Content available at: iponlinejournal.com

IP Archives of Cytology and Histopathology Research

Journal homepage: www.innovativepublication.com

Case Report

Varied presentation of skin adnexal tumors - A case series with review of literature

Binjul Juneja[1], Shafaque Zabin[1], Mohd Talha[1], Kafil Akhtar[1,*]

[1] *Dept of Pathology, J. N. Medical College - Aligarh Muslim University, Aligarh, Uttar Pradesh, India*

ARTICLE INFO

Article history:
Received 02-01-2020
Accepted 23-01-2020
Available online 18-03-2020

Keywords:
Adnexal Tumours
Benign
Histopathology
Malignant
Skin

ABSTRACT

Skin adnexal tumours are a heterogeneous group of uncommon and rare tumours usually misdiagnosed clinically due to different types and similar presentation. They include a diverse group of benign and malignant neoplasms. They may present as single or multiple lesions and can be sporadic or familial in inheritance. They might be a superficial manifestation of an internal malignancy. Benign lesions are more common but malignant tumours are rare and locally aggressive, with potential nodal and distant metastasis with a poor clinical outcome. Histopathology is the basis of critical diagnosis, which has therapeutic and prognostic implications. Here we report a case series of skin adnexal tumours with aggressive behaviour on histopathology which appeared benign clinically. This case series emphasises the need for early detection and proper follow up of skin adnexal tumours to reduce the morbidity and mortality.

1. Introduction

Skin adnexal tumours represent only 1.0 to 2.0% of skin lesions. They have a varied presentation and may show the tumoral proliferation of the pilosebaceous unit, apocrine and eccrine sweat glands, though some tumours may show elements of mixed differentiation.[1,2] The tumor characteristics may be due to their origin from pluripotent stem cells.[2,3]

Skin adnexal tumours may be papular or nodular firm, surface lesions but with distinct histological features.[4] Most of the tumours are benign and may be superficial manifestations of some syndromes associated with internal malignancies, such as trichilemmomas in Cowdens disease and sebaceous tumours in Muir –Torre syndrome.[5]

Benign adnexal tumours usually have a malignant neoplastic counterpart. Although malignant tumours are rare; they are highly aggressive, with a poor clinical course. They have the clear potential for nodal and distant metastasis.[1] Therefore, establishing the diagnosis of malignancy in any skin adnexal tumour is of utmost therapeutic and prognostic importance.

2. Case Summary

2.1. Case 1

A 53 -year-old female presented to the Dermatology outpatient department with complaints of swelling and pain i n her right eyelid for 3 years. On examination, the mass was firm and solid, 2 x 3 cm in size. It was excised completely and on microscopic examination, large round to polygonal cells arranged in trabeculae, nests and cords was seen in a hyalinized stroma, with abundant clear to granular cytoplasm and oval hyperchromatic nuclei with prominent nucleoli. A diagnosis of poorly differentiated sebaceous cell carcinoma was given (Figure 1). The patient was administered adjuvant radiotherapy of 5 0 Gy in 30 fractions. She developed recurrence after 8 months of completion of therapy with cervical lymph node involvement. Subsequently she was given chemotherapy (Cisplatin 50mg/m^2 x 6 cycles) and irradiation with 60 Gy in 30 fractions. She did not tolerate the therapy and succumbed to her illness.

* *Corresponding author*.
E-mail address: drkafilakhtar@gmail.com (K. Akhtar).

https://doi.org/10.18231/j.achr.2020.020

2.2. Case 2

A 65-year-old male presented to the Dermatology outpatient clinic with multiple white patches in the buccal mucosa for the last 4 years and swelling on his scalp for last 3 months. He had history of tobacco consumption for the last 20 years. CT scan of the head showed heterogenous mass in the scalp with intracranial extension and involvement of the parietal bone. Biopsy from both oral cavity and scalp lesion showed features of basaloid squamous cell carcinoma (Figure 2) Due to inoperability of the lesion, he was given radiotherapy of 60 Gy in 30 fractions and cisplatin (50 mg/m^2 x 6 cycles) based chemotherapy. There after he developed metastasis to the lumbar vertebrae after a year of therapy and died subsequently.

