Case Report

Extra-ocular sebaceous carcinoma: Rare skin adnexal tumor

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ABSTRACT

Sebaceous carcinoma is a rare, highly malignant, and potentially lethal tumor of the skin, which most commonly occurs in the eyelid. The neoplasm arises from sebaceous glands, such as those of the eyebrow. It represents 1-5.5% of eyelid malignancies and is considered to be the third most common eyelid malignancy after basal cell carcinoma and squamous cell carcinoma, although few reports placed this tumor as second most common after basal cell carcinoma. Cutaneous extra-ocular sebaceous carcinoma is a rare tumor that frequently occurs on the face and scalp, constituting about 25% of all sebaceous carcinoma. It usually affects elderly women and characterized by the high rate of local recurrence, regional and distant metastases. We describe a case of sebaceous carcinoma of the left cheek in an 84-year-old male.

Key words: Cheek, extraocular type, sebaceous carcinoma

INTRODUCTION

Sebaceous carcinoma is a rare adnexal tumor that follows an aggressive course, with a risk of local recurrence and distant metastases. Its etiology is unknown. Three-quarters of cases occur in the perioccular region, although the tumor can present on any part of the body.[1] Sebaceous gland tumor of the eyelids may arise from the meibomian glands, glands of Zeis or glands associated with the caruncle. Sebaceous gland carcinoma might be the second most common lid malignancy after basal cell carcinoma. Its multifocal origin and pagetoid spread gives it an unique place among eyelid malignancies.[2]

Sebaceous carcinoma may resemble an inflammatory disease or another tumor, leading to delay in diagnosis, inappropriate treatments, and increasing morbidity and mortality. It is also one of the cutaneous markers of Muir-Torre syndrome (MTS). Therefore, patients with sebaceous carcinoma should be carefully assessed by taking an extensive personal and family oncologic history, performing a detailed physical examination, and ordering any additional tests with close follow-up.[1]

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CASE REPORT

We present a case of 84-year-old man who presented with an ulcer over the left cheek since 2 years. The lesion started initially as a small, slow growing, firm, deep seated mass over left cheek 2 years ago. The lesion was painless which gradually increased in size and resulted in a nodule of size about 2 cm. It got eroded and evolved into an ulcer with a tendency to bleed. No palpable regional lymph nodes were detected. A provisional clinical diagnosis of basal cell carcinoma was made. The tumor was excised and sent to the Department of Pathology for the histopathological examination.

Skin covered soft to firm tissue piece measuring 2.5 × 1.5 × 0.5 cm was received. The skin was ulcerated in the center, which showed a grey white area measuring 1 × 0.8 × 0.2 cm underlying it on the cross section. The whole tissue was processed with relevant sections from the base and excised margings.

The microsections examined showed a nodular lesion with ulcerated epithelium covering an unencapsulated

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neoplasm in the dermis composed of lobules of varying sizes. Lobules showed larger oval cells of varying sizes with clear or pale, bubbly cytoplasm. The nuclei were centrally located, showed nuclear grooves, clear chromatin, and prominent nucleoli [Figure 1]. They also showed moderate degree of atypia and occasional mitotic figures. Diagnosis of sebaceous neoplasm was made. Immunohistochemical (IHC) staining showed patchy positivity for epithelial membrane antigen (EMA) [Figure 2a], negative for bcl-2 and Ki-67 proliferation index of >5% [Figure 2b], which helped us to come at the conclusion of sebaceous carcinoma.

DISCUSSION

Ocular sebaceous carcinoma is a rare adnexal tumor of unknown etiology characterized by its aggressive course. It can present in any part of the body where sebaceous glands are common. Three-quarters of cases occur in the perioccular region, particularly on the eyelids.[1] The tumor is very rare, slow growing, and commonly found in the elderly population with female predisposition. Mean age at diagnosis is mid-sixties; however, the tumor has been reported in children as young as 3.5 years old. It is rare in Caucasians and common in oriental Asiatics with incidence of 0.5-5% in USA and 28% in China.[2]

Extra-ocular sebaceous carcinoma constitutes ¼ of all cases of sebaceous carcinoma that has a potential for aggressive behavior. Extra-ocular sebaceous carcinoma commonly occurs in the head and neck region. Males are twice more commonly affected than females and patients are usually in the 7th decade with rare cases reported in childhood.[3] Only fewer than 120 cases of sebaceous carcinoma previously reported; and approximately 75% involved the head and neck region, since it contains the highest density of sebaceous glands in the body.[4]

Clinically, sebaceous carcinoma presents as an occasionally pedunculated subcutaneous pinkish red nodule or mass of firm consistency with a tendency for ulceration and spontaneous bleeding in one-third of cases. In general, these tumors are painless.[1] Sebaceous carcinoma is one of the great masqueraders - it can mimic other, more benign lesions. It can mimic chalazia, chronic blepharitis, basal cell carcinoma, squamous cell carcinoma, superior limbic keratoconjunctivitis, and ocular cicatricial pemphigoid.[5]

