ABSTRACT

Sebaceous carcinoma is a rare neoplasm of the sebaceous gland. It is diagnosed mainly on histopathology and, clinically, it may mimic other neoplasms like squamous or basal cell carcinoma. We came across a patient presenting with a non-healing ulcer over the left temporo-parietal region of the scalp since 4 years and a single asymptomatic nodule over the occipital region since 3 years. Histopathology from an ulcer was pathognomonic of sebaceous carcinoma and that from a nodule was suggestive of proliferating trichilemmal cyst. The patient was screened for Muir Torre syndrome. We referred the patient to the oncologist for further management, where the patient was advised complete excision of the lesion.

Key words: Proliferating trichilemmal cyst, scalp, sebaceous carcinoma

INTRODUCTION

Sebaceous gland carcinoma is one of the rare appendageal tumors.[1] An increased frequency is seen in the Asian population.[2] It is three-times more common in the periorcular area than in other sites. Sebaceous carcinoma over the scalp is less frequently reported. We report a case of sebaceous carcinoma over the scalp presenting as a non-healing ulcer and associated with proliferating trichilemmal cyst.

CASE REPORT

A 42-year-old, married, moderate-built female presented with complaints of a solitary ulcer over the left side of the scalp since 4 years. The ulcer started as a hard swelling over the same site that gradually increased in size and ulcerated over a few months. The ulcer kept increasing in size despite systemic antibiotics and local care. She had no history of weight loss, gastrointestinal complaints, breathlessness, nausea, vomiting and recurrent fever. She had no eye complaints.

On examination, a single 6 cm X 6 cm-sized circular ulcer with erythematous indurated base with elevated margin was present over the left temporal and parietal region of the scalp [Figure 1]. Margins were rolled up and the base bled on touch. No significant lymphadenopathy of cervical or other group of lymph nodes was seen. A nodule of size around 3 cm was present over the occipital region. It was firm to hard in consistency, mobile and adherent to the overlying skin. A presumptive diagnosis of basal cell carcinoma or syringomatous carcinoma for the ulcer and a diagnosis of metastasis and trichilemmal cyst was thought for the nodule.

On investigation, blood biochemistry showed hemoglobin 10 gm%, total leucocyte count 9100 cells/ml and with normal liver and kidney function tests. Chest and skull radiograph was normal and no abnormality was detected on computed tomography of the brain and ultrasound examination of the abdomen. Enzyme-linked immunosorbent assay for human immunodeficiency virus and venereal disease research laboratory (VDRL) tests were negative. Pus culture and sensitivity test from an ulcer showed *Pseudomonas aeruginosa* growth with sensitivity to ceftazidime and piperacillin.

Skin biopsy taken from the edge of an ulcer showed invasion of dermis by poorly developed sebaceous lobules and moderate atypia and scalloped nuclei of some cells without peripheral pallisading or retraction spaces and moderately fibrotic stroma, sebaceous ducts at many places and foamy appearance to some cells suggestive of sebaceous carcinoma [Figure 2]. Epithelial
membrane antigen (EMA) staining to differentiate it from sebaceous carcinoma could not be done due to unaffordability of the patient; however, majority of the cells were sebocytes with a few basaloid cells and, thus, on clinicopathologic correlation, a diagnosis of sebaceous carcinoma was made. A biopsy from a nodule showed a cystic lesion in the dermis with a wall showing bland squamous proliferation. The wall of the cyst also showed multiple squamous eddies and abrupt trichilemmal keratinization, suggestive of proliferating trichilemmal cyst [Figure 3].

The patient was referred to an oncologist for further management, where she was advised complete excision of the ulcer.

**DISCUSSION**

The tumors of sebaceous glands are separated into three major categories: sebaceous adenoma, basal cell carcinoma with sebaceous differentiation (sebaceous epithelioma) and sebaceous carcinoma.[1] An increased frequency is seen in the Asian population.[2] It is estimated that approximately 25% of the sebaceous carcinomas occur in extraorbital sites, about 70% of which are in the head and neck region.[2] A tumor of sebaceous glands usually starts as a solitary, erythematous or sometimes pale yellow-colored, firm to hard, slowly growing nodule mainly over the head and neck and, less commonly, over the trunk, which may or may not ulcerate over time. It may also occur at the genitals.[3] It is a rare neoplasm comprising only less than 1% of the skin malignancies. The most common site of sebaceous carcinoma is the eyelids; however, lesions at other sites are not uncommon. It typically presents in old-aged women. It may be associated with Muir-Torre syndrome, which involves multiple low-grade visceral neoplasias with multiple sebaceous gland neoplasm of skin.[4] Our patient was screened for Muir Torre syndrome with colonoscopy, ultrasound examination of abdomen and chest and routine and microscopic urine examination, all of which turned out to be negative. Peri-orbital sebaceous carcinoma is approximately three-times more common than the extraorbital one. Some extraorbital tumors also may show rapid growth, and metastases are reported.[5]

