International Journal of Health Sciences and Research
Vol.11; Issue: 2; February 2021
Website: www.ijhsr.org
Case Report *ISSN: 2249-9571*

Sebaceous Carcinoma of Scalp: A Rare Occurrence with a Diagnostic Challenge

Anshika Rai[1], Shilpi Sahu[2], Reeta Dhar[3]

[1]Junior Resident, [2]Head of Department & Professor, [3]Professor,
Department of Pathology, MGM Medical College, Navi Mumbai.

Corresponding Author: Anshika Rai

ABSTRACT

Sebaceous carcinoma (SC) is an aggressive malignant adnexal tumour, which tends to be an exception in adults, more so in males. Common among the females, in seventh decade, the extraocular type of SC usually occurs over the face and scalp. They are derived from the sebaceous gland epithelium. Although no obvious aetiology is identified, a few cases show association with Muir-Torre syndrome (MTS). It has a diverse clinical presentation which often causes it to be mistaken for common benign entities.[10] The definite diagnosis is confirmed on histopathology. The characteristic histologic criteria include lobular architecture, foamy vacuolization of the cytoplasm, nuclear pleomorphism, and mitotic activity.

Keywords: Sebaceous carcinoma, malignant adnexal tumour, extra-ocular, scalp.

INTRODUCTION

Sebaceous carcinoma (SC) is a malignant adnexal tumour, which tends to be an exception in adults, more so in males. [1] Till a decade earlier, nearly 150 cases of extra orbital sebaceous carcinomas were reported. [2] Derived from sebaceous gland epithelium, [3] these are aggressive tumours, 75% arising in the periocular region. [4] Extraocular SC is extremely uncommon, occurring usually in the face and scalp[5] but, can also occur on the trunk, extremities, genitalia and rare sites like lungs, salivary glands, breast. [6] It usually arises in the seventh decade and have no obvious aetiology with a few cases showing association with Muir-Torre syndrome (MTS) [7,8] These tumours have a high incidence of local recurrence approximately 30%-40% and regional metastasis around 10%-15%. [8,9] It has a diverse clinical presentation which often causes it to be mistaken for common benign entities.[10] The definite diagnosis is mainly on histopathology. [9,11] Adnexal tumours in general, represents neoplasms varying in behaviour and malignant potential and poses a diagnostic challenge, [12] which holds true for malignant tumours of the scalp, which are rare and associated with a poor prognosis. [1]

CASE REPORT

A 70 years old male, presented to the surgery outpatient department with a lesion on scalp, for 10 years which had started growing rapidly within the past month. On physical examination, an irregular, exophytic, firm to hard, immobile growth with ulcerated appearance, over the right parietal region was noted. (Figure 1) It bled on touch and was infested with maggots. The tumour was excised and sent for histopathological examination. The gross examination revealed, a single, grey-brown, tissue mass measuring 8.5 X 9 X 4 cm in size, with hair-bearing skin attached to it. External surface was grey-white, irregular, hair-bearing, fungating mass. (Figure 2) Cut surface showed grey-white to grey-brown

areas with areas of haemorrhages along with a few congested blood vessels. (Figure3) Several sections were taken and slides were reported.