2.3. Case 3

A 42 year old male presented to the surgery outpatient clinic with recurrent ulcer on the scalp for the last 3 years. Local examination revealed a 5x4x3 cm fungating ulcer over the right occiput. There was no tenderness at the local site. There was no enlarged cervical lymph nodes noted and systemic examinations were no n-significant with no features of raised intracranial pressure. Wide local excision of the lesion was performed. Histopathologic feature was consistent with eccrine porocarcinoma with positive deep resection margin (Figure 3). On post-operative follow up after 6 months, the disease showed recurrence of the ulcerative growth. Computed tomography was performed which showed an ill-defined soft tissue mass at the primary site with metastasis to the right posterior cervical lymph nodes . Fine needle aspiration cytology from the cervical lymph node showed metastatic deposits. The patient underwent repeat wide local excis ion with posterior neck node dissection. There was microscopic evidence of porocarcinoma in the resected specimen with metastasis to cervical nodes. Concurrent chemo-radiotherapy in dose of 50 Gy of Co-60 teletherapy in 30 fractions and 50 mg/m^2 of Cisplatin for 6 cycles was administered . Our patient is doing well after 12 months of follow up period.

2.4. Case 4

A 67 years old male presented to the surgery outpatient clinic with complaints of bleeding ulcer on the right eye. On examination, a 5 $\times$ 6 cms excoriated ulcer with everted edges was seen in the right lower eyelid. There were no enlarged regional lymph nodes in the neck. There was a past history of similar lesion on the nose 5 years back, for which he was operated. There was also a history of shortness of breath and few episodes of altered sensorium for the last one month. Computed tomogram of the head was performed which showed a 3x 5 mm heterogenous mass in the left frontal lobe with perilesional edema. The chest radiograph showed a well defined radio-opaque mass in the left upper lobe of the lung. The liver function test and other routine blood investigations were within normal limits. Excision biopsy of the eyelid lesion and CT guided biopsy of the left lung lesion was performed. Microscopic examination of the facial lesion showed dysplastic stratified squamous epithelium with irregular nests of small uniform round to oval tumor cells with hyperchromatic nucleus and scant cytoplasm with nuclear palisading in the underlying stroma. Histopathological features were consistent with basal cell carcinoma (Figure 4). CT guided biopsy from the lung lesion showed foci of metastatic basal cell carcinoma. Prognosis of the disease was explained to the patient and he is presently on palliative radiotherapy and chemotherapy.

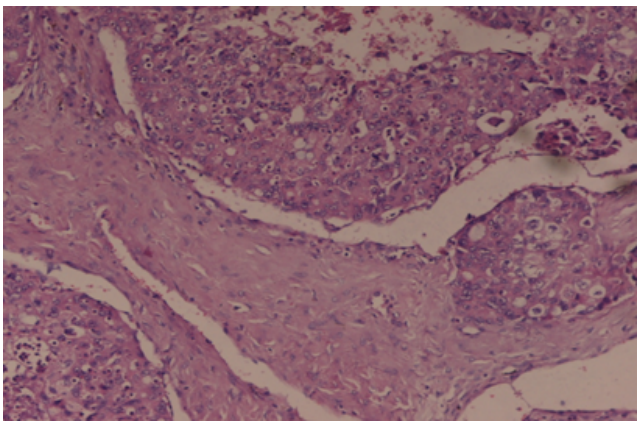

Fig. 1: Sebaceous Carcinoma: Microscopic examination shows large round to polygonal tumor cells arranged in trabeculae, nests and cords in a hyalinized stroma, with abundant clear to granular cytoplasm and oval hyperchromatic nuclei with prominent nucleoli. Haematoxylin and Eosin, x 40.

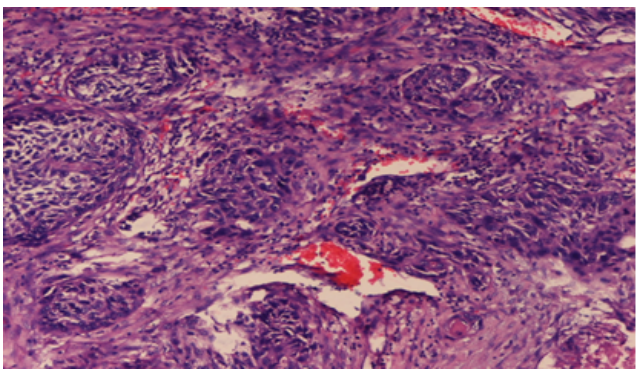

Fig. 2: Squamous cell carcinoma: Sections shows solid nests of large round to polygonal tumor cells of varying sizes with a squamous differentiation with hyperchromatic nucleus and well defined eosinophilic cytoplasm with distinct intercellular bridges and evident foci of keratinization. Haematoxylin and Eosin, x 40.

