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Microcystic adnexal carcinoma of the cheek—a case report with dermatoscopy and dermatopathology

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ABSTRACT We present a case report of a microcystic adnexal carcinoma on the cheek of a 67-year-old man. Clinical, dermatoscopic and dermatopathologic images are presented. A search of the literature has not discovered any previously published dermatoscopy images of microcystic adenexal carcinoma.

Introduction

Microcystic adnexal carcinoma (MAC) is a rare malignant cutaneous neoplasm with pilar and eccrine gland differentiation. MAC was first described as a separate clinical entity by Goldstein et al in 1982 [1]. It is locally aggressive but rarely metastasizes, usually presenting as a slow growing asymptomatic lesion on the head and neck. MAC arises from pluripotent keratinocytes that possess the capability for adnexal differentiation. Predisposing factors include exposure to UV radiation, immunosuppression, and history of radiotherapy [2]. Fewer than 300 cases have been reported worldwide, according to an analysis by Yu et al [3].

Case presentation

A 67-year-old man presented to a primary care skin cancer clinic in Melbourne, Australia for a routine six-month skin cancer examination. There was a long history of recreational sun exposure. His brother had a history of melanoma of his thigh in his 40s. Seven separate basal cell carcinomas had required excision from his forehead, nose, pinna, posterior neck, mid back calf in the last decade. Most recently a moderately differentiated squamous cell carcinoma had been excised from his right upper forehead some six months previous.

A whole body skin examination was undertaken with the aid of a Heine Delta 20 non-polarizing dermatoscope (Heine Optotechnik, Herrshing, Germany). Digital clinical and dermatoscopic images were taken with a Medicam 800 Fotofinder non-polarizing camera (Fotofinder Systems GmbH, Aichner, Birnbach, Germany), the dermatoscopy images being at 20x magnification. Examination confirmed Fitzpatrick skin type 2 with severe actinic damage to the skin of his face, upper trunk and distal limbs with multiple solar lentigines and actinic keratoses. Significant actinic damage to the lower lip (actinic cheilitis) was apparent.

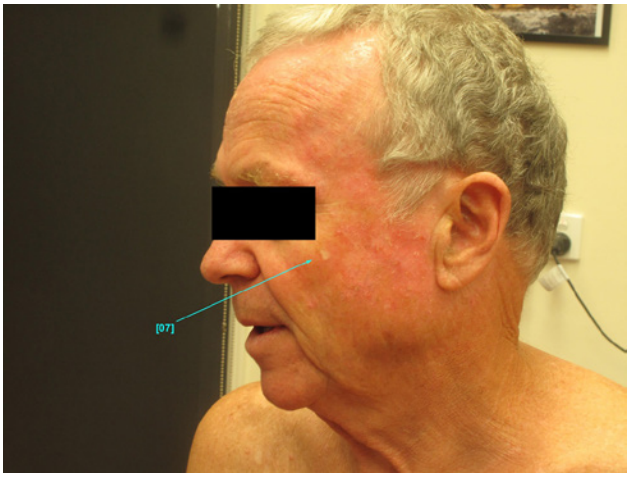

Figure 1. Clinical image of a non-pigmented skin lesion on the left mid cheek of a 67-year-old man. [Copyright: ©2014 Inskip, Magee.)

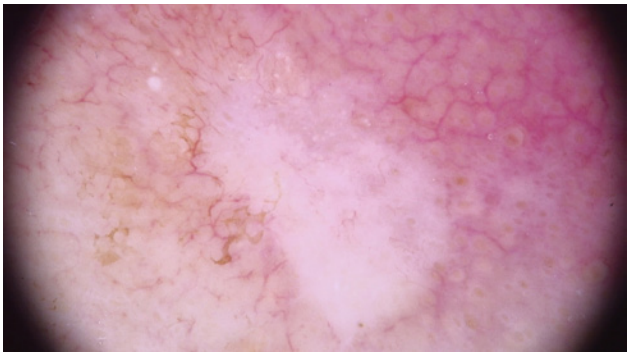

Figure 2. Dermatoscopy showing a dense white structureless area with fine linear branched blood vessels centrally. Note the white clods of variable diameter superiorly. (Copyright: ©2014 Inskip, Magee.)

