

Squamoid Eccrine Ductal Carcinoma: A Diagnostic Dilemma

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ABSTRACT

Eccrine carcinomas comprise less than 0.01% of all cutaneous malignancies. Squamoid eccrine ductal carcinoma is an exceedingly rare variant that exhibits both squamous and eccrine ductal characteristics. It presents in patients older than 50 years on the head, neck, and trunk as nodule or plaque. It is commonly misdiagnosed as squamous cell carcinoma from which it must be differentiated as its recurrence and metastasis rates are much higher. Very few cases have been reported, we hereby report an additional case of this rare tumor.

A 50 years old male presented with a slow-growing nodule in the frontal region of scalp since one year. Histopathology showed a dermal based infiltrative epithelial tumor with extensive squamous differentiation arranged in interlacing cords and nests, with focal formation of ductal lumina which was highlighted by immunohistochemistry. The diagnostic identification of eccrine carcinoma is hampered by their rarity. Awareness of this rare entity will help in accurate diagnosis and management.

Keywords: Squamoid, Eccrine, Ductal, Carcinoma, p63, CEA

Introduction

Eccrine carcinomas comprise less than 0.01% of all cutaneous malignancies. [1] Squamoid eccrine ductal carcinoma is an exceedingly rare variant of eccrine carcinomas that exhibits both squamous and eccrine ductal characteristics. [2] It is commonly misdiagnosed as squamous cell carcinoma from which it must be differentiated as its recurrence and metastasis rates are much higher. [3] Very few cases have been reported, we hereby report an additional case of this rare tumor.

Case Report

A 50 years old male presented with a slow growing hard nodule measuring one centimetre in diameter in the frontal region of scalp since one year. The skin over the nodule showed hypopigmentation with telangiectasia and was not pinchable. A superficial incisional biopsy was taken with a clinical diagnosis of benign appendageal tumor. Histopathology showed cords of atypical cells infiltrating the dermis with no specific differentiation. Excisional biopsy was advised with possible differentials of malignant appendageal or metastatic tumor. Histopathology showed a dermal based infiltrative epithelial tumor with extensive squamous differentiation arranged in interlacing cords and nests with intervening fibro-myxoid stroma [Figure 1]. Cells were polygonal, had moderate amount of eosinophilic cytoplasm, moderately pleomorphic nuclei with prominent nucleoli. Frequent mitoses were seen. Foci of squamoid differentiation were noted in the form of intracytoplasmic keratin and intercellular bridges

[Figure 2a]. Few cells showed intracytoplasmic vacuoles containing eosinophilic material pushing the nucleus to periphery [Figure 2b]. Occasional duct formation was noted on careful examination. Some of the nests showed central necrosis along with individual cell necrosis. A grenz zone separated the tumor from the overlying epidermis. The overlying epidermis was unremarkable and showed no connection with the tumor on serial sectioning. The tumor was extending into the subcutis and was reaching close to deep resected margin. However, the lateral margins were free. Immunohistochemistry showed positive nuclear staining for p63 [Figure 3a] favouring cutaneous primary and luminal positivity for CEA [Figure 3b] highlighting the inconspicuous ductal lumina. There is no evidence of recurrence or metastasis at five months follow-up.

Discussion

Eccrine carcinomas are uncommon tumors of eccrine glands as was highlighted in a review at Mayo clinic wherein they reported only 14 tumors in a span of 75 years. [4] The diagnostic identification of eccrine carcinomas is hampered by their rarity. Myriad of clinical presentations plus a lack of uniform classification further add to difficulties in diagnosis. [5]

It presents in patients older than 50 years on the head, neck, and trunk with varied appearances such as nodule, plaque, exophytic growth or completely benign looking lesion. [2,3] However, the commonest clinical presentation is that of a nodule on the scalp as was seen in our case too. Histologically, this tumor presents as an infiltrative