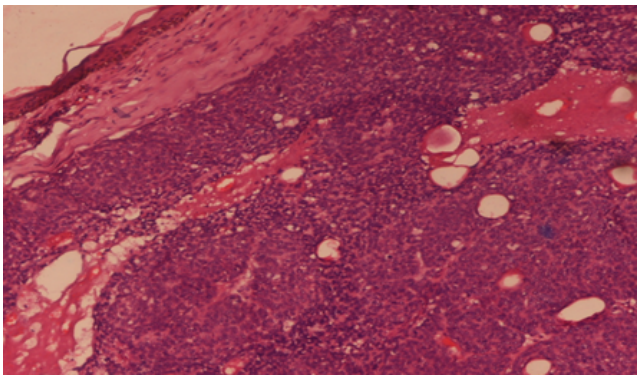

Fig. 3: Porocarcinoma: Microscopically tissue section shows atypical tumor cells arranged in cords and lobules, with marked anisonucleosis, with foci of necrosis and high mitotic activity. Haematoxylin and Eosin, x10.

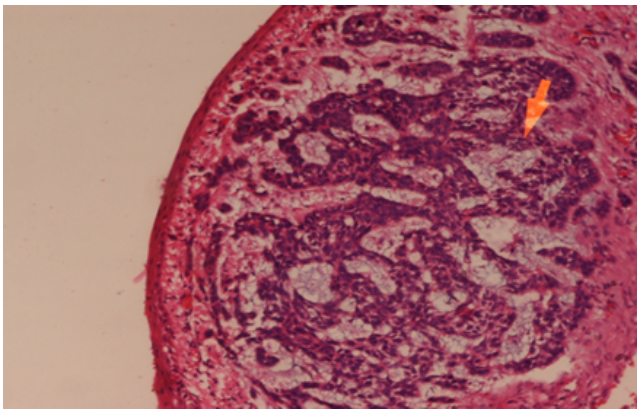

Fig. 4: Basal cell carcinoma: Tissue section shows dysplastic stratified squamous epithelium with irregular nests of small uniform round to oval tumor cells with hyperchromatic nucleus and scant cytoplasm with nuclear palisading in the underlying stroma. Haematoxylin and Eosin, x10.

3. Discussion

The benign tumours of epidermal appendages are a result of the controlled proliferation of the hair follicles, sebaceous glands, apocrine glands and eccrine glands.[2,3] The malignant adnexal tumours mainly arise from the malignant transformation of the sebaceous glands, eccrine glands and apocrine glands.[2,4] Few cases of malignant tumours of hair follicles have also been reported such as pilomatrical carcinoma, malignant proliferating trichilemmal cyst and trichilemmal carcinoma.[6,7] Varied presentation of the lesions, huge number of tumors with variants and complicated nomenclature of skin adnexal tumours are a diagnostic dilemma.

Adnexal tumours originate from multipotent stem cells rather than mature cells, which have the capability to differentiate along a particular pathway.[6,7] Among the adnexal tumours, the most frequent line of differentiation is sweat gland differentiation (56 .0%), followed by hair follicle differentiation (28.0%) and the least frequent being sebaceous gland differentiation (16.0%).[8] The overall incidence of skin adnexal tumours in India is very low.[1,2] Head and neck region is the most common location of adnexal tumours, followed by the axilla, trunk and the legs.[8,9]

Sebaceous carcinoma is a rare malignant neoplasm of adult males.[10] Usually presents as peri-ocular exfoliative sebaceous neoplasms, with the upper eyelid more often affected than the lower due to numerous meibomian glands.[10,11] Extraocular sebaceous carcinoma is seen more often in the head and neck region followed by trunk, salivary glands, genitalia, breast, ear canal and the oral cavity.[7,8] The usual clinical presentation is a painless subcutaneous nodular growth.

