

A long-standing plaque in the cutaneous upper lip of a late middle-aged man

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Our patient was a 68-year-old man who presented with a firm, immobile, asymptomatic, colorless nodule on the upper lip (Figure 1), which he described as being present for the past twenty years. The lesion had been growing recently without any other symptoms. The patient had no history of facial trauma, immunodeficiency, and radiotherapy. However, the patient had a positive history of prolonged sun exposure as a part of his occupational hazards. The cystic lesion was excised from his upper lip and was sent for histopathological examination, which reported: “numerous islands and some cords of basaloid cells with small keratinous cyst formation involving the superficial and deep dermis and subcutaneous fat” (Figure 2). There were foci of duct differentiation, a few with comma-like tails (Figure 2). Low mitotic figures & mild pleomorphic cellular shapes were seen. The stroma was dense and hyalinized, consisting of thickened collagen bundles.

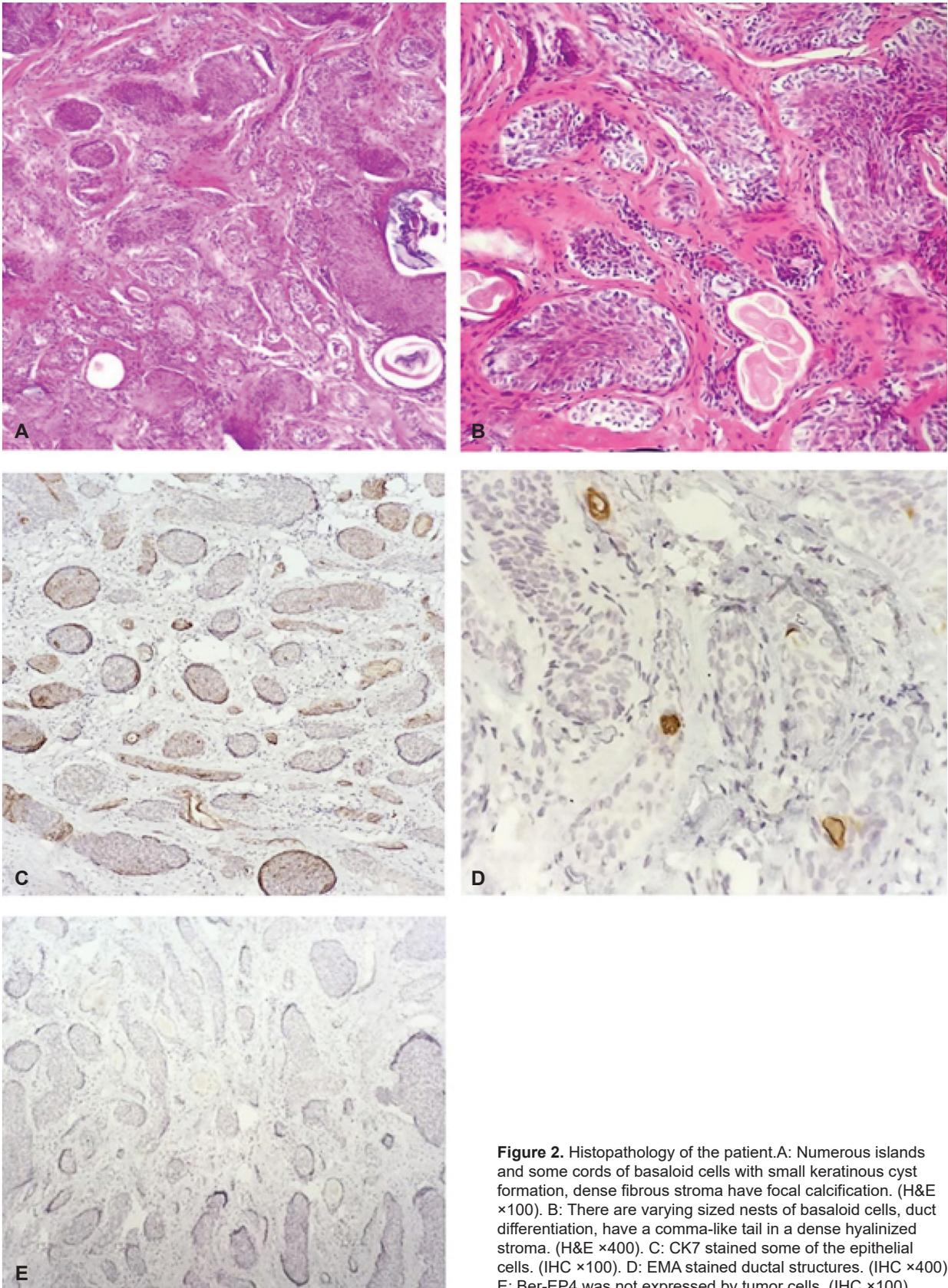
What is your diagnosis?

