

EXTRAOCULAR SEBACEOUS CARCINOMA ARISING FROM NEVUS SEBACEOUS

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Abstract- Sebaceous carcinoma is a relatively rare cutaneous tumor that is most commonly derived from the sebaceous glands located in periorbital area. The extraocular occurrence of this tumor is very rare. We report a case of an 85 years old woman who presented with a slowly enlarging mass on the right temporal region. Histological examination of the lesion revealed a malignant sebaceous carcinoma occurring in conjunction with nevus sebaceus. The lesion was surgically excised completely.

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INTRODUCTION

Sebaceous carcinoma is a rare malignant tumor of the skin and is most commonly found on eyelids but rare cases may arise at other sites. It derives from epithelium of sebaceous glands. This tumor has a high incidence of local recurrence and regional metastasis. Lesions appear as asymptomatic nonencapsulated nodules, usually on the upper eyelids. The diagnosis of this tumor can be difficult and it may mimic ruptured cyst, conjunctivitis, basal cell carcinoma (BCC) or other adnexal tumors (1-6).

CASE REPORT

An 85-year-old woman presented with a long history of an enlarging mass on the right temporal region (Fig. 1). She had first noted the lesion in childhood and slowly enlarged. In the past 3 years, the patient noted a faster increase in size that was associated with ulceration of the lesion. Physical

examination revealed a 5×5 cm firm tumor with telangiectatic blood vessels.

There was no regional lymphadenopathy and general examination including abdominal examination was normal. A definite clinical diagnosis was not possible and an incisional biopsy was performed. Microscopic examination of the lesion showed an unencapsulated dermal neoplasm characterized by pleomorphic epithelial cells with amphophilic to vacuolated cytoplasm, poorly defined cell borders and oval vesicular nuclei. Occasional abnormal mitotic figures were seen. These findings were compatible with sebaceous carcinoma (Fig. 2).

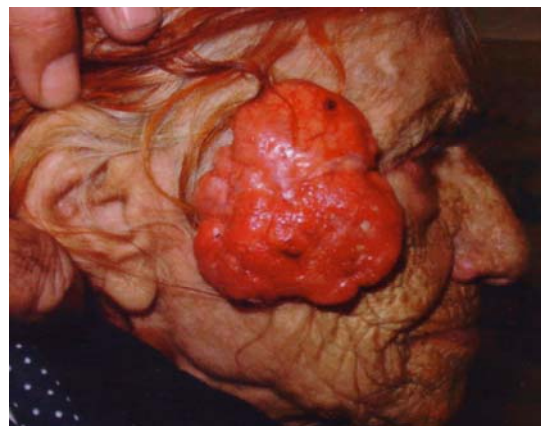


Fig. 1. Sebaceous carcinoma on the right temporal region.

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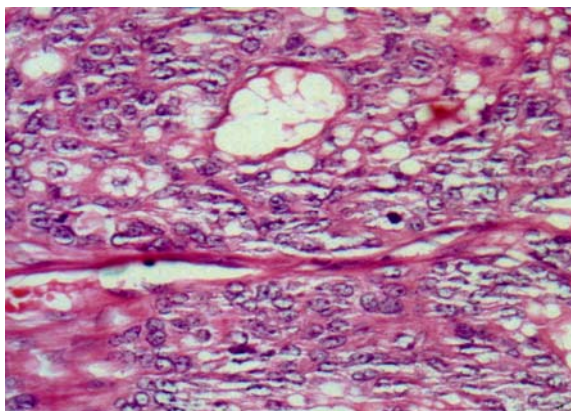


Fig. 2. Microscopic examination of the lesion.

The overlying cutaneous epithelium displayed papillomatous hyperplasia with prominent sebaceous glands, as well as eccrine and apocrine glands and rudimentary hair follicle (Fig. 3), therefore a diagnosis of underlying nevus sebaceous was established.

Histologically, the margins of resection were clear of tumor. The patient made good postoperative recovery with good healing.

The patient was therefore submitted to instrumental examination (chest X-ray, complete ultrasound examination of the abdomen and occult blood test) to evaluate the possible existence of metastasis and to exclude the existence of carcinoma in other sites as in Muir-Torre syndrome, all with a negative result. She was treated by surgical excision of the lesion with a 1 cm clinical margin.