The characteristic histologic feature of sebaceous cell carcinoma is the presence of large sebaceous tumor cells, with vacuolated cytoplasm filled with fat.[10] The main differential of sebaceous cell carcinoma is basal cell carcinoma with sebaceous differentiation, which typically shows peripheral palisading and clefting from the adjacent stroma. Our case did not show small round to oval dense basaloid cells with palisading as seen in basal cell carcinoma. Immunohistochemistry shows diffuse strong cytokeratin express ion and focal EMA positivity in tumor cells with sebaceous differentiation.[9,10] Based on the histomorphological and immunohistochemical findings, a diagnosis of sebaceous carcinoma was given by us. 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.[5,10] Distant metastases and recurrence rates are more common in the ocular type of sebaceous carcinoma, as was evident in our case. Wide surgical excision and radiotherapy is the treatment of choice, though mortality rates are as high as 50.0%.[9,11]

Basaloid squamous cell carcinoma (BSCC) is an aggressive variant of squamous cell carcinoma. Occurs in the head and neck region, is usually multifocal, infiltrative in nature with high incidence of metasta sis at initial presentation.[6,7] This tumor has more common in males, with history of tobacco and alcohol a ddiction. Human papilloma virus and herpes simplex virus and previous history of head and neck radiation have also been implicated in the etiology of BSCC.[12] The tumors ha ve an origin from the totipotent basal epithelial cell s of skin surface and the minor salivary glands of the submucosa.[13]

BSCC usually presents as an ulcerated mass with extensive submucosal induration, which often can be confused with a minor salivary gland tumor. Histologically, round to oval dark basaloid cells coexists with a squamous cell carcinoma component which can be either

in situ carcinoma or invasive keratinizing squamous cell carcinoma.[14] The lobular configuration of the basaloid cells are seen in solid sheets with prominent peripheral palisading. They have a propensity to invade the soft tissue and bones, but brain metastases a re extremely rare.[12,13] Liver, lungs and bones are the sites of distant spread.[14] Therefore, preoperative tumour staging with CT scan of the thorax, radionuclide bone scans and ultrasound of the liver is usually recommended. In our case, the patient had histologically confirmed brain metastasis of BSCC.

Eccrine porocarcinoma are rare malignant tumors of sweat gland, first described by Pinkus and Mehregan in 1963.[7] Mishisma and Moriko gave the term "eccrine porocarcinoma" in 1969.[15]Porocarcinoma occurs in the elderly individuals of both sexes. It presents as an ulcerated nodular growth or plaque like lesions in the lower extremities, trunk and head and neck. Studies suggest that this cancer arises from eccrine poroma with rare bone involvement.[6,7] Microscopically tissue section shows atypical tumor cells in cords and lobules with marked anisonucleosis with foci of necrosis and high mitotic activity, infiltrating the dermal stroma.

Wide excision or Mohs micrographic surgery is the treatment of choice, but eccrine porocarcinoma have a high recurrence rate of 20.0%.[7,15]Regional lymph nodes should be assessed, as porocarcinoma spread by lymphatics in about 20% of the cases.[15]The infiltrative nature of the disease has been responsible for high local recurrence rates. So adjuvant radiotherapy is advised to reduce local recurrence. In our patient wide local excision was done with lymph node dissection and adjuvant radiotherapy was given in view of extra capsular extension and positive resected margin even after re-excision. Chemotherapy is mainly reserved for metastatic disease.[15] Intra-lesional photodynamic therapy can be an alternative mode of therapy.[6]Lymph node and distant metastasis are associated with poor prognosis.[15]

Basal cell carcinoma is a slow growing but aggressive skin neoplasm when giant lesions are involved.[16] It is most commonly seen in the exposed area of the head and neck but few cases have been reported in the unexposed areas of the genitalia.[16] Metastatic BCC is extremely rare and are almost fatal. The associated risk factors of metastatic basal cell carcinoma are long duration of disease, large size, multiple lesions, increased depth of invasion, with infiltrative margins and incomplete surgical resection and history of previous radiation therapy.[16–18] The superficial, infiltrative and micronodular histological subtypes of BCCs have a high propensity for recurrence and metastasis.[18] Metastasis to the regional lymph nodes, lung and bone are common. But secondaries may be seen in the spinal cord, parotid gland, bone marrow, spleen, liver, brain and the kidney.[17,18] In our case of metastatic BCC, the primary lesion was significantly large.

Treatment of metastatic BCC consists of wide surgical excision of the prmary with concurrent chemo -irradiation therapy. Combination chemotherapy with cisplatin, cyclophosphamide, vincristine and bleomycin has shown significant response.[17,18]The prognosis is poor with a mean survival time of only eight months from the time of diagnosis.[17,18]

4. Conclusions

This case series highlights multiple aspects of malignant adnexal carcinomas, such as clinical aspects, treatment modalities used, locoregional failure and histopathological diversity and emphasizes the need for early detection of metastatic adnexal tumours so as to reduce the morbidity and mortality associated with it. But it has a few limitations such as short follow-up and a small study sample to comment on the epidemiology of the disease. None of the patients was treated with definitive radiotherapy to evaluate its role as a single modality of therapy. Studies from multiple centers will help to establish guidelines to address these rare but aggressive tumors.

