Extra ocular sebaceous carcinoma: A rare case report

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ABSTRACT

Extra ocular Sebaceous Carcinoma is a rare malignancy when compared to Peri ocular variant and these are derived from sebaceous gland epithelium. The aggressive types of extra ocular sebaceous neoplasm are reported with lymph node and visceral metastasis associated with poor prognosis. Here we report a case of extensive cutaneous extra ocular sebaceous cell carcinoma confined to large area of scalp proven by Immunohistochemistry without intra cranial involvement, distant metastases or evidence of Muir-Torre syndrome.

Key words: Extraocular, Sebaceous Carcinoma, Immunohistochemistry

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INTRODUCTION

Sebaceous carcinoma, first described by Allaire in 1891 accounts for less than 1% of all cutaneous malignancies.[1] It is a rare but aggressive malignant neoplasm that arises from sebaceous glands with a tendency for both local recurrence and distant metastases.[2] Sebaceous carcinoma can either be ocular or extraocular, and extraocular type is rare.[3] We report a case of an extraocular type of sebaceous carcinoma of the scalp with no distant metastases.

CASE REPORT

A 65-year elderly man presented with a large asymptomatic swelling over the vertex of the scalp, which gradually progressed over a period of twelve months. He had no history of trauma, neavi, or bleeding at this site. Examination showed a large swelling of 15 × 12 × 5 cm size situated over the vertex of the scalp extending to the occipital and tempero-parietal regions.The tumor showed focal ulceration with yellowish malodorous seropurulent discharge [Figure 1]. The swelling was soft to firm in consistency, base indurated with minimal tenderness. There was no localized or generalized lymphadenopathy. Clinically, a differential diagnosis of cylindroma, angiosarcoma, deep fungal infections, nocardiosis, mycobacterial or atypical mycobacterial infections were considered. Routine blood investigations, chest X-ray and ultrasound abdomen were normal.

The culture of the seropurulent discharge revealed *Staphylococcus aureus* and *Acinetobacter baumanni* suggestive of secondary bacterial infection. AFB stain and culture for both mycobacteria and atypical mycobacteria and fungal culture were negative. CT scan of the brain showed only age related cerebral atrophy, and there was no evidence of extension of the tumour or metastatic deposits.

A wedge biopsy was sent for histopathology and immunohistochemical marker studies. Histopathology revealed an infiltrating neoplasm in the dermis composed of cells arranged in lobules separated by fibrovascular stroma. [Figure 2] Neoplastic cells were round having moderate amount of eosinophilic to vacuolated cytoplasm and large nuclei with prominent eosinophilic nucleoli. [Figure 3] Cells exhibited marked nuclear pleomorphism and increased mitotic activity. [Figure 3] Focal sebaceous differentiation was also noted. The epidermis was normal.

Immunohistochemical marker study showed diffuse strong positivity for cytokeratin and focal positivity for epithelial membrane antigen (EMA) [Figure 4].

