Extraocular Sebaceous Carcinoma

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Sebaceous carcinoma is an uncommon neoplasm usually associated with ocular adnexa. This malignancy may occur at any anatomic site that contains sebaceous glands. Despite the widespread anatomic distribution of sebaceous glands, extraocular sebaceous carcinoma occurs with far less frequency. We report a case of an 87-year-old Korean woman with extraocular sebaceous carcinoma treated with total excision. (Ann Dermatol 16(1) 13~15, 2004)

Key Words: Sebaceous carcinoma, Extraocular

INTRODUCTION

Sebaceous carcinomas are rare malignant appendageal tumors[1] usually associated with ocular adnexa, without definite clinical feature. They are classified into two main variants on the basis of their location, ocular and extraocular sebaceous carcinomas. About 50% of ocular sebaceous carcinoma arises from meibomian glands[2], with a marked preference for the upper eyelid[3]. Extraocular sebaceous carcinoma mostly involves the head and neck region, it may occur at any anatomic site that contains sebaceous glands or the potential for their development, including trunk, extremities, and external genitalia[4].

Despite the widespread distribution of sebaceous glands, extraocular sebaceous carcinoma occurs with far less frequency[5]. Herein we report a case of extraocular sebaceous carcinoma that occurred without primary lesion on the face of an 87-year-old Korean woman with a review of literature.

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CASE REPORT

An 87-year-old Korean woman was referred to our department for evaluation of a slowly-growing mass on her face. The patient had been aware of the mass for the previous 3 years. The lesion had gradually increased its size to 1.0 × 1.5 cm, yellowish dome-shaped nodule with no subjective symptom (Fig. 1). There was no history of any significant medical problem with her.

A 3 mm-punch biopsy specimen was obtained from the lesion. The tumor was non-encapsulated but relatively circumscribed and consisted of variably sized lobules of neoplastic cells (Fig. 2). The peripheral areas of the tumor lobules were composed of

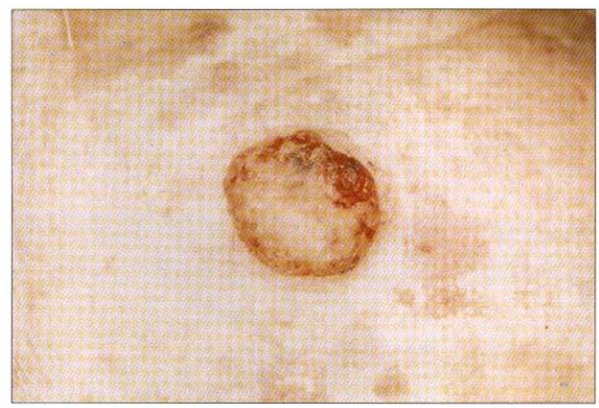

Fig. 1. Asymptomatic solitary, 1×1.5 cm sized, pinkish dome-shaped nodule on the left cheek.

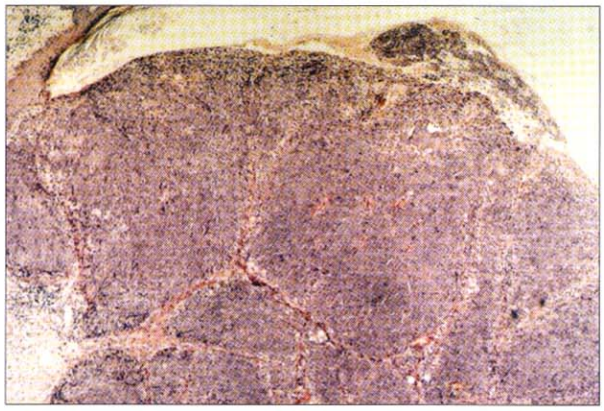

Fig. 2. Well-demarcated, irregular, variable sized, lobular tumor nests are seen (H&E, ×40).

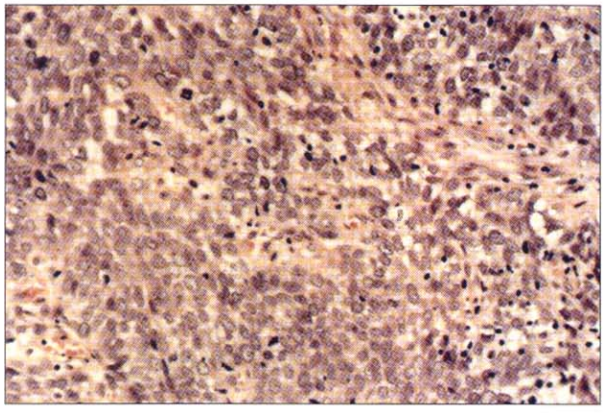

Fig. 3. Tumor lobules are composed of pleomorphic, foamy-cytoplasmic, and atypical cells with mitotic figure (H&E, × 200).

undifferentiated cells with eosinophilic cytoplasm. They appeared atypical with malignant cytologic features including nuclear pleomorphism and hyperchromatism (Fig. 3). Special staining with Oil-red-O was positive, and immunohistochemical staining with CEA and cytokeratin showed no response.

She underwent total excision of the lesion and no tumor cell was found at the margin of resection. No recurrence was observed during 6 months after the excision.

DISCUSSION

Sebaceous carcinoma occurs most frequently in the sixth and seventh decades of life, and is reported to occur in women more often than men. There is a higher frequency in Asian population, but no cause for this has been documented[6]. The clinical presentations of sebaceous carcinoma are diverse and often elude diagnosis for months to years. The most common presentation is a non-specific, slow-growing, firm, nodular mass ranging from pink to red in color that may bleed spontaneously[7,8].

The histopathologic examination of sebaceous carcinoma reveals irregular lobular formations showing great variations in size of the lobules. Although many cells are undifferentiated, distinct sebaceous cells with a foamy cytoplasm are present in the center of most lobules. Many undifferentiated cells and sebaceous cells appear atypical, showing considerable variation in the size and shape of their nuclei[9]. Special stains such as Oil-red-O or Sudan IV are helpful for the diagnosis of sebaceous carcinoma, and fresh tissue is often necessary to prevent lipid dissolution with alcohol in routine hematoxylin-eosin-stained sections[10]. Immunohistochemical stains such as EMA or LeuM1 can be used for the diagnosis, and electron microscopic examination showing intracytoplasmic lipid droplets in neoplastic cells also may be helpful[10]. In our study, Oil-red-O stains showed intracellular lipid deposition in tumor cells, and immunohistochemical staining with CEA and cytokeratin showed no response.

A differential diagnosis for sebaceous carcinoma would encompass other primary cutaneous malignant neoplasms, including balloon cell melanoma, clear cell variants of squamous cell carcinoma, basal cell, and eccrine carcinomas[11]. These neoplasms composed of clear cells show no evidence of sebaceous differentiation, and neoplastic cells exhibit a signet-ring cell appearance with the nuclei positioned eccentrically due to glycogen accumulation within their cytoplasm. Metastases to the skin from malignant neoplasms composed of clear cells, namely, renal, breast, bladder, and prostatic carcinoma or melanoma, also may be sebaceous carcinoma. Histochemical methods (PAS, mucicarmin, etc.) and immunohistochemical investigations (PSA for prostatic carcinoma, S-100 and HMB-45 for melanoma) may be useful for the differential diagnosis[12].

Sebaceous carcinoma may be seen as a component of the Muir-Torre syndrome, which is characterized by sebaceous neoplasm manifesting in conjunction with underlying, often multiple, visceral malignancies (mainly colorectal, genitourinary, breast, and

hematologic) and occasionally keratoacanthomas [13,14]. In our patient there was no familial history of hereditary cancers or sebaceous neoplasms and no other malignancies were discovered. Therefore, a diagnosis of Muir-Torre syndrome may be ruled out.

The treatment of choice for sebaceous carcinoma is surgical intervention[12]. Great care must be taken in primary treatment to ensure complete tumor excision with proven free margins of resection. Radiotherapy is used if metastatic disease or a high risk of recurrence is present[3]. Multi-agent chemotherapy has been used to treat recurrent disease[15]. Combined treatment with isotretinoin and interferon-alpha appears to be successful in cancer development in patients with Muir-Torre syndrome[16].

Ocular type of sebaceous carcinoma is aggressive and has a tendency to recur locally after local excision and Mohs microsurgery. Also, there may be orbital invasion, and in 22% of cases reported in one study, death resulted from visceral metastasis[17]. In contrast to ocular type, extraocular sebaceous carcinoma has been considered as a locally invasive malignant neoplasm that only rarely metastasizes, with a far better prognosis than that of ocular adnexa. Some investigators, however, have argued that in their reported cases of extraocular sebaceous carcinoma the clinical aggressiveness was similar to that of ocular sebaceous carcinoma[18].

In summary, we report a case of extraocular sebaceous carcinoma that occurred on the face of an 87-year-old Korean woman treated with total excision.

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