Postauricular sebaceous cell carcinoma

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ABSTRACT

Introduction: Sebaceous cell carcinoma is an aggressive tumor arising from the sebaceous glands. It is relatively rare tumor and seen almost exclusively on the eyelids (75%). Despite the high concentration of sebaceous glands over head and neck region, true neoplasm, i.e. sebaceous cell adenoma and carcinomas are infrequent. It accounts for just 0.2–0.7% of all eyelid tumors in USA and very few cases that have originated in areas other than the eyelids have been reported. Case Report: A 70-year-old male presented with complaints of rapidly growing swelling (3×4 cm), on the right post auricular region, since one and half months. The patient developed discharging ulcer over the swelling associated with progressive tinnitus and hoarseness of voice, since last twenty days. Fine needle aspiration cytology (FNAC) was advised, which suggested the diagnosis of sebaceous cell carcinoma and excision biopsy was done. Histopathological examination of excised tissue confirmed the diagnosis.

Conclusion: 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 fulminating course, serious associations with Muir–Torre syndrome and high potential for regional and distant metastasis.

Keywords: Sebaceous gland, Sebaceous cell carcinoma, Extraocular

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INTRODUCTION

Sebaceous cell carcinoma is an uncommon, cutaneous tumor first well-described by Allaire in 1891 [1]. Most sebaceous gland carcinomas have no obvious etiology and only a few are associated with Muir–Torre syndrome. This tumor is thought to arise from sebaceous glands in the skin and, thus, may crop up anywhere on the body, where these glands exist, including the genitalia [2]. Despite this relatively high concentration, true neoplasm, i.e., sebaceous cell
adenomas and carcinomas are infrequent and very few cases have been reported in scientific journals [3]. This tumor has a fulminant clinical course, with a considerable propensity for both 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]. Ocular region accounts for nearly 75% of 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 older population at 6-7th decade, and arises in ocular region from the meibomian gland of 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]. So far only few cases of extra orbital sebaceous carcinoma are reported in literature and none in the postauricular region [7].

CASE REPORT

A 70-year-old male, presented in surgery outpatient department with complain of rapidly growing swelling, on the right postauricular region (Figure 1), since one and half months. Initially, a very small asymptomatic lesion was present for which no medical opinion was sought. The lesion progressively increased in size and for last 20 days patient had developed discharging ulcer over the swelling associated with progressive tinnitus and hoarseness of voice. His past history was unremarkable and there was no family history of similar lesion. On local examination, a moist greyish pink ulcerated growth of 3x4 cm was present behind his right pinna with features suggestive of facial nerve paresis. The lesion was non tender, fixed to overlying skin and underlying tissue. No regional lymph-adenopathy was present. ENT examination and systemic examination was unremarkable. His routine investigation on blood and urine were within normal limits. Further his radiological investigations, i.e. X-ray skull, X-ray chest and USG whole abdomen too revealed normal study.

Fine needle aspiration (FNA) of swelling was advised which suggested the diagnosis as sebaceous cell carcinoma and excision biopsy was performed. Biopsy tissue on histo-pathological examination of cut surface revealed yellowish-grey tumor with 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 was characteristically finely vaculated to foamy. Some areas showed focal necrosis with comedo-like pattern (Figure 3-Inset), while other areas displayed several mitotic 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 first well described by Allaire in 1891 [1]. But this disease was not firmly accepted until 1956 when Straatsma thoroughly studied the histological and clinical presentation of this disease. This tumor is thought to arise from sebaceous gland in the skin and thus, may arise anywhere on the body where these glands exist [1]. It may appear on the top of pre-existing dermatosis, such as nevus sebaceous and actinic keratosis or may follow radiation therapy for other diseases. It may also occur in Muir–Torre syndrome, characterized by occurrence of sebaceous tumors in association with visceral malignancies. Sebaceous carcinoma is traditionally classified into two groups: tumor arising from the ocular adnexa, particularly the meibomian glands and glands of Zeiss, 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 and histological growth patterns that the diagnosis is often delayed for months to years. The clinical appearance of extraocular sebaceous carcinoma is not pathognomic, but the lesion may be a pink to red-yellow nodule. Extraocular sebaceous carcinoma, in association with nevus sebaceous, in the postauricular region or in external auditory canal is very rarely described in 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 Muir–Torre syndrome 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 lymphadenopathy nor any evidence of other internal malignancies as associated with Muir–Torre syndrome. However, immunohistochemistry, an important aid to diagnosis of Muir–Torre syndrome, could not be performed on the tumor to look for expression of mismatch repair genes because of local non-availability and financial constraints. Metastases have been reported to occur as late as five 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 basaloïd 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]. Histochmically, the clear cells of sebaceous carcinomas are negative with periodic acid-Schiff and Alcian blue staining. Immunohistochemically, the tumor cells of sebaceous carcinomas show positive reactions for EMA and Ber-EP4, in contrast to squamous cell carcinoma which are negative for Ber-EP4. Human milk fat globules subclass 1 and 2 (HMFG1 and HMFG2) are positive in basal cell carcinoma which helps it to distinguish it from sebaceous cell carcinoma in which it
CONCLUSION

We conclude that sebaceous carcinoma is a rare tumor and 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 Muir–Torre syndrome and high potential for regional and distant metastasis.

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Author Contributions
Suresh K Bhatia – Acquired the data, Revised it critically for important intellectual content, Final approval of the version to be published
Shivani Atri – Conception and design, Acquired the data, Drafted the article, Collected review articles, Final approval of the version to be published
Minakshi Sardha – Drafted the manuscript, Revised it critically for important intellectual content, Final approval of the version to be published
Sufian Zaheer – Acquired the figures, Revised the manuscript, Final approval of the version to be published
Syed Asmat Ali – Revised the manuscript critically for important intellectual content, Final approval of the version to be published
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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES