Unusual Presentation of Ulcerative Postauricular Swelling as Sebaceous Cell Carcinoma

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ABSTRACT

Sebaceous glands have high concentration over head and neck region. Despite high concentration, sebaceous cell adenoma and carcinomas are infrequent. Sebaceous cell carcinoma is an uncommon, cutaneous aggressive tumor arising from the sebaceous glands and seen almost exclusively on the eyelids (75%). It accounts for just 0.2–0.7% of all eyelid tumors in the USA and very few cases that have originated in areas other than the eyelids have been reported. A 67-year-old male presented with swelling (3 cm × 4 cm), on the right postauricular region, since about 1-month. The swelling became ulcerative and associated with progressive tinnitus and hoarseness of voice. The patient was investigated. Fine-needle aspiration cytology suggested sebaceous cell carcinoma. Then excision biopsy was done, and histopathological examination of excised tissue confirmed the diagnosis. Extraorbital sebaceous cell carcinoma is an aggressive and invasive malignancy. It clinically mimics other diseases and is difficult to diagnose. Hence, an accurate and prompt diagnosis is crucial because of its fulminant course, serious associations with Muir-Torre syndrome and high potential for regional and distant metastasis.

KEYWORDS: Extraocular, sebaceous cell carcinoma, sebaceous gland

INTRODUCTION

Sebaceous cell carcinoma is an uncommon, cutaneous tumor, arising from the sebaceous glands. It was first well-described by Allaire in 1891.^[1] Mostly these tumors have no obvious etiology, and only a few are associated with Muir-Torre syndrome (MTS). Sebaceous gland tumor may crop up anywhere on the body, where these glands exist, including the genitalia.^[2] Despite high concentration, true neoplasm, that is, sebaceous cell adenomas and carcinomas are infrequent and very few cases have been reported.[3] This tumor has a fulminant clinical course, with a considerable propensity for local recurrence and distant metastasis. Diagnosis and therapy tend to be delayed because sebaceous carcinoma is frequently mistaken for more common benign entities, further complicating treatment of this aggressive malignancy.^[4] In addition to its varied clinical appearance, a varied histologic appearance may occur, and delayed diagnosis or misdiagnosis following a biopsy is not uncommon.^[5] The ocular region accounts for nearly 75% of

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all reported cases, as head and neck region of the body has the greatest density of sebaceous glands and its ectopias. Classically, it occurs in females and the older population at sixth to a seventh decade, and arises in the ocular region from the meibomian gland of the tarsal plate and upper eyelid.^[6] The parotid gland is most common site outside the ocular region, accounting for about 20% of cases.^[7] Overall, it is uncommon tumor with orbital sebaceous carcinoma accounting for only 0.2–0.7% of all eyelid tumors.^[8]

CASE REPORT

A 67-year-old male, presented with complaints of swelling, on the right postauricular region [Figure 1], since about 1-month. The swelling increased in size (3 cm \times 4 cm) rapidly and developed discharging ulcer over the swelling associated with progressive tinnitus and hoarseness of voice. The lesion was nontender, fixed to overlie skin and underlying tissue with no regional lymph-adenopathy. Ear-nose-throat examination and systemic examination was unremarkable. All investigations of blood and urine were within normal limits. No abnormality was detected

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Figure 1: Postauricular swelling (3 cm × 4 cm)

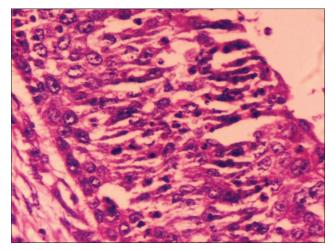


Figure 3: Cells arranged in sheets with finely vacuolated to foamy cytoplasm (H and E stain: ×40)

on radiological investigations, that is X-ray skull, X-ray chest, and USG whole abdomen.

Fine needle aspiration cytology suggested sebaceous cell carcinoma and excision biopsy was performed. Cut surface revealed yellowishgrey tumor with an area of necrosis and hemorrhages [Figure 2]. Microscopic examination [Figures 3 and 4] exhibited lobules of tumor cells separated by fibrovascular stroma. The nuclei showed pleomorphism, hyperchromasia and cytoplasm of the cell were characteristically finely vacuolated to foamy. Some areas showed focal necrosis with the comedo-like pattern [Figure 3], while other areas displayed several mitiotic figures, globules and gland-like structure formation. The distinctive microscopic findings confirmed the diagnosis of sebaceous cell carcinoma, and as the extent of the lesion precluded further surgical treatment, radiotherapy was advised for palliation.