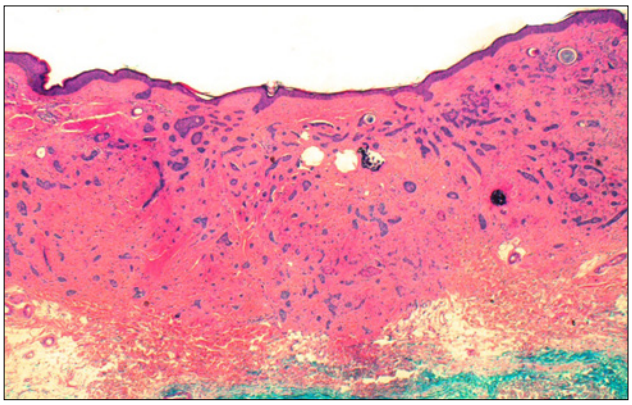

Figure 3. Low power view demonstrates a poorly circumscribed, deeply invasive, infiltrative neoplasm, with superficial cyst formation and calcification, and a more morphoeiform desmoplastic deep component (Copyright: ©2014 Inskip, Magee.)

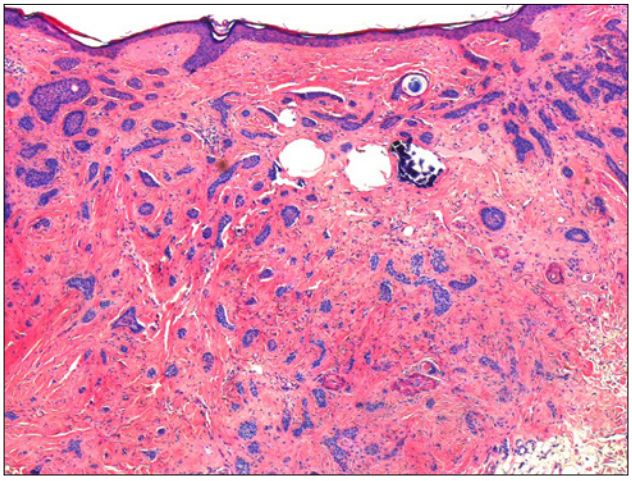

Figure 4. Intermediate power image shows superficial follicular differentiation with cysts and calcification, and more infiltrative deep component. (Copyright: ©2014 Inskip, Magee.)

During the examination the patient pointed out a white, scar-like lesion on his mid left cheek measuring 8 x 4 mm in diameter. It was non-pigmented and was composed of a clearly demarcated flat white plaque (Figure 1). Dermatoscopically the lesion exhibited a dense white structureless area with fine linear branched blood vessels centrally. A notable feature was the white clods of variable diameter superiorly (Figure 2). Differential diagnosis included morphoeic or fibrosing basal cell carcinoma and desmoplastic trichoepithelioma [4]. Eccrine syringoid carcinoma, a rare malignant cutaneous adnexal tumor, should also be included in the differential diagnosis [5].

An excisional biopsy was performed using an elliptical excision, and the specimen was submitted for assessment by a specialist dermatopathologist.

Histology

Examination of the histological sections revealed a deeply invasive dermal neoplasm composed superficially of keratin filled cysts with calcification, and in the underlying reticular dermis, of infiltrative aggregates of basaloid cells in slender strands and syringomatoid aggregates. These extended to the subcutis. There were areas suspicious for small nerve invasion present within the tumor. Excision appeared complete.

The differential diagnosis was between a microcystic adnexal carcinoma and a fibrosing basal cell carcinoma with follicular differentiation. Immunohistochemical stains were performed. The lesion stained positively for Ber-EP4. There was also strong CK15 positivity. CEA stained some lumina within the lesion. This staining pattern, although not entirely specific, was more in favour of microcystic adnexal carcinoma than fibrosing basal cell carcinoma. Ber-EP4 expression has been noted in 38% of microcystic adnexal carcinomas and 100% of basal cell carcinomas. However CK15 is expressed in 92% of microcystic adnexal carcinomas, whereas basal cell carcinomas are negative [6]. Hence, in conjunction with the positive CEA, the findings favoured a microcystic adnexal carcinoma (Figures 3 to 11).

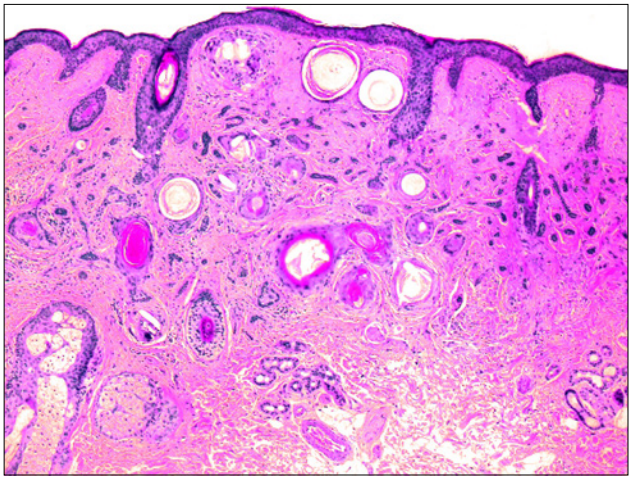

Figure 5. Intermediate power of superficial follicular keratin-filled cysts. (Copyright: ©2014 Inskip, Magee.)