DISCUSSION

Sebaceous carcinoma is a rare malignant

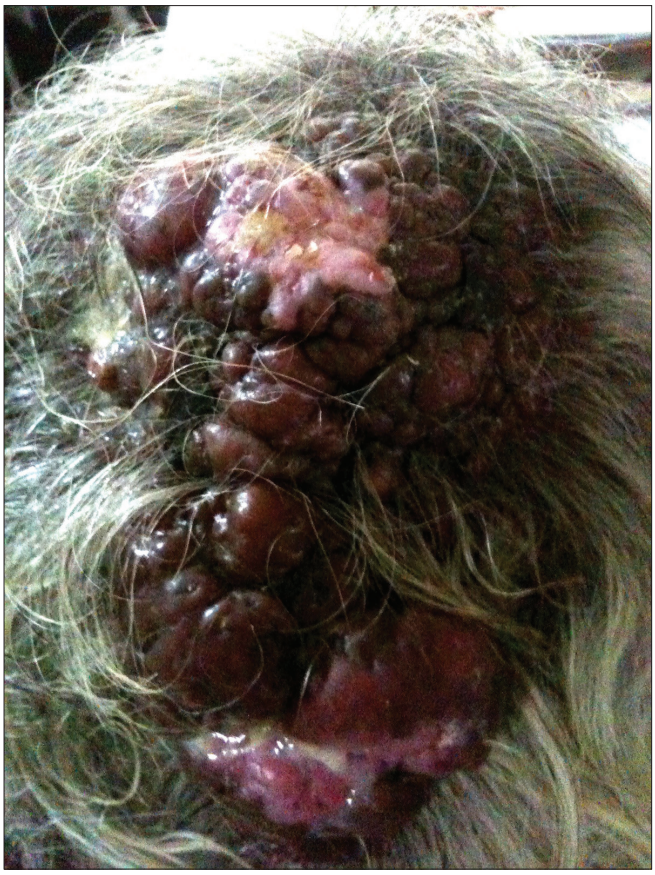

Figure 1: A large multi- lobulated swelling with ill-defined margins and irregular surface showing focal ulceration and yellow sero-purulent discharge

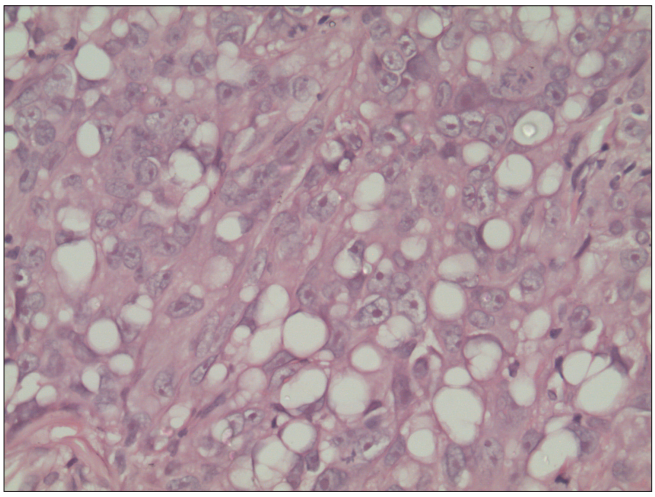

Figure 3: Neoplastic cells having moderate amount of eosinophilic to vacuolated cytoplasm and large nuclei with prominent eosinophilic nucleoli and increased mitoses (H and E, ×40)

neoplasm, often occurs in adults with a slight male preponderance[4] This malignancy can occur as peri-ocular and extraocular types and the former variant contributes to 75% of sebaceous neoplasms.[3,5] The upper eyelid is affected two to three times more often than the lower eyelid

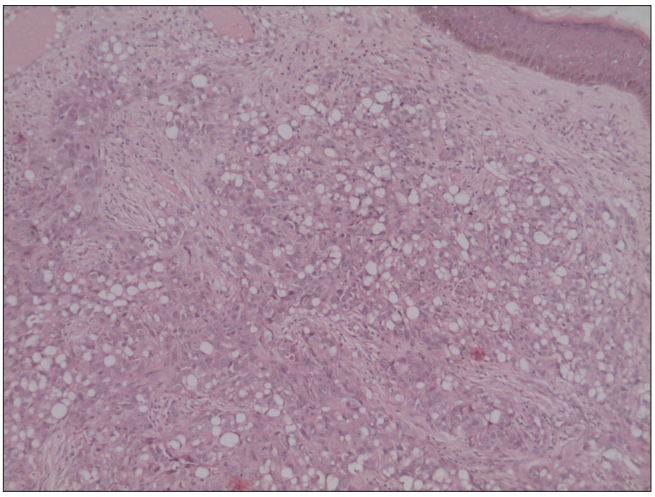

Figure 2: Dermis showing a neoplasm composed of cells arranged in lobules separated by fibrovascular stroma (H and E, ×4)