Figure 1

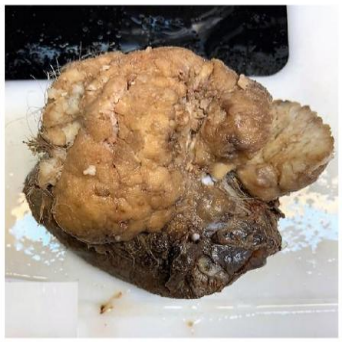


Figure 2

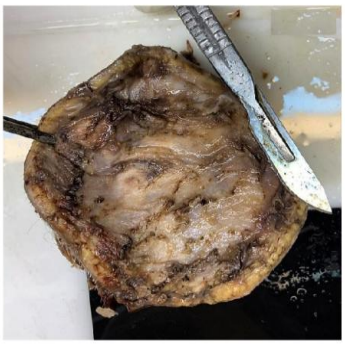
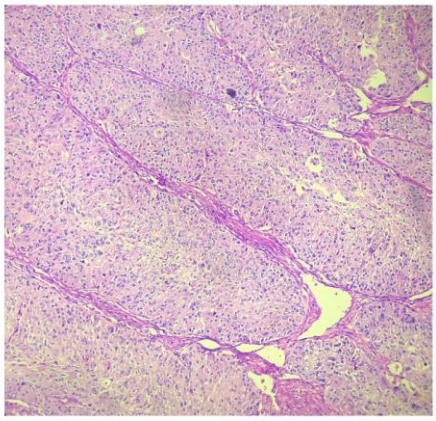
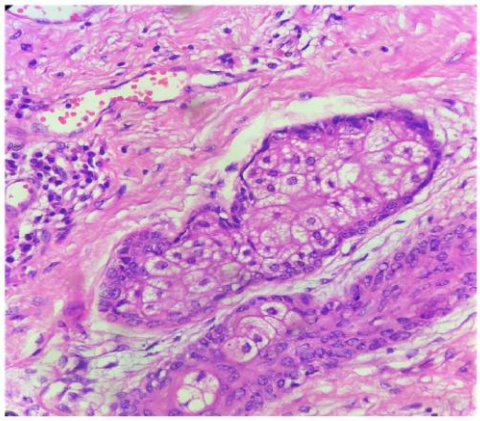


Figure 3

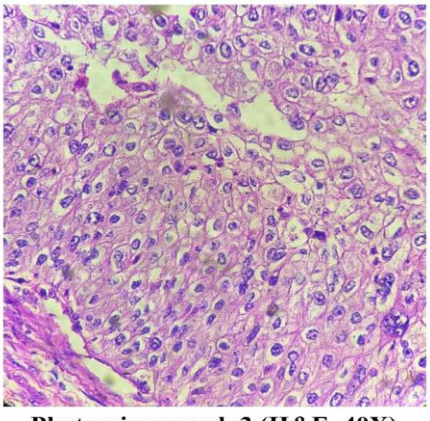
Microscopically, the sections studied revealed lobular arrangement of cells. The lobules were irregular, and highly variable in size comprising predominantly of basaloid and squamoid cells, along with the few cells in transition. (Photomicrograph 1) The basaloid cells were the undifferentiated cells arranged in periphery having slightly eosinophilic cytoplasm, pleomorphic nuclei and prominent nucleoli, while the squamoid cells were the differentiated sebaceous cells with more clear and foamy cytoplasm, having scalloped nuclei and few cells showing prominent nucleolus. Plenty of abnormal mitosis was also noted. (Photomicrograph 2 and 3) The intervening stroma is composed of eosinophilic, spindly, fibrotic hyalinized connective tissue. (Photomicrograph 4)



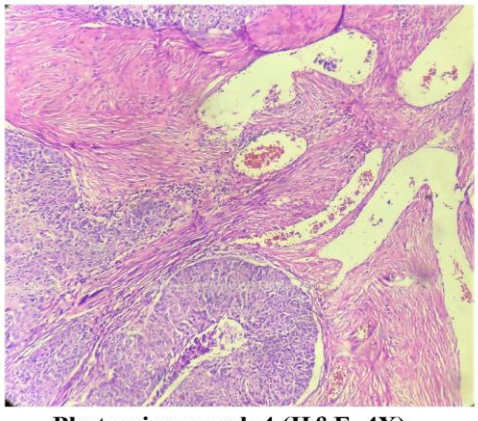
Photomicrograph 1 (H&E, 4X)



Photomicrograph 2 (H&E, 40X)

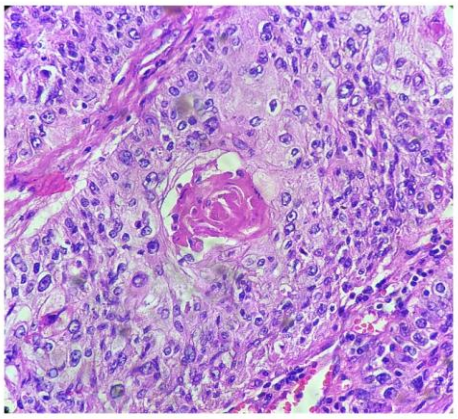


Photomicrograph 3 (H&E, 40X)