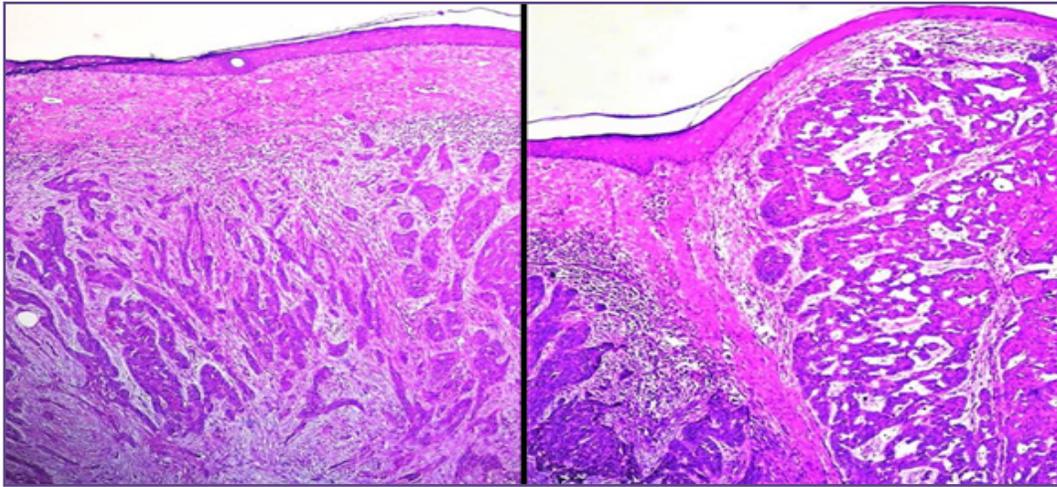


Fig. 1: Dermal based infiltrative epithelial tumor arranged in intertwinning cords and nests [H&E, 100x].

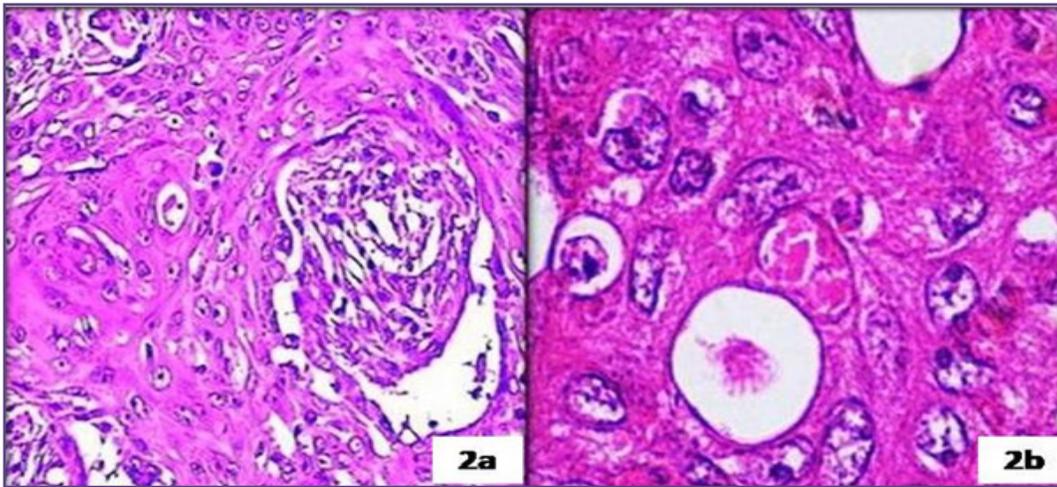


Fig. 2a: Tumor showing squamous differentiation [H&E, 400x], Figure 2b - Tumor cells showing intracytoplasmic vacuoles [H&E, 1000x].

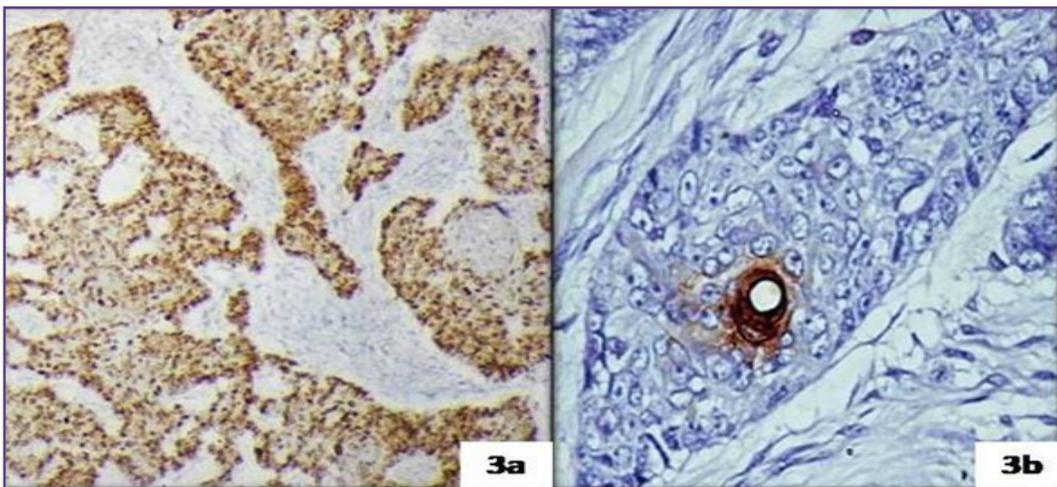


Fig. 3a: Diffuse nuclear p63 positivity [H&E, 200x], Figure 3b - CEA highlighting the ductal lumina formation [H&E, 200x].

