplasm, hyperchromatic large nuclei and prominent nucleoli without definite intercellular bridge or keratin pearl. The other revealed well circumscribed nests of basaloide cells with peripheral palisading and scattered microcystic areas. The pathologic diagnosis was basal squamous cell epithelioma at that time.

Fifty-five days after the first diagnostic biopsy a wide excision with skin graft was performed. The polypoid lesion measured 3 x 2.5 cm in maximum cross and 1.7 cm in height. The outer surface was shiny and yellowish tan with focal reddish crusted areas. The cut surface showed sharply delineated margin between the tumor and normal dermis. The tumor was composed of bright yellow tan compact small lobules which had tiny central cystic areas (Fig. 1). The entire tumor was blocked and stained with hematoxylin-eosin and periodic acid-Schiff reagent. Additionally oil-red-O staining (ORO) on frozen sections of formalin fixed specimen was done. Microscopically, the tumor consisted of many variable sized lobules of relatively monotonous basaloide cells with peripheral palisading. From the margin to the center of each lobule, the tumor cells were increased in size and showed increasing number of cytoplasmic vacuoles (Fig. 2). Most of the lobules had central microcystic portion containing pinkish amorphous material, bubbly histiocytes or cell debris. Abortive hair follicles were also seen in some lobules near the center of lobules. Except the lobules of basaloide cells, small nests of large pleomorphic cells were aggregated in some portions, especially in deeper area (Fig. 3). These pleomorphic cells were similar to the squamous cell com-
ponent of previous biopsy and showed blurred margins with individual cells in fibrous stroma. A few keratin pearl-like materials were also seen in focal areas. PAS stain revealed a few weakly positive granules in microcystic areas, but no positive granule was seen in the cytoplasm of any tumor cell in spite of strong positive reaction in hair sheath cells in the same section. Oil-red-O stain revealed bright red droplets which increased in size and number from the margin toward the center of each lobule (Fig. 4). These vacuoles were also noted in the small nests of the more undifferentiated tumor.

The patient is well and free of recurrence 10 months after resection.

DISCUSSION

The extraocular sebaceous malignancy is of a very rare occurrence and most reports are based on only a few cases. This neoplasm is difficult to make a diagnosis because many have failed to distinguish pure sebaceous carcinoma from basal or squamous cell carcinoma with focal sebaceous differentiation. Sebaceous carcinomas of the eyelid were documented and have much more aggressive behavior than their extracocular counterparts. Hernandez-Perez and Banos (1978) reported two cases of extraocular sebaceous carcinoma with lymph node metastasis and King et al. (1979) reported a case with visceral metastasis. This neoplasm may be confused with tumors composed of basal cells, squamous cells, clear cells and balloon nevus cells as well as other sebaceous neoplasm. However, fat stain of frozen section would reveal clear sebaceous differentiation and marked cytologic atypia with stromal, vascular or perineural invasion could make differentiation from other sebaceous neoplasm.

In this case, vascular or perineural invasion was not found. However, cytologic atypia and stromal invasion were definite. Clinically and grossly, this case resembled the proliferating trichilemmal tumor or giant hair matrix tumor. But intracytoplasmic glycogen granule was not evident. Microscopically, there were some abrupt keratin pearl formations and areas of tumor cells with plump eosinophilic cytoplasm. But the areas accounted for only a small portion of the tumor, and ORO stain revealed positive reaction around these areas, again indicating that this tumor is sebaceous carcinoma.

REFERENCES


안외 피지암종
-1증례 보고-

서울대학교 의과대학 방리학교실 및 외부과학학교실*
김상윤·자체근·이유신*

40세 남자의 우측 전두부의 두피에 생긴 피지암종을 보고한다. 이 병변은 뒷뇌년 때부터 발생하여 있던 부위가 10년전에 자라뉘 같은 구긴으로 변하고 다시 최근에 둔져진 종괴로 되었는데, 쉽게 충혈하는 경향을 보였다고 한다. 육안적으로 종괴는 크기 가 3×2.5×1.7cm이었고, 아래쪽의 진피와 두꺼이가 구분되었으며, 밝은 황색의 아주 작은 소열들로 구성되어 있었다. 현미경 소견으로, 이 종양의 대부분은 기저세포양 세포로 이루어진 소열들로 이루어져 있었는데, 각 소열의 중심부분은 세포가 없는 낡은변화를 보이는 것이 많았고, 각 소열의 주변부에서 중심부로 점수록 세포질에 종괴가 증가하는 것을 보여 주었는데, 이 종괴들은 지방질에 대한 특수형식약 양성반응을 보였다. 그리고 간피존으로는 천장상피세포양의 세포 들이 작은 소(nests)를 이루고 있었는데, 주위의조직을 침습하고 있는 형태를 보였다. 안외 피지암종은 안경에 생긴 피지암종과 조직소견은 유사하나, 예후가 아주 좋으므로 단리 취급되고 있으며, 그 예가 극히 드문이 주의를 깨닫히 필요하다고 생각되어 본 예를 보고하는 바이다.

LEGENDS FOR PLATES

Plate 1. Gross specimen (cut section). Yellowish protruding lesion is quite characteristic with its solid lobulated and sharply delineated margin.

Plate 2. Low power photomicograph showing lobules of monotonous basloid cells with peripheral palisading. H&E, X40.

Plate 3. Pleomorphic squamoid cells are aggregated with abundant clear cytoplasm which is weakly positive for PAS. PAS, X320.

Plate 4. Oil-red-O stain of the tumor, showing strongly positive hair sheath cells. The fat droplets increase in size and number from the margin toward the center of a lobule. ORO, X320.