DISCUSSION

Sebaceous gland carcinoma is an aggressive, uncommon, cutaneous tumor, may arise anywhere on the body where these

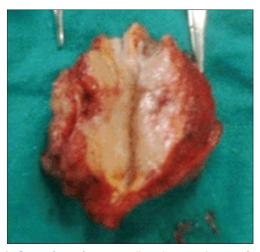


Figure 2: Cut surface of tumour-yellowishgrey with area of necrosis and hemorrhages

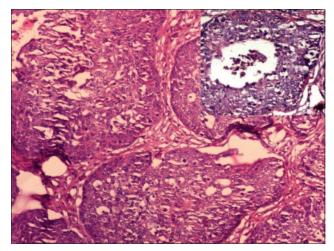


Figure 4: Section showing lobules of tumor cells separated by fibrovascular stroma, cytoplasm of the tumor cells within the lobules is finely vacuolated to foamy (H and E stain: ×10)

glands exist, first well described by Allaire in 1891.^[1] It may appear on preexisting dermatosis, such as nevus sebaceous and actinic keratosis or may follow radiation therapy for other diseases. It may also occur in MTS, characterized by the occurrence of sebaceous tumors in association with visceral malignancies. Sebaceous carcinoma is traditionally classified into two groups: Tumor arising from the ocular adenexa, and those arising in extra ocular sites. Extraocular sebaceous carcinoma most commonly involve the head and neck region, the parotid and submandibular glands, the external auditory canal, the trunk and upper extremity, sole, the dorsum of the great toe, and laryngeal or pharyngeal cavities. The sex distribution of extra-orbital sebaceous carcinoma appears to be about equal for male and female patients, and the mean age of occurrence is 63 years.^[9]

The disease exhibits such a variety of clinical presentations, that the clinical appearance of extraocular sebaceous carcinoma is not pathognomic, but the lesion may be a pink to the redyellow nodule. Diagnosis and therapy tend to be delayed because sebaceous carcinoma is frequently mistaken for more common benign entities, further complicating treatment of this aggressive malignancy.^[4] In addition to its varied clinical appearance, a varied histologic appearance may occur, and delayed diagnosis or misdiagnosis following a biopsy is not uncommon.^[5] Extraocular sebaceous carcinoma, in association with nevus sebaceous, in the postauricular region or in the external auditory canal is very rarely described in the literature.^[10] Although extraocular sebaceous carcinoma, compared to orbital sebaceous carcinoma, is generally considered less aggressive, visceral metastasis has been reported.^[11] Draining lymph nodes may be involved in few cases. The possibility of MTS must be considered in every case of sebaceous tumor. Criteria for diagnosis of MTS include the presence of at least one sebaceous adenoma, sebaceous epithelioma, or sebaceous carcinoma and at least one visceral cancer.^[12]

In our patient, there was neither any regional lymph adenopathy nor any evidence of other internal malignancies as associated with MTS. Metastases have been reported to occur as late as 5 years after the initial diagnosis, lending support to the continual surveillance of patients with sebaceous carcinoma.^[9]

Histologically, sebaceous carcinomas are often poorly differentiated neoplasms mainly within the dermis. Multiple lobules of basaloid undifferentiated cells are present within the dermis. In the central portion of lobules, more mature cells are present. Marked nuclear atypia, pleomorphism, and mitosis are common.^[13] This neoplasm may be confused with tumors composed of basal cells, squamous cells (mucoepidermoid and spindle cell carcinoma), clear and balloon nevus cells as well as other sebaceous neoplasm.^[14]

CONCLUSIONS

Sebaceous carcinoma is an uncommon tumor. Extraorbital sebaceous cell carcinoma is an aggressive and invasive malignancy. It clinically mimics other diseases and is difficult to diagnose. Further, correct preoperative diagnosis of sebaceous cell carcinoma is rarely made and its malignant potential is usually underestimated by the surgeon. Hence, an accurate and prompt diagnosis is crucial because of its fulminant course, serious associations with MTS and high potential for regional and distant metastasis.

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Conflicts of interest

There are no conflicts of interest.

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