neoplasm extending into deep dermis or subcutis. There is prominent squamoid differentiation in the superficial aspect of the tumor whereas deeper reaches of the tumor show ductal differentiation in the form of eosinophilic cuticle lined lumina.^[6] Our case also showed squamous differentiation and glandular differentiation in the form of both ductal lumina and intracytoplasmic eosinophilic vacuoles. The cytoplasmic features like intracytoplasmic vacuoles were distinctive and have not been described by other authors. Also some of the morphological features in our case were divergent from other reported cases like lack of epidermal continuity and intimate admixture of squamous and ductal components. Similar divergent features have been reported previously in only one case report by Magro et al.^[7]

Histological differential diagnoses include microcystic adnexal carcinoma with squamous features, squamous cell carcinoma and metastasis.^[2,8] Microcystic adnexal carcinoma shows presence of many keratinous cysts. Deeper components include smaller nests and strands of cells. A dense fibrous stroma surrounds all components which was loose fibromyxoid in our case.^[5,8] Squamous cell carcinoma is differentiated on the basis of epidermal involvement and lack of ductal differentiation which can be confirmed on immunohistochemistry for CEA and EMA. Recent studies have shown that p63 is a reliable marker to differentiate primary adnexal sweat gland carcinomas from metastatic tumors.^[9] Our case showed diffuse p63 immunoreactivity. Additionally, radiologic screen and clinical evaluation ruled out the possibility of metastasis.

Squamoid eccrine ductal carcinoma is noted to be locally aggressive and frequently courses along perineural tissue.^[3] Reports of local recurrence rates after surgical excision are up to 70%.^[1] However, Mohs micrographic excision has been found to reduce the recurrence rates. It has been proposed that FFDG PET/CT imaging is indicated to stage patients with a diagnosis of SEDC, based on the high likelihood for metastatic disease.^[10] Due to rarity of this tumor, no definite treatment protocol exists.

Conclusion

Squamoid eccrine ductal carcinoma closely mimicks squamous cell carcinoma. It can have benign clinical presentation further adding to misdiagnosis. To conclude, this case highlights many new must to be known facts about this rare tumor like ductal differentiation may be inconspicuous unless highlighted using immunohistochemistry, epidermal connection of the tumor may be absent and squamoid and ductal components may be closely intermingled. Review by an experienced dermatopathologist and awareness about this rare entity will help in accurate diagnosis and management.

References

1. Wick MR, Swanson PE. Cutaneous adnexal tumors: a guide to pathological diagnosis. Chicago: American Society of Clinical Pathologists Press; 1991. p. 10.
2. Jung YH, Jo HJ, Kang MS. Squamoid eccrine ductal carcinoma of the scalp. *Korean J Pathol* 2012; 46(3): 278-81.
3. Terushkin E, Leffell DJ, Futoryan T, Cowper S, Lazova R. Squamoid eccrine ductal carcinoma: a case report and review of the literature. *Am J Dermatopathol* 2010 ;32(3):287-92.
4. Wick MR, Goellner JR, Wolfe JT 3rd, Su WP. Adnexal carcinomas of the skin. *Eccrine carcinomas*. *Cancer* 1985 ;56(5):1147-62.
5. Wildemore JK, Lee JB, Humphreys TR. Mohs surgery for malignant eccrine neoplasms. *Dermatol Surg* 2004 ;30:1574-9.
6. Herrero J, Monteagudo C, Jordá E, Llombart-Bosch A. Squamoid eccrine ductal carcinoma. *Histopathology* 1998 ;32(5):478-80.
7. Magro C, Glass S. Squamoid Eccrine Ductal Carcinoma. *The Dermatologist* 2016 ;24(5)
8. Antley CA, Carney M, Smoller BR. Microcystic adnexal carcinoma arising in the setting of previous radiation therapy. *J Cutan Pathol* 1999; 26: 48–50.
9. Qureshi HS, Ormsby AH, Lee MW, Zarbo RJ, Ma CK. The diagnostic utility of p63, CK5/6, CK 7, and CK 20 in distinguishing primary cutaneous adnexal neoplasms from metastatic carcinomas. *J Cutan Pathol* 2004; 31: 145–52.
10. Wang B, Jarell AD, Bingham JL, Bonavia GH. PET/CT imaging of squamoid eccrine ductal carcinoma. *Clin Nucl Med* 2015 ;40(4):322-4.

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