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Figure 1. A 68-year-old man with a cystic, immobile lesion on the left upper lip



ANSWER

Microcystic adnexal carcinoma

DISCUSSION

Microcystic adnexal carcinoma (MAC) is a rare and low-grade form of malignant skin sweat gland carcinoma, known to occur in young and middle-aged women. The head and neck region is particularly susceptible, and facial involvement is usually seen. The tumor typically demonstrates local invasion with low metastatic potential¹. However, there have been reports of MAC with lymphatic spread². This tumor was first described as an adnexal keratinocyte tumor in 1982 by Goldstein *et al.*³; the initial description of MAC as well-differentiated keratinocyte cysts along with well-defined ducts is still regarded as the histological characteristics of this condition⁴.

Due to its superficial cystic appearance on the skin, MAC may be misdiagnosed as other adnexal lesions such as syringoma, desmoplastic trichoepithelioma, and infiltrative basal cell carcinoma. Several studies reported eccrine carcinoma with squamous differentiation and squamous cell carcinoma as the differential diagnoses for a similar-appearing skin lesion⁵.

Treatment of MAC involves excision of the lesion, with Mohs micrographic surgery being the treatment of choice. In addition to excision, long-term follow-up is needed as there have been case reports of recurrence of MAC following excision of the lesion. The longest period recorded for recurrence was reported in a case report of MAC re-occurring thirty years following excision. Therefore, a whole-body skin examination with the examination of lymph nodes is recommended to identify recurrence and lymphatic spread of MAC. In our case, the lymph nodes of the patient were examined and did not demonstrate lymphadenopathy, and the patient was scheduled for a follow-up appointment after six months.

In summary, our case involved a MAC lesion occurring on the left upper lip of a late middle-aged man. Therefore, we suggest that MAC should be considered as a differential diagnosis for patients presenting with similar lesions. In a case series, Chiller K *et al.* reported that this neoplasm has a

tendency to involve the left side of the face, and our case also demonstrated this phenomenon. There is also the possibility that excessive exposure to the sun may be a contributing factor in the causation of MAC⁵. Thus, we suggest that a deep histological biopsy should be carried out in any suspected facial lesion to rule out the possibility of MAC, especially in middle-aged women presenting with suspected syringomas and trichoblastomas⁵.

LEARNING POINTS

1. Microcystic adnexal carcinoma (MAC) is a rare and low-grade form of malignant skin sweat gland carcinoma.
2. The head and neck region is particularly susceptible and facial involvement is usually the rule.
3. The tumor usually demonstrates local invasion with low metastatic potential.
4. The histopathological appearance of MAC includes differentiated keratinocyte cysts along with well-defined ducts.
5. Deep histological biopsy with immunohistochemistry (IHC) should be carried out in any suspected facial lesion to rule out the possibility of MAC, especially in middle-aged women presenting with suspected syringomas and trichoblastomas.

Conflict of interest: None declared.

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