5. Source of funding

None.

6. Conflict of Interest

None.

References

1. Guerrissi JO, Quiroga JP. Adnexal carcinomas of the head and neck. *Indian J Plast Surg*. 2008;41(02):229–234.
2. Radhika K, Phaneendra BV, Rukmangadha N, Reddy MK. A study of biopsy confirmed skin adnexal tumours: experience at a tertiary care teaching hospital. *J Clin Scientific Res*. 2013;2:132–138.
3. S SG, Alavandar E, S AK. AN ANALYSIS OF SKIN APPENDAGEAL TUMORS IN SOUTH INDIA. *Journal of Evolution of Medical and Dental Sciences*. 2012;1(6):907–912. Available from: https://dx.doi.org/10.14260/jemds/145. doi:10.14260/jemds/145.
4. Pantola C, Kala S, Agarwal A, Amit S, Pantola S. Cutaneous adnexal tumours: A clinicopathological descriptive study of 70 cases. *World J Pathol*. 2013;2:77–82.
5. Lazar AJ, Lyle S, Calonje E. Sebaceous neoplasia and Torre–Muir syndrome. *Curr Diagn Pathol*. 2007;13(4):301–319.
6. Das N, Gharami R, Chowdhury S, Datta P, Saha A. A clinico-histopathological study of appendageal skin tumors, affecting head and neck region in patients attending the dermatology OPD of a tertiary care centre in Eastern India. *Indian Journal of Dermatology*. 2011;56:33–36.
7. Jindal U, Patel R. Study of adnexal tumors of the skin: A three year study of 25 cases. *Int J Pathol*. 2012;p. 13–33.
8. Shields JA, Demirci H, Marr BP, Eagle RC, Shields CL. Sebaceous Carcinoma of the Ocular Region: A Review. *Surv Ophthalmol*. 2005;50:103–122.
9. Callahan EF, Appert DL, Roenigk RK, Bartley GB. Sebaceous carcinoma of the eyelid: A review of 14 cases. *Dermatol Surg*. 2004;30:1164–1168.
10. Schwartz RA, Torre DP. The Muir-Torre syndrome: A 25-year retrospect. *J Am Acad Dermatol*. 1995;33(1):90–104.

11. Dowd MB, Kumar RJ, Sharma R, Murali R. DIAGNOSIS AND MANAGEMENT OF SEBACEOUS CARCINOMA: AN AUSTRALIAN EXPERIENCE. *ANZ J Surg*. 2008;78(3):158–163.
12. Radhi J. Basaloid squamous cell carcinoma. *Coratia: Intech publications*. 2012;p. 132–137.
13. Gopinath D, Beena VT, Stephen M, Sivakumar R, Choudhary K. Basaloid squamous cell carcinoma of oral cavity: Report of two cases. *Int J Dent Clin*. 2012;4:62–64.
14. Ruzevick J, Olivi A, Westra WH. Metastatic squamous cell carcinoma to the brain: an unrecognized pattern of distant spread in patients with HPV-related head and neck cancer. *J Neuro-Oncol*. 2013;112(3):449–454.
15. Mishima Y, Morioka S. Oncogenic differentiation of the intraepidermal eccrine sweat duct: eccrine poroma, poroepithelioma and porocarcinoma. *Dermatol*. 2009;138:238–250.
16. Aldhaban S, Marc S, Eshki M, Girod A, Boissonet H, et al. Giant basal cell carcinoma with regional lymph node and distant lung metastasis. *Eur J Dermatol*. 2011;21(6):972–975.
17. Mehta KS, Mahajan VK, Chauhan PS, Sharma AL, Sharma V, et al. Metastatic Basal Cell Carcinoma: A Biological Continuum of Basal Cell Carcinoma? *Case Rep Dermatol Med*. 2012;4:221–224.
18. Vu A, Laub D. Metastatic basal cell carcinoma. In: Madan V, editor. Basal Cell Carcinoma. Croatia: In Tech ; 2012,. p. 79–92.

Author biography

Binjul Juneja Resident

Shafaque Zabin Resident

Mohd Talha Resident

Kafil Akhtar Professor

Cite this article: Juneja B, Zabin S, Talha M, Akhtar K. Varied presentation of skin adnexal tumors - A case series with review of literature. *IP Arch Cytol Histopathology Res* 2020;5(1):99-103.