

CASE REPORT

A case report of aggressive sebaceous carcinoma of the scalp

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Abstract

Malignant pilosebaceous neoplasm of the scalp is a very rare tumor. A 60-year-old man presented with a rapidly enlarging, ulcerated, and firm nodular mass over the scalp for a duration of 3 months. A few months back, the patient noticed a subcutaneous nodule at the same site, and it was reported as sebaceous adenoma on histopathology. The swelling recurred at the same site and was surgically excised and sent for histopathology. A histological diagnosis of sebaceous carcinoma of the scalp was made. On follow-up, there was no recurrence or distant metastasis. Due to the rarity and aggressive behavior of the malignant pilosebaceous neoplasm of the scalp, we present this case along with clinical and histopathological findings.

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<https://doi.org/10.36922/td.v1i2.203>**Received:** September 23, 2022**Accepted:** November 17, 2022**Published Online:** December 15, 2022**Copyright:** © 2022 Author(s). This is an Open Access article distributed under the terms of the Creative Commons Attribution License, permitting distribution, and reproduction in any medium, provided the original work is properly cited.**Publisher's Note:** AccScience Publishing remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

1. Background

Malignant pilosebaceous neoplasm or sebaceous carcinoma (SC) is a rare tumor of the sebaceous gland. It can be classified into ocular and extraocular types. It commonly affects the head-and-neck region, with the periocular area being the most common site^[1]. SC typically presents as a firm subcutaneous nodule that enlarges gradually. The aggressive behavior of this tumor is debatable^[2].

2. Case presentation

A 60-year-old man presented with a rapidly enlarging, ulcerated, and firm nodular mass over the scalp for a duration of 3 months. He had no family history of malignancy or other contributory family history. Several months back, he noticed a subcutaneous nodule at the same site and it was diagnosed as sebaceous adenoma on histopathology by a pathology laboratory. After an interval of 2 months, it recurred, following which he underwent wide local excision again.

On examination, a single, nodular swelling over the scalp at the occipital region was observed with surface ulceration, measuring 2.5 × 1.8 cm. It was soft to firm in consistency and adhered to the overlying skin. He had no lymphadenopathy and his systemic examination was normal.

The tumor was surgically excised with wide margins. On gross examination, it was a single nodular mass measuring $2.5 \times 1.5 \times 1.0$ cm with surface ulceration. On cut section, it was firm to hard, greyish-brown, and fleshy (Figure 1).

Histopathological examination (Figures 2 and 3) with hematoxylin and eosin showed neoplastic cells of basaloid, basosquamous, and epidermoid type with varying degrees of differentiation and arranged in irregular lobules and sheets. The intervening stroma was fibrovascular. The tumor lobules had mild pleomorphic cells with hyperchromatic nuclei and a moderate amount of vacuolated or foamy cytoplasm. There was also increased mitoses. In poorly differentiated areas, the tumor cells were highly pleomorphic, hyperchromatic, or vesicular with prominent nucleoli. The neoplastic cells had large multivacuolated foamy cytoplasm. Multifocal epidermal

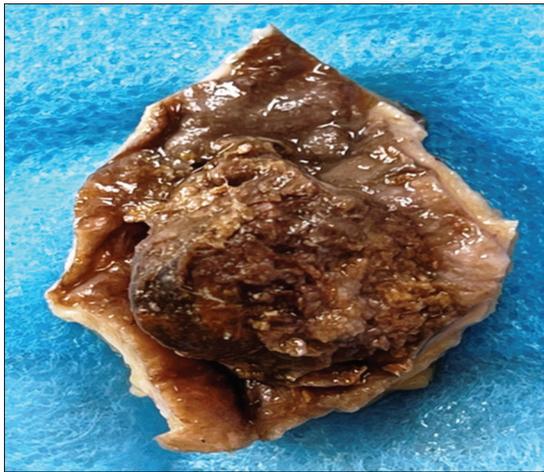


Figure 1. Gross examination showing a nodular mass lesion measuring $2.5 \times 1.5 \times 1.0$ cm with surface ulceration.

