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CASE AND RESEARCH LETTER

Epidemiological and Clinical Features of 49 Hispanic Patients With Microcystic Adnexal Carcinoma



Características epidemiológicas y clínicas de 49 pacientes hispanos con carcinoma aneal microcístico

To the Editor,

Microcystic adnexal carcinoma (MAC) is a slow-growing, aggressive, cutaneous neoplasm with a tendency for perineural invasion and high recurrence rates.¹ There are less than 2000 reported cases worldwide, mostly in Caucasian individuals.² Although UV exposure, radiotherapy, and genetics may increase susceptibility, the exact pathogenesis remains uncertain. MAC presumably originates from pluripotent adnexal keratinocytes, as it shows elements of eccrine (keratinous cysts) and follicular (epithelial cords and ducts) differentiation.¹ It primarily affects the head and neck region; and may appear as an ill-defined, erythematous, skin-coloured, or yellowish plaque, nodule, or cyst. Due to the clinical and histopathological similarity with other skin or adnexal cancers, the disease may be particularly underappreciated in populations that have not been investigated. Therefore, our study aimed to describe the epidemiology and clinical presentation of MAC in Hispanic patients from a tertiary dermatology hospital in Mexico.

We performed a retrospective analysis from 2010 to 2020 including complete patient records with a postoperative, histopathological diagnosis of MAC. A total of 49 cases were found, corresponding to 0.05% of the 83,364 samples reviewed in the dermatopathology department during the study period. The mean age was 67 years (median, 67 years; range, 24–101 years). Females (69.4%) were more commonly affected than males (30.6%). The average time-to-onset was 3 years (median, 2 years; range, 0.08–22 years). The head was involved in 91.8% of the cases, and the trunk in 8.2%. In the head region, most lesions occurred on the cheeks (40%), followed by the nose (28.9%), lips (11.11%), forehead (6.7%), eyelids (6.7%), scalp (4.4%), and ears (2.2%). The nodular aspect (71.4%) predominated over plaque (26.5%) and cyst (2%). Additional features such as a pearly border,

ulceration, atrophy, and telangiectasias were described in 42.9%, 38.8%, 16.3% and 16.3% of the cases, respectively. The average lesion diameter was 1.6 cm (median, 1 cm; range, 0.3–10 cm).

Apart from the presence of a pearly border, our results are in line with the published literature. In a systematic review of 1968 patients, 92.8% of the subjects were Caucasian, and only 0.2% were Hispanics.² In total, mean age was 61.8 years, females represented 54.1% of the cases, and latency ranged from 1 to 11 years. Although the location and morphology were not quantitated, it includes other case series where head and neck involvement ranges from 60 to 100%.^{3–8} The mean diameter at diagnosis was 2.8 cm.²

Since MAC may be confused with other tumours, we also evaluated the clinical and histopathological diagnostic accuracy. Prior to the histopathology result, most were clinically diagnosed as basal cell carcinoma (BCC) (71.4%) and squamous cell carcinoma (SCC) (16.3%), followed by nevus (4.1%), lichen planus pilaris (2%), actinic keratosis (2%), trichodiscoma (2%), and adnexal tumour (2%). As expected, documentation of a pearly border was significantly associated with the presumptive clinical diagnosis (Fisher–Freeman–Halton $p=0.041$), from which BCC represented 90.5% of the cases (Table 1). The frequency of preoperative histopathological misdiagnosis was 20.4%, still below the 27–69% in other case series.^{3–5,9} In our sample, this included BCC (60%), basosquamous cancer (20%), and SCC (20%).

The limitations of this study are inherent to its retrospective nature. We were not able to re-evaluate the clinical morphology of the lesions, since descriptions were obtained directly from the medical records. However, these were elaborated by qualified dermatologists from our institution. In the future, we hope to create a picture database to improve our descriptions, and also study various clinical–histopathological correlations with a focus on prognosis.

In conclusion, this is the first study to characterise MAC in Hispanics, specifically of Mexican origin. Our data should complement previous epidemiological and clinical knowledge in a population that has been underrepresented in larger studies. Importantly, our results demonstrate that BCC was the main differential diagnosis of MAC, and that nearly half of our cases had a documented pearly border. Also, our analysis shows that MAC clinical suspicion remains

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Table 1 Pearly border and presumptive clinical diagnoses.

Pearly border	Presumptive clinical diagnoses							Total n (%)
	Basal cell carcinoma	Squamous cell carcinoma	Lichen planus pilaris	Nevus	Actinic keratoses	Trichodiscoma	Adnexal tumour	
<i>Absent</i> n (%)	16 (57.1)	7 (25)	1 (3.6)	2 (7.1)	1 (3.6)	1 (3.6)	0 (0)	28 (57.1%)
<i>Present</i> n (%)	19 (90.5)	1 (4.8)	0 (0)	0 (0)	0 (0)	0 (0)	1 (4.8)	21 (42.9%)

Caption: A pearly border was documented in 42.86% of all MAC cases. Prior to the postoperative histopathology result, basal cell carcinoma was the most common presumptive clinical diagnosis. As expected, basal cell carcinoma was also the most frequent presumptive clinical diagnosis that had a documented pearly border (90.48%).

low, with only 1 case (2%) being considered as an adnexal tumour. Moreover, the frequency of histopathological misdiagnosis suggests that initial sampling may be insufficient to establish the definitive diagnosis in more than a fifth of the cases. Therefore, increasing disease awareness is crucial to select an adequate biopsy technique that can improve the diagnostic accuracy of histopathology. Together, this may result in earlier, well-planned treatment strategies and better patient outcomes.

Conflict of interest

The authors declare that they have no conflict of interest.

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