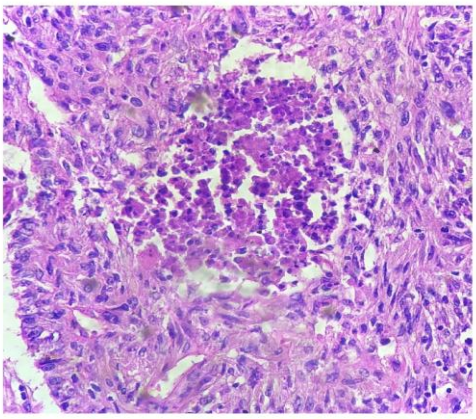


Photomicrograph 4 (H&E, 4X)

Few foci showed keratinizing cells arranged in loose clusters. (Photomicrograph 5) Also, noted were the areas of necrosis and plenty of inflammatory cell infiltrate. (Photomicrograph 6)



Photomicrograph 5 (H&E, 40X)



Photomicrograph 6 (H&E, 40X)

DISCUSSION

Scalp comprises a characteristic stratified structure consisting of epidermis, dermis, subcutis, epicranial aponeurosis, adjacent periosteum and the closely arranged adnexa such as sebaceous glands, hair follicles, eccrine and apocrine glands, surrounded by numerous blood vessels and lymphatics. [1] The tumours affecting scalp are mostly benign, only 1–2 % of scalp tumours are malignant. [1] Sebaceous carcinoma is a rare malignant tumour, commonly affecting adults with a slight female preponderance. [5] Although the aetiology is not known, hypothesis is that the tumour originates in the epithelium of sebaceous glands. And hence, this tumour can occur in any part of the body where sebaceous glands are present (face and scalp). [4,5,8] It may also be derived from a pluripotent cell able to differentiate into any cell line, including sebaceous cells. [4] Some relation has been shown to the germ line mutations in the MSH2 and MLH1 genes found on chromosomes 3p and 2p, respectively. [9] Associated risk factors are advanced age, Asian or South Asian race, previous irradiation, Muir-Torre syndrome, and immunosuppression following renal transplantation. [7,9] SC is classified into those arising from the ocular adnexa, and those arising in extraocular sites. Complete surgical excision is the treatment of choice. [11,12] It presents most commonly as a gradually enlarging, pink to red-yellow, firm, irregular, painless localised papular or nodular subcutaneous growth. [8,10] These are poorly circumscribed, deeply infiltrative, asymmetric tumor. [12] Definitive diagnosis is only by pathology, established via incisional or partial-thickness biopsy. [8,10] The presence of intracytoplasmic lipid by fat stains such as Oil Red O and Sudan IV may aid the diagnosis,[13] but requires frozen section.[3] More recently, antibodies to proteins associated with lipid droplets have also been identified such as adipophilin, perilipin and TIP47/PP17. [13] Common positive immunohistochemical markers include cytokeratin (CK), epithelial membrane antigen (EMA), Cam5.2 and anti-breast carcinoma associated antigen-225 antibody (anti-BCA-antibody-225). [3,10] Poor prognostic factors include multicentricity, size >1 cm in diameter, poor differentiation, extensive tissue infiltration, vascular or lymphatic involvement, pagetoid change, duration of more than six months and previous irradiation. [2,14] Although, distant metastases and recurrence rates are more common in the ocular type of sebaceous carcinoma, and not the extra-ocular type. [3,8]

On histopathology, sebaceous carcinoma is classified as well, moderately,

or poorly differentiated tumour. [1-3] The hallmark of sebocytic differentiation is the presence of multivesicular and vacuolated cytoplasm. [3] The characteristic histologic criterias are lobular architecture, foamy vacuolization of the cytoplasm, nuclear pleomorphism, hyperchromatic nuclei and mitotic activity. [2,4] The tumour is dermal, non-encapsulated and composed of clear cells with various levels of differentiations, [7] such as basaloid, squamoid, organoid, pseudo-neuroendocrine. [6,11] The tumour is composed of multiple irregular sebaceous lobules of varying sizes, [11] usually arranged in the dermis with a fibrovascular stroma.6 Less well differentiated cells contain a lipid-rich eosinophilic cytoplasm, giving a foamy appearance. [1,2] While, mature sebocytes may show multiple cytoplasmic vacuoles and scalloped nuclei. [11] These tumours present with a varied morphology and are considered great mimics of different types of neoplasms, which leads to the diagnostic difficulty. [6] As they possess subtle clinical features, they remain undiagnosed for years. [12] Previous studies have reported an incorrect initial histological diagnosis in 40–75 % of patients. [13] Recently, needle aspiration cytology procedure has been shown to be increasingly useful in establishing the diagnosis. [15]

CONCLUSION

The variations in the clinical picture as well as the histomorphology, often causes a delay in the diagnosis. Sometimes, there uncanny resemblance to benign pathology may possess unprecedented morbidity and mortality. Hence, owing to the rarity and diagnostic dilemma related to sebaceous carcinoma, a vigilant and careful approach is needed from the pathologist.