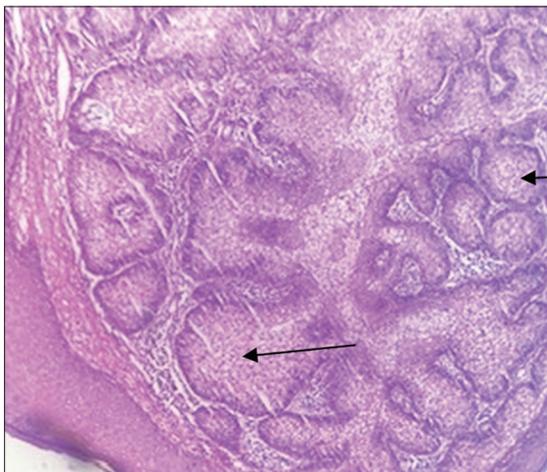


Figure 2. Photomicrograph showing neoplastic cells with various degrees of differentiation and arranged in irregular lobules of atypical sebaceous cells (arrows) (H&E stain, $\times 40$).

ulceration was also observed. A histological diagnosis of sebaceous carcinoma of the scalp was made.

On follow-up, the patient was asymptomatic, without any evidence of locoregional recurrence or metastasis. We present this case in view of its aggressive nature.

3. Discussion

SC is a rare tumor with sebaceous differentiation. Although it is a slow-growing tumor, it occasionally shows rapid and aggressive behavior. It constitutes 0.2–4.6% of all malignant epithelial lesions^[3]. SC predominates in the periorcular region and occurs more frequently in Asian population and in women more than 40 years of age^[4]. Since there are abundant sebaceous glands over the face and scalp, these areas are often affected by extraocular SC. Although SC rarely occurs in other parts of the body, it may occur in certain areas, including the trunk, extremities, genitalia, and external auditory meatus. Although aggressive behavior in SC of the scalp is rare, in our case, the tumor, which was located over the scalp, showed aggressive behavior.

The risk factors for SC include patient's weak immune system, advanced age, excess exposure to ultraviolet rays from the sun, medications, radiation, immunosuppression, inherited diseases such as Muir-Torre syndrome^[5]. Patients with Muir-Torre syndrome may have malignancy along with sebaceous tumor-like adenoma or SC. Clinically, SC presents as an asymptomatic and yellowish nodular lesion, often with ulceration. The primary sites include the eyelid (38.7%), scalp, and neck (8.7%)^[6].

The pathogenesis of SC is unknown. It may begin as an inflammatory condition, which is often overlooked. The

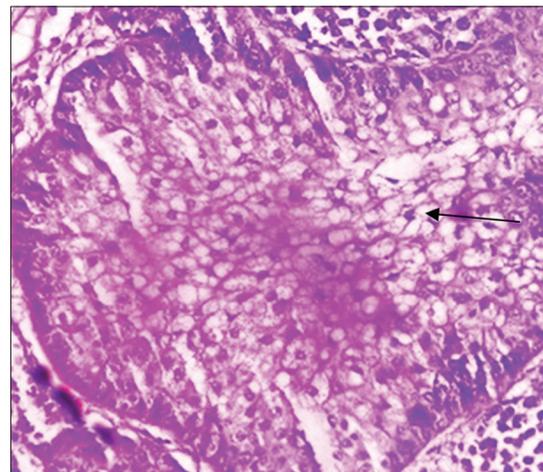


Figure 3. Photomicrograph showing neoplastic cells having hyperchromatic and pleomorphic cells with multivacuolated or foamy cytoplasm (arrow) (H&E stain, $\times 100$).

pathogenic germline variants of DNA mismatch repair genes *MSH2*, *MSH6*, and *MLH1* have been identified in 8–29% of individuals with SC^[7]. In these patients, the tumors are characterized by microsatellite instability.

On microscopy, sebaceous neoplasm shows a wide range of differentiation: Sebaceous adenoma, basal cell carcinoma with sebaceous differentiation, and sebaceous carcinoma^[8]. In SC, the tumor cells are arranged in cords and lobules, the neoplastic cells show varying degrees of sebaceous differentiation, and there is tumor infiltration to adjacent soft tissue, nerve, or the lymphatic system. Less commonly, it exhibits a broad superficial intraepidermal pattern.