There is also a rare syndrome called Muir-Torre of visceral neoplasms associated with sebaceous carcinoma on the skin, which is an autosomal dominant condition.[6] The diagnosis of MTS requires the presence of a sebaceous neoplasm in combination with at least one internal malignancy. The skin lesions are most commonly sebaceous adenomas, but sebaceous epitheliomas and carcinomas also occur in MTS.[7] The sebaceous carcinomas in MTS are less aggressive and less likely to metastasize than solitary sebaceous carcinoma.[8] The second key feature of MTS is internal malignancy. While most patients have only 1 malignancy, 2 or 3 different malignancies can be seen in 37% of the cases. The most common visceral neoplasm is colorectal carcinoma (51%) with others including carcinomas of the genitourinary tract (including bladder, renal pelvis, ovary, and uterus), breast carcinomas, and hematologic, head and neck, and small intestinal malignancies.[7]

Histologically, sebaceous carcinoma can be well, moderately, or poorly differentiated. In well-differentiated lesions, there is lobular arrangement and vacuolization of the cytoplasm. Cells present will include sebaceous and undifferentiated cells.[9] Tumor cells have hyperchromatic nuclei of variable shapes and sizes, with high mitotic activity and basaloid characteristics. Less well-differentiated cells contain a lipid-rich eosinophilic cytoplasm, giving a foamy appearance. The presence of sebaceous differentiation can be shown by staining with Oil Red O or Sudan IV, or with IHC markers such as EMA and Leu-M1.[1]

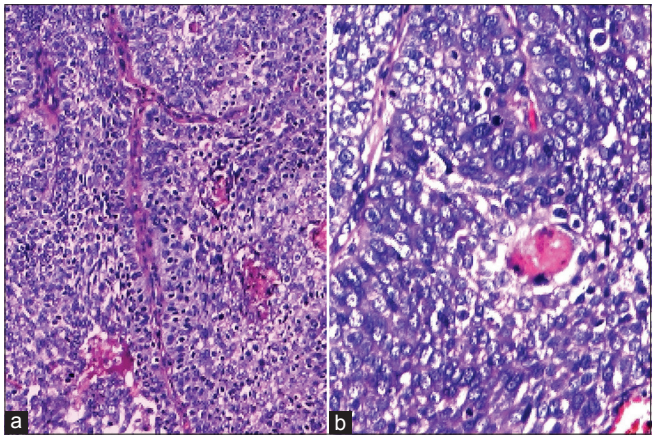

Figure 1: Photomicrograph showing lobules of large oval cells of varying sizes with pale bubbly cytoplasm. The nuclei are centrally placed with clear chromatin and prominent nucleoli (a and b) (H and E, ×100 and ×200)

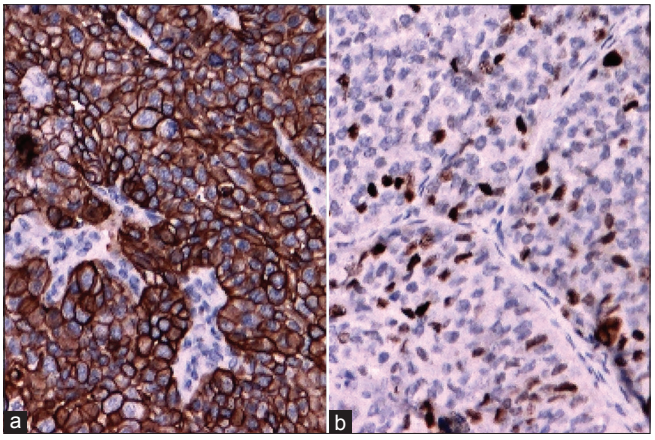

Figure 2: Photomicrograph showing (a) positivity for epithelial membrane antigen (immunohistochemical [IHC], ×200). (b) Proliferation index >5% for Ki-67 (IHC, ×200)

Wide excision with margins of 5-6 mm is the treatment of choice for well differentiated sebaceous carcinoma.[1,3] In cases of poorly differentiated lesions, wide excision is followed by radiotherapy and regular follow-up of skin, lymph nodes and other organ systems.[3] Local recurrence occurs in 32% of the cases. Use of Moh's micrographic surgery has been reported, with lower recurrence rates (approximately 12%).[1] Poor prognostic factors include multicentricity, size >1 cm in diameter, poor differentiation, extensive tissue infiltration, vascular or lymphatic involvement, pagetoid change and duration of symptoms >6 months. tumors <6 mm have an excellent prognosis.[2] Mortality rate varies from 20% to 22%.[3]

This is a case of rare adnexal tumor of unknown etiology characterized by aggressive course. The role of the pathologist is essential for the confirmation of diagnosis, treatment, and prognosis of these patients.

CONCLUSION

Though a rare tumor, sebaceous carcinoma should be kept in the differential diagnosis even in extraoccular sites. It follows an aggressive course; hence an early diagnosis is mandatory.

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