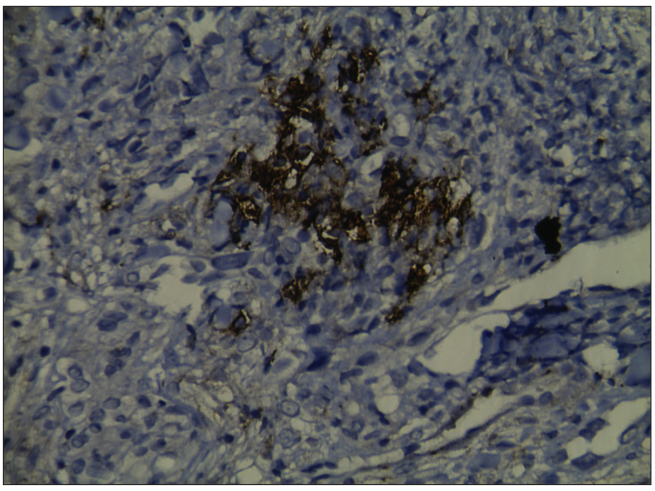

Figure 4: Focal positivity for EMA (Immunohistochemistry marker, ×40)

due to greater number of meibomian glands[6,7] Extraocular sebaceous carcinoma which constitutes 25%[8] of the sebaceous neoplasms has been reported more commonly on the head and neck areas[4,8] followed by trunk, salivary glands, genitalia, breast, ear canal, and the intra-oral cavity. The most frequent clinical presentation is a painless subcutaneous nodule but it can also present as pedunculated lesions, irregular mass or diffuse thickening of the skin. This protean appearance frequently masquerades as other benign tumors or inflammatory conditions thereby leading to delay in diagnosis, inappropriate treatment, increased morbidity, and mortality.

Sebaceous carcinoma histologically may be classified as well, moderately, or poorly differentiated. The morphological hall mark of sebaceous differentiation is the detection of sebaceous cells and demonstration of fat in vacuolated tumor cells. Special stains such as oil red O may be helpful in confirming the presence of fat, but requires frozen section. This was not done as the specimen had already been fixed in formalin.

Other differential diagnosis includes basal cell carcinoma with sebaceous differentiation for poorly differentiated sebaceous cell carcinoma. Basal cell carcinomas exhibit peripheral palisading and clefting from the adjacent stroma. Our case did not show poorly differentiated basaloid cells and other features of basal cell carcinoma.

Sebaceous carcinoma cells express immunohistochemical markers such as cytokeratin, epithelial membrane antigen (EMA), Cam5.2 and anti-breast carcinoma associated antigen-225 antibody. The current case showed diffuse strong cytokeratin expression and EMA was focally positive in cells with sebaceous differentiation. Considering the morphological, histopathological and immunohistochemical marker study findings we favoured a diagnosis of sebaceous carcinoma. The common associations of sebaceous carcinoma are Muir-Torre syndrome, an autosomal dominant condition comprising of sebaceous neoplasm with one or more low-grade visceral malignancies and Nevus sebaceous of Jadassohn.[9]

Distant metastases and recurrence rates are more common in the ocular type of sebaceous carcinoma[3,10] when compared to extraocular sebaceous carcinoma as seen in our case. Recurrence rates of ocular sebaceous carcinoma ranges from 11% to 30% with distant metastases occurring in 3% to 25%[3,10] Though extraocular sebaceous carcinoma is less aggressive, a study with 91 cases of extraocular sebaceous carcinoma showed a recurrence rate of 29% with 21% developing metastases.[11] Mortality rates irrespective of ocular or extraocular type ranges from 9% to 50%.[12,13] Wide excision and selective use of radiotherapy is the ideal treatment of choice.[14,15]

In summary, extraocular sebaceous carcinoma is a very rare and aggressive malignancy. This tumor should suggest the possibility of Muir-Torre syndrome and alert the clinician to search for occult malignancies.

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