The architecture usually consists of sheets or lobules separated by fibrovascular stroma. The various histologic patterns of SC include lobular, cystic with the central comedo-type necrosis, papillary, and mixed type. The atypical sebocytes may be well, moderately, or poorly differentiated. They are arranged as rounded nodular aggregates or angulated infiltrative aggregates. A well-differentiated SC shows increased proportion of mature appearing multivacuolated sebocytes with nuclear indentation, mild nuclear pleomorphism, and minimal mitoses and necrosis. In contrast, anaplastic cells, with prominent nuclear pleomorphism and frequent mitoses and necrosis, are observed in moderate to poorly differentiated SC. In terms of tumor dissemination, these tumors may occasionally spread in a pagetoid manner^[9]. Tumor multicentricity, differentiation, pagetoid spread, and perineural, vascular, and lymphatic invasion should be emphasized in pathological reporting since this information can aid clinicians in treating patients with SC.

Sebaceous adenoma, basal cell carcinoma with sebaceous differentiation, clear cell melanoma, clear cell squamous cell carcinoma, clear cell hidradenocarcinoma, metastatic renal cell carcinoma, and prostate carcinoma are among the benign and malignant conditions that are considered differential diagnoses for SC^[10].

Sebaceous adenoma is a benign epithelial neoplasm with hyperplasia of sebaceous lobules associated with expansive aggregates of basaloid germinative cells. It is a well circumscribed neoplasm that principally demonstrates organoid and lobular configuration and contains a significant percentage of mature and lipid-rich sebaceous cells.

Another differential is basal cell carcinoma with sebaceous differentiation. Its tumor cells are small basaloid with peripheral palisading, surrounded by fibromyxoid stroma, with focal differentiation toward mature, benign-appearing, and multivacuolated sebocytes. In contrast, there is no peripheral palisading in SC.

Clear cell squamous cell carcinoma, on the other hand, shows clear cells; however, their cytoplasm is not as multivacuolated as sebocytes. Without lobular arrangement, it is difficult to determine sebaceous differentiation in squamous cell carcinoma^[11].

On immunohistochemistry, SC tumor cells are reactive for epithelial membrane antigen (EMA), cytokeratin, Ber-EP4, and adipophilin (adipose differentiation-related protein, ADP), which has a membranous vesicular pattern, and androgen receptor^[12]. Androgen receptors and ADPs are not observed in squamous cell carcinoma, but EMA, cancer antigen (CA)15-3, Ber-EP4, and ADP are observed in basal cell carcinoma, thus differentiating them from SC. Immunohistochemistry can also be used to evaluate *MSH2*, *MSH6*, *MLH1*, and *PMS2* for microsatellite instability in SC, as SC may metastasize in 2.4% of cases. According to the literature, local recurrence is more common in extraocular SC^[12].

One of the features that is indicative of poor prognosis is tumor size >1 cm (associated with a 5-year mortality rate of 50%). The other features include moderate-to-poor sebaceous differentiation, tumor necrosis, increased mitotic activity, infiltrative growth, and lymphovascular invasion.

For local disease, wide local excision is the preferred treatment. Other modalities of treatment include Mohs micrographic surgery, radiation, and systemic chemotherapy, which may be considered for recurrent or metastatic disease^[13].

Bailet *et al.* have reported a local recurrence rate of 29%, regional nodal metastasis in 15%, and a disease-related mortality of 20%^[9]. It has also been reported that the 5-year survival rate of this SC is 92.7%. SC is primarily treated with wide local excision, and in cases of localized tumor, the prognosis is good following surgical removal. However, in scalp SC, adjuvant treatment with radiation and chemotherapy is required. A study conducted by Angela Orcurto *et al.* noted the recurrence of aggressive SC of the scalp even after multiple excisions and local radiotherapy^[14]. In another study, Bhavaraju noted aggressive SC over the scalp and suggested the need for close follow-up of these patients to detect recurrence and distant metastasis^[15]. In our case, the patient was asymptomatic without any evidence of locoregional recurrence on follow-up.

4. Conclusion

Extraocular SC is an aggressive malignant neoplasm of the skin. The management of SC is challenging and patient assessment may be necessary depending on the prognostic features. We present a case of SC of the scalp along with

clinical and histopathological findings in light of its rarity and aggressive behavior.

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Conflict of interest

The authors declare no conflicts of interest.

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Ethics approval and consent to participate

Not applicable.

Consent for publication

Not applicable.

Availability of data

The data can be requested from the corresponding author following reasonable reason.

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