Milia-like papules and cutaneous atrophy on the infraorbital area

A 59-year-old Chinese female presented with 2 years' duration of asymptomatic milia-like papules over her right infraorbital area. One year later, apparent skin atrophy appeared at the periphery of these papules.

Cutaneous examination revealed multiple pinhead-sized whitish, transparent, very firm and hardly mobile papules with surrounding telangiectatic surface and distinct depression [Figure 1]. Submandibular and cervical lymph nodes were not palpable during physical examination.

Abdominal ultrasonography and computed tomography of the head, neck and thorax demonstrated no evidence of systemic or local metastasis. A skin punch biopsy (4 mm) showed a poorly circumscribed, deeply infiltrative, asymmetric tumor composed of basaloid nests, keratin-filled cysts, and ductal structures set in desmoplastic stroma [Figures 2-4]. Immunohistochemistry showed negativity for CK5/6, CK7, CK20, CD34, and Bcl-2.

The patient was treated surgically with excision of the tumor with a 1-cm margin and is on follow-up for the past 27 months, with no recurrence.

Question

What is your diagnosis?

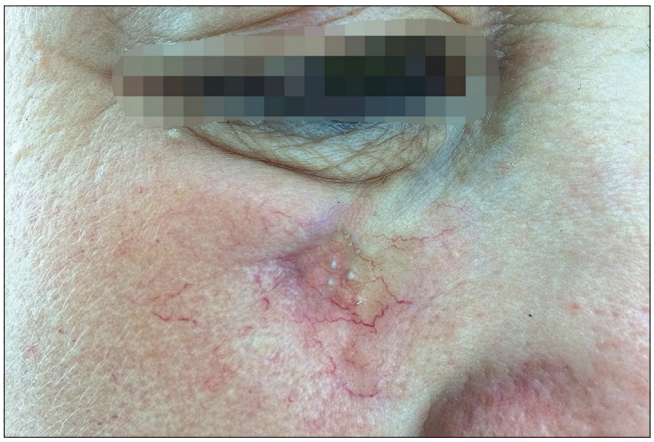

Figure 1: Multiple milia-like papules and their surrounding region with a telangiectatic surface and distinct depression on the right infraorbital area

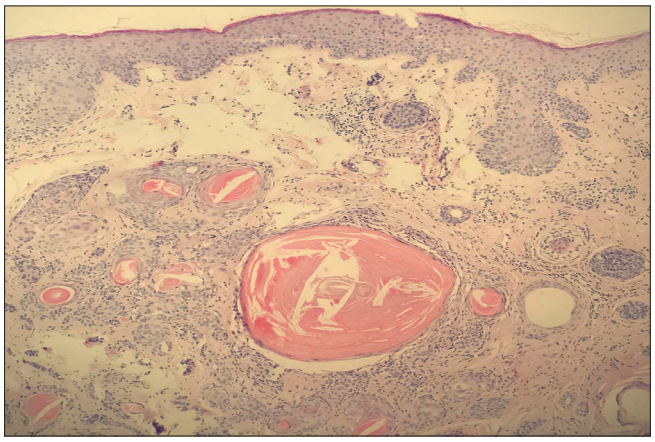

Figure 2: Small nests and keratin-filled cysts in the dermis (H and E, ×100)

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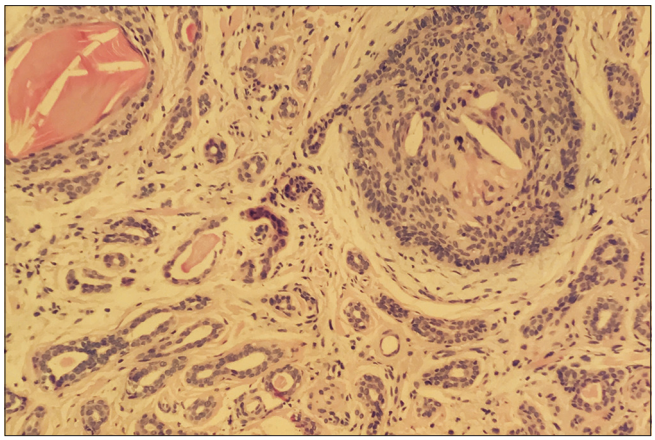

Figure 3: Variable numbers of tubular structures in the deep dermis (H and E, ×200)

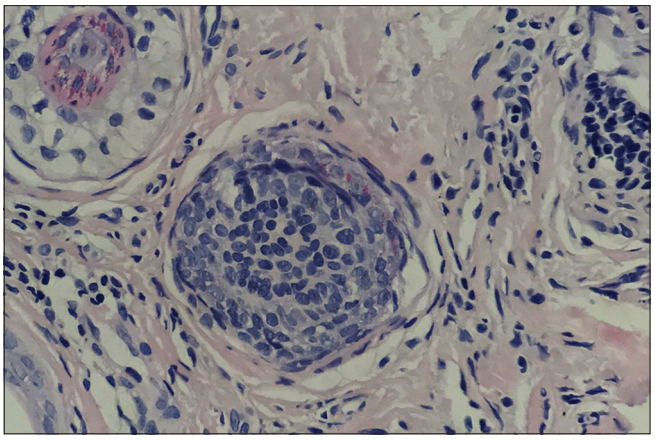

Figure 4: Small nests of basaloid cells presented in a desmoplastic stroma in thicker sections and scarce cellular atypia and mitoses (H and E, ×400)

Diagnosis

Microcystic adnexal carcinoma.