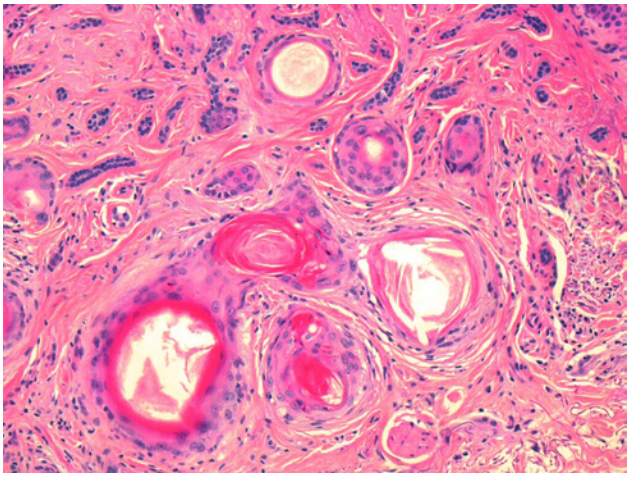

Figure 6. Intermediate power of superficial follicular keratin-filled cysts. (Copyright: ©2014 Inskip, Magee.)

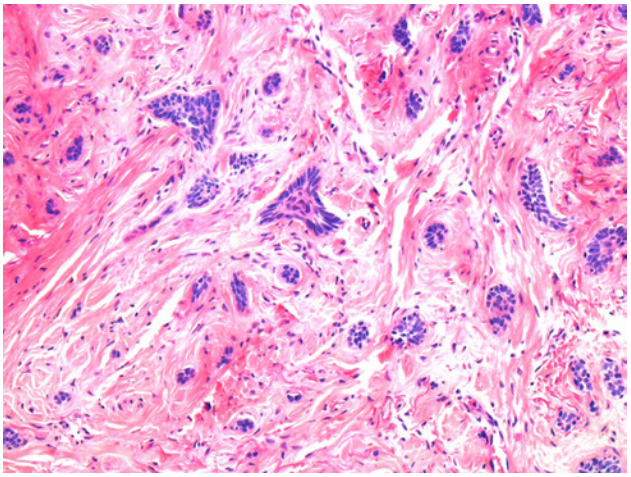

Figure 7. High power of basaloid infiltrative aggregates in deeper portion of tumor .(Copyright: ©2014 Inskip, Magee.)

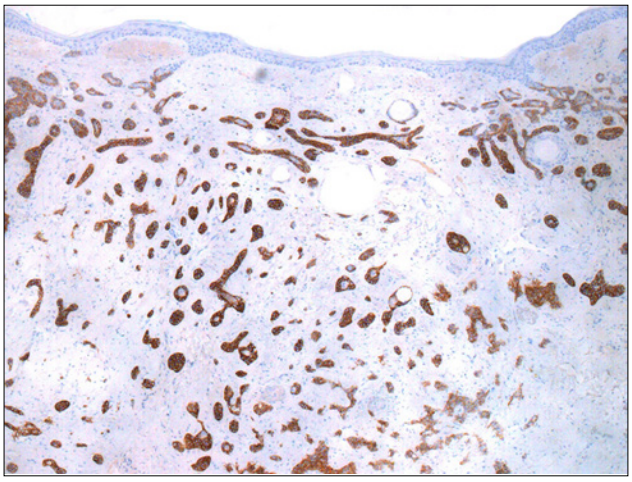

Figure 9. Ber-Ep4 stain is strongly positive. This finding is not specific as Ber-Ep4 is positive in basal cell carcinoma. One study showed Ber-Ep4 positivity in 38% of microcystic adnexal carcinomas, 57% of desmoplastic trichoepitheliomas, 100% of basal cell carcinomas, and 38% of squamous carcinomas [6]. (Copyright: ©2014 Inskip, Magee.)