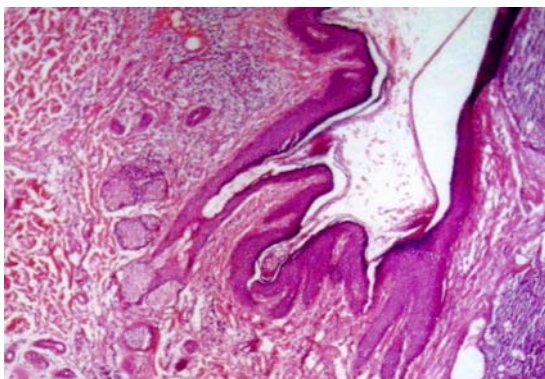


Fig. 3. Microscopic examination of nevus sebaceous, showing papillomatous hyperplasia as well as some sebaceous glands and a rudimentary hair follicle.

DISCUSSION

Sebaceous carcinoma of the skin, especially the extraocular type is a very rare tumor. It occurs most frequently on the eyelids, where they originate usually from the Meibomian glands and less commonly from the glands of Zeis. However they may occur in any part of body that contains sebaceous glands, including skin, major salivary glands, larynx and pharynx. Extraocular sebaceous carcinoma (EOSC) accounts for approximately 25% of all sebaceous carcinoma. The common site of EOSC is head and neck, with parotid glands being the most common location, followed by nose, face and neck (1-3). EOSC in external genitalia, external auditory canal, trunk and upper extremity has also been reported (7). It usually presents as a painless enlarging mass ranging from pink to red in color that may bleed spontaneously (1, 2). The age distribution for EOSC is similar to that of ocular sebaceous carcinoma, with a mean age of 63 years. In contrast to female predilection for ocular lesions, EOSC shows a nearly equal gender distribution (4). EOSC may cause regional metastasis but it is very uncommon and seems to have a better prognosis than ocular sebaceous carcinoma (2).

Histopathologically, sebaceous carcinomas are often poorly differentiated neoplasms present mainly within the dermis. Multiple lobules of basaloid undifferentiated cells are present within dermis. In the central portion of the lobules, more mature sebocytes are present. There is marked nuclear atypia and pleomorphism, and mitoses are common (1, 8). Sebaceous carcinomas may involve the epidermis and there may even be pagetoid intraepidermal spread of tumor cells. Pagetoid involvement is particularly noted in sebaceous carcinoma of the eyelids and rarely present in EOSC (9). The cause of most cases of sebaceous carcinomas is unknown. It may be associated with Muir-Torre syndrome, an autosomal dominant disorder with many sporadic cases, which is characterized by sebaceous neoplasms manifesting in conjunction with underlying, often multiple, visceral malignancies and occasionally keratoacanthomas.

The most frequent location for sebaceous carcinoma related to Muir-Torre syndrome is ocular

area, and risk for metastasis from sebaceous carcinoma in these patients seems to be lower than in sebaceous carcinoma not associated with Muir-Torre syndrome (10, 11). In our patient there was no family history of hereditary cancers or sebaceous neoplasms and no other malignancies were discovered. Therefore, a diagnosis of Muir-Torre syndrome may be ruled out.

Nevus sebaceous is a rare cutaneous malformation that is often seen at birth or arises in infancy, producing a slowly enlarging plaque. This lesion occurs almost always on the scalp and face, areas rich in sebaceous glands. The histologic findings include epidermal acanthosis and surface irregularity, sebaceous gland hyperplasia or hypoplasia with alteration in maturation, and structural anomalies and numerical reduction of the sweat glands and hair follicles. Nevus sebaceous is often associated with the development of benign and malignant neoplasms such as BCC and apocrine carcinoma, but the occurrence of sebaceous carcinoma in association with this nevus, as in our case, is extremely rare (12). The treatment of choice for sebaceous carcinoma is surgical, with complete excision verified by negative margins. Radiotherapy is used if metastatic disease and/or a high risk of recurrence is present. Multiagent chemotherapy has been used to treat recurrent disease (1, 13).

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