ACKNOWLEDGMENT

We would like to thank Department of Dermatology, for their contribution to this case.

Financial support and sponsorship: Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Prodinger CM, Koller J, Laimer M. Scalp tumors. JDDG: Journal der Deutschen Dermatologischen Gesellschaft. 2018 Jun;16(6):730-53.
2. Bhavarajua VM, Shamim SE, Naik VR, Shaari S. Sebaceous cell carcinoma of scalp– A rare presentation. The Malaysian journal of medical sciences: MJMS. 2007 Jan;14(1):67.
3. Natarajan K, Rai R, Pillai SB. Extra ocular sebaceous carcinoma: a rare case report. Indian dermatology online journal. 2011 Jul;2(2):91.
4. Cieza-Díaz DE, Cano-Martínez N, Barchino-Ortiz L, Longo-Imedio I. Extraocular sebaceous carcinoma: a report of 2 cases. Actas Dermo-Sifiliográficas (English Edition). 2012 Dec 1;103(10):919-22.
5. Dowerah S, Borgohain M. Sebaceous carcinoma of the scalp: Recurrence after treatment and utility of diagnostic cytology. Indian Journal of Dermatopathology and Diagnostic Dermatology. 2016 Jul 1;3(2):71.
6. Panjwani PK, Tirumalae R, Crasta JA, Manjunath S, Rout P. Extraocular sebaceous carcinoma: a series of three cases with varied presentation. Dermatology Practical & Conceptual. 2012 Jan;2(1):39.
7. Grigoryan KV, Leithauser L, Gloster HM. Aggressive extraocular sebaceous carcinoma recurring after mohs micrographic surgery. Case reports in oncological medicine. 2015 Jan 1;2015.
8. VimalReddy K, Yohesuwary G. Rare Tumour-Sebaceous Carcinoma of the Scalp.
9. Chitra Subramanian, Saravanan Krishnasamy, Ashoka Chakravarthi Dhamodaran, Manoharan Appavu. “A Rare Case of Sebaceous Carcinoma of Scalp with Parotid and Neck Secondaries”. Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 52, November 30, 2015; Page: 8699-8702, DOI: 10.18410/jebmh/2015/1207
10. Swali R, Nwannunu C, Tyring S. Sebaceous Carcinoma: A Rare Extraocular Presentation of the Cheek.

11. Chikhalkar S, Garg G, Gutte R, Khopkar U. Sebaceous carcinoma of scalp with proliferating trichilemmal cyst. Indian dermatology online journal. 2012 May;3(2):138.
12. Waqas O, Faisal M, Haider I, Amjad A, Jamshed A, Hussain R. Retrospective study of rare cutaneous malignant adnexal tumors of the head and neck in a tertiary care cancer hospital: a case series. Journal of medical case reports. 2017 Dec 1;11(1):67.
13. Muthusamy K, Halbert G, Roberts F. Immunohistochemical staining for adipophilin, perilipin and TIP47. Journal of clinical pathology. 2006 Nov 1;59(11): 1166-70.
14. Karkuzhali P, Gomathy N, Ahamed PB. Sebaceous carcinoma of cheek arising in a lesion of solar keratosis. Indian Journal of Dermatology. 2009 Jan 1;54(5):16.
15. Bailet JW, Zimmerman MC, Arnstein DP, Wollman JS, Mickel RA. Sebaceous carcinoma of the head and neck: case report and literature review. Archives of Otolaryngology–Head & Neck Surgery. 1992 Nov 1;118(11):1245-9.

How to cite this article: Rai A, Sahu S, Dhar R. Sebaceous carcinoma of scalp: a rare occurrence with a diagnostic challenge. *Int J Health Sci Res.* 2021; 11(2):100-104.

******