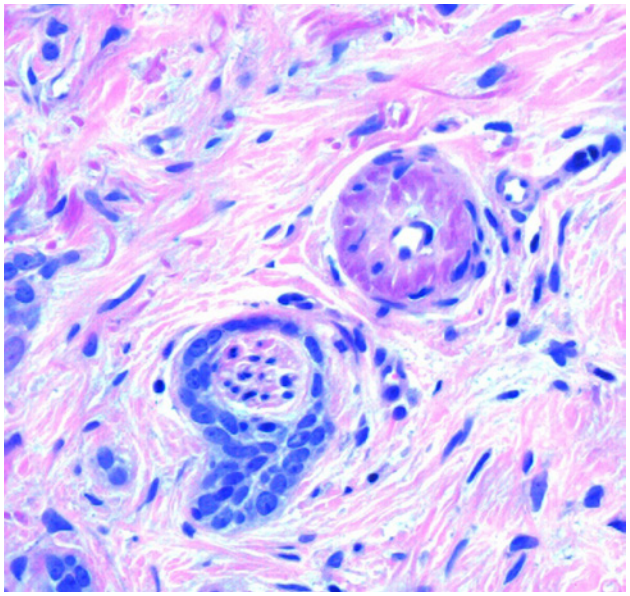

Figure 8. Focus of perineural invasion in deeper portion of tumor. (Copyright: ©2014 Inskip, Magee.)

Conclusion

A search of the literature has not discovered any previously published dermatoscopy images of a microcystic adenexal carcinoma. The two most notable dermatoscopic features of the lesion we present were the dense white structureless area centrally and the white clods of variable diameter peripherally. White clods of this pattern have also been observed in the more common adnexal skin tumor trichoepithelioma and may represent keratin retention cysts [7].

Microcystic adenexal carcinoma is currently considered a rare tumor. However, such rarities will present more often as the world population increases in age and has increased access to modern medicine. Dermatoscopy is a relatively new diagnostic tool. The authors feel it is important to publish such dermatoscopic images as ours to as wide an audience as possible to aid clinical diagnosis in future.

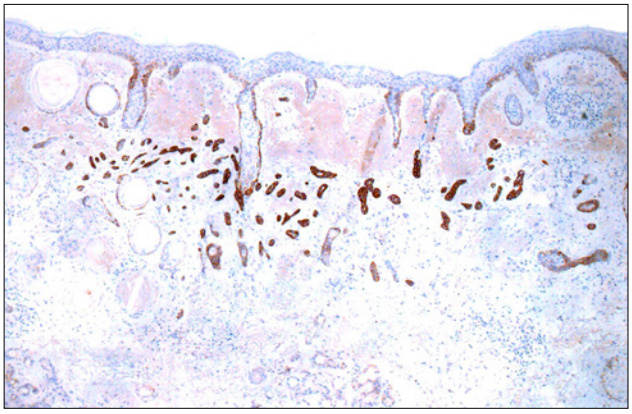

Figure 10. Cytokeratin 15 stain is also strongly positive. CK15 is a marker of the stem cells in the hair follicle bulge area. This finding favours microcystic adnexal carcinoma. Hoang et al found 92% of MAC and 100 % of desmoplastic trichoepitheliomata expressed CK15 whilst morphoeic BCC and squamous cell carcinomata were all negative. Thus this is a helpful marker to distinguish MAC from morphoeic BCC with adnexal differentiation [6]. (Copyright: ©2014 Inskip, Magee.)

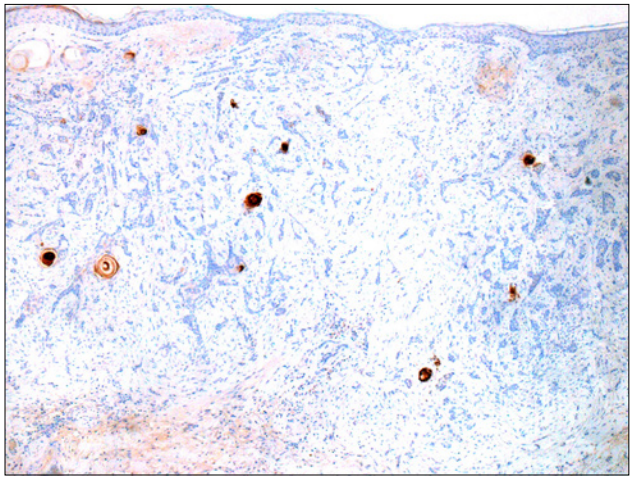

Figure 11. CEA stain marks some of the ductular lumina within the aggregates. This is a marker of eccrine and apocrine ducts, which would favor microcystic adnexal carcinoma. (Copyright: ©2014 Inskip, Magee.)

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