In addition to its varied clinical appearance, a varied histologic appearance may occur, and delayed diagnosis or misdiagnosis following a biopsy is not uncommon.[6-8]

Histopathologically, the tumor is composed of multiple irregular sebaceous lobules of various sizes. The cells in the lobules are mainly undifferentiated, showing nuclear atypia and atypical mitotic figures. Necrosis may be also present in the center.[9] With fat stains on frozen sections, the cells are found to contain fine lipid globules. Some mature sebocytes may show multiple cytoplasmic vacuoles and scalloped nuclei. Some large lobules may show atypical keratinization, as seen in squamous cell carcinoma.[10] Tumor cells in sebaceous carcinoma are often large and may show squamoid changes. In this case, it should be differentiated from squamous cell carcinoma with hydropic changes. Sometimes, tumor cells show basaloid differentiation with inconspicuous lipidization, and the tumor must be distinguished from basal cell carcinoma with sebaceous differentiation.[11] Immunohistochemical staining for EMA can differentiate sebaceous carcinoma from basal cell carcinoma and squamous cell carcinoma.[11]

Complete surgical excision is the treatment of choice.[12] Reported local recurrence rates range from 9 to 36%, with larger series reporting recurrence rates in the range of 30%. Local recurrence tends to occur within 5 years.[13] Metastasis to liver, lungs, bones and brain may occur in 14–25% of the patients.[14,15]

Concurrent occurrence of ulcerated extraorbital sebaceous carcinoma with proliferating trichilemmal cyst in our patient may be a chance finding.
REFERENCES

1. Rulon DB, Helwig EB. Cutaneous sebaceous neoplasms. Cancer 1974;33:82-102.
2. Wick MR, Goellner JR, Wolfe JT 3rd, Su WP. Adnexal carcinomas of the skin. II. Extraocular sebaceous carcinomas. Cancer 1985;56:1163-72.
3. Urban FH, Winkelmann RK. Sebaceous malignancy. Arch Dermatol 1961;84:64.
4. Rütten A, Burgdorf W, Hügel H, Kutzner H, Hosseiny-Malayeri HR, Friedl W, et al. Cystic sebaceous tumors as marker for the Muir Torre syndrome: A histopathologic and molecular genetic study. Am J Dermatopathol 1999;21:405-13.
5. King DT, Hirose FM, Gurevitch AW. Sebaceous carcinoma of skin with visceral metastasis. Arch Dermatol 1979;115:862-3.
6. Izumi M, Tang X, Chiu CS, Nagai T, Matsubayashi J, Iwaya K, et al. Ten cases of sebaceous carcinoma arising in nevus sebaceus. J Dermatol 2008;35:704-11.
7. Doxanas MT, Green WR. Sebaceous gland carcinoma. Review of 40 cases. Arch Ophthalmol 1984;102:245-9.
8. Khan JA, Grove AS Jr, Joseph MP, Goodman M. Sebaceous carcinoma. Diuretic use, lacrimal system spread, and surgical margins. Ophthal Plast Reconstr Surg 1989;5:227-34.
9. Song A, Carter KD, Syed NA, Song J, Nerad JA. Sebaceous cell carcinoma of the ocular adnexa: Clinical presentations, histopathology, and outcomes. Ophthal Plast Reconstr Surg 2008;24:194-200.
10. Rulon DB, Helwig EB. Cutaneous sebaceous neoplasm. Cancer 1974;33:82-102.
11. Friedman KJ, Boudreau S, Farmer ER. Superficial epithelioma with sebaceous differentiation. J Cutan Pathol 1987;14:193-7.
12. Kuo T. clear cell carcinoma of skin. A variant of squamous cell carcinoma that stimulates sebaceous carcinoma. Am J Surg Pathol 1980;4:573-83.
13. Sinard JH. Immunohistochemical distinction of ocular sebaceous carcinoma from basal cell and squamous cell carcinoma. Arch Ophthalmol 1999;117:776-83.
14. Graham R, McKeever P, McGibbon D. Torre-Muir syndrome: An association with isolated sebaceous carcinoma. Cancer 1985;55:2868-73.
15. Nelson BR, Hamlet KR, Gillard M, Railan D, Johnson TM. Sebaceous carcinoma. J Am Acad Dermatol 1995;33:1-15; quiz 16-8.

Cite this article as: Chikhalkar S, Garg G, Gutte R, Khopkar U. Sebaceous carcinoma of scalp with proliferating trichilemmal cyst. Indian Dermatol Online J 2012;3:138-40.

Source of Support: Nil, Conflict of Interest: None declared.