Discussion

Microcystic adnexal carcinoma is a locally aggressive tumor of the eccrine sweat glands. As a group, sweat gland neoplasms accounts for 0.005% of all malignant epithelial neoplasms.[1] The tumor is extremely rare in Asian individuals. A PubMed search of articles indexed for MEDLINE using the terms microcystic adnexal carcinoma revealed seven cases published in the last 3 years [Table 1].[2] However, cutaneous manifestations of milia-like papules and apparent atrophy in the disease have not been reported so far in the literature. Ours is such a case of microcystic adnexal carcinoma occurring in a Chinese woman. Ideally, deeper infiltration of the dermis/fat with or without perineural extension should be present to make a definitive diagnosis. Though a deep biopsy was not done in our case, the histological appearance was typical and hence we did not repeat the biopsy again.

Microcystic adnexal carcinoma commonly occurs in older patients in the fifth to seventh decades of life. Usually, there is no systemic metastasis. It demonstrates predilection for the head, particularly the periorbital and nasolabial regions. The disease typically presents as a smooth-surfaced, slow-growing, rarely ulcerated, firm, solitary nodule. The lesion commonly develops on the face for 3–5 years.[3] The tumor often extends far beyond its visible margins, probably leading to insufficient excision.[4] Our case is unusual in the following: a non-Caucasian patient, atypical lesion, and duration of less than 3 years.

Clinically, the current case shows many cutaneous similarities to morpheaform basal cell carcinoma. Their common features are the following: location on the face, smooth surface, slow growth, and depressed area of induration with telangiectasia. Other clinical differential diagnoses include desmoplastic trichoepithelioma and lichen sclerosus. Microcystic adnexal carcinoma is an ill-defined, fixed tumor rather than a small, well-circumscribed papulonodule seen in desmoplastic trichoepithelioma. Lichen sclerosus is less commonly seen on extragenital skin. Due to the high prevalence of microcystic adnexal carcinoma in Caucasians, a diagnosis of the disease may be significantly overlooked in non-Caucasian individuals. Thus, the present case stresses the importance of the differential diagnosis in non-Caucasian patients.

Histologically, microcystic adnexal carcinoma is a poorly circumscribed and deeply infiltrative tumor, composed of superficial keratinous cysts and solid aggregates of tumor cells in the mid-dermis. As the tumor cells invade the reticular dermis, they present as ductal structures similar to glands covered by one to two layers of flat or cuboidal cells. Histological features vary at different depths of the

Table 1: Main clinicopathological features of microcystic adnexal carcinoma

References	Sex/age, years	Location	Clinical presentation	Histopathological features	Immunohistochemical features
Haga *et al*.[6]	Female/78	Philtrum	Red tumor	Keratinous cysts and small islands of basaloid and squamous epithelium with ductal differentiation	BerEP4−
Hamsch and Hartschuh[4]	Male/52	Right external auditory canal	Palpable nodule	Keratinous cysts, irregular ductal structures, and solid epithelial nests	NA
Mukherjee *et al*.[7]	Male/69	Left eye	Loss of vision associated with pain	Severe nuclear atypia and tumor lobules	Cytokeratin+ CK7+ CD15+ BerEP4+ EMA+
Chen *et al*.[8]	Male/68	Left thumb	Subcutaneous nodule	Adenocarcinoma with a small component of sarcomatous differentiation	Pankeratin+ CK5/6+ CAM 5.2+ MOC31+ CEA+
Namiki *et al*.[9]	Female/79	Cheeks, forehead, and nose	Sclerotic erythematous plaque	Tubular structures similar to a tadpole appearance	EMA+
Waqas *et al*.[10]	Female/59	Forehead	NA	Infiltrating tubules and cords	NA
Waqas *et al*.[10]	Male/53	Left temporal scalp	NA	NA	NA
Our case	Female/59	Right infraorbital area	Milia-like papules and apparent skin atropy	Small nests of basaloid cells and keratin-filled cysts	CK5/6− CK7− CK20− CD34− Bcl-2−

NA: not available; +: positive, −: negative, CK: cytokeratin, CD: cluster of differentiation, EMA: epithelial membrane antigen, CAM: calmodulin, CEA: carcinoembryonic antigen, Bcl-2: B-cell lymphoma-2

tumor. Because it is the deeper component that contains the characteristic features of microcystic adnexal carcinoma, superficial biopsies can often result in misdiagnosis, which occurred in the cases previously reported.[1] The histopathological differential diagnoses include syringoma, desmoplastic trichoepithelioma, and squamoid eccrine ductal carcinoma. Syringoma is a purely ductal neoplasm without keratocyst formation and is confined to the superficial dermis. In contrast, desmoplastic trichoepithelioma lacks ductal structures. Squamoid eccrine ductal carcinoma is often characterized by conspicuous cytological atypia, pleomorphism and mitotic activity. CK5/6 is also positive in a recent report.[5]

Surgical excision of the tumor is the treatment of choice for locally invasive microcystic adnexal carcinoma. The surgical techniques most commonly used are Mohs surgery and conventional surgical excision with significantly wider margins. Recurrence rate can be as high as 60% after conventional surgical excision.[3] Therefore, sufficiently wide excision and careful follow-up are essential in patients such as the one reported here to prevent and detect early recurrence and/or metastasis of microcystic adnexal carcinoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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