

subsequent appearance of similar lesions and rapid response to immunosuppressive therapy.

Two cases of pyoderma gangrenosum mimicking squamous cell carcinoma have been reported in the literature. In 2012, Wolfe et al.⁴ described the case of a patient with an ulcerated lesion on the left hand that was histologically compatible with a moderately differentiated squamous cell carcinoma but that progressed rapidly following excision. Evaluation by the dermatology team and a second biopsy showed a neutrophilic infiltrate in the reticular dermis; the patient was diagnosed with pyoderma gangrenosum and improved rapidly after initiation of treatment with oral prednisone. In 2015, Čuk Radović et al.⁵ published the case of a patient with large, ulcerated lesions on the trunk who had been initially treated for chronic infection and subsequently for squamous cell carcinoma on observation of compatible histologic findings. The lesions were treated with several wide local excisions, but the disease recurred rapidly at the edges and there was additional progression of ulcers.

We have presented a new case of pyoderma gangrenosum with histopathologic findings consistent with squamous cell carcinoma. This case highlights the importance of integrating clinical and histologic findings to aid the establishment of a definitive diagnosis, avoiding diagnostic delays and unnecessary treatments, including invasive surgery, which can lead to clinical exacerbation due to pathergy reactions.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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M. González-Sabín,^{a,*} E. Rodríguez-Díaz,^a
P. Gonzalvo-Rodríguez,^a I. Astola-Hidalgo^b

^a Servicio de Anatomía Patológica, Hospital Universitario de Cabueñes, Gijón, Asturias, Spain

^b Servicio de Medicina Intensiva, Hospital Universitario Central de Asturias, Oviedo, Asturias, Spain

* Corresponding author.

E-mail address: martagonzalezsabin@gmail.com
(M. González-Sabín).

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Clear Cell Papulosis: A Case Series[☆]



Papulosis de células claras: una serie de casos

To the Editor:

Clear cell papulosis (CCP) is a rare entity of unknown cause that was first described in 1987.¹ It is reported more frequently in Asian countries and usually affects children under 2 years of age, with comparable prevalence in both sexes.²

It is characterized by flat, hypopigmented, asymptomatic macules and/or papules of 2 mm to 10 mm located on the lower abdomen and pubic area, and less frequently on the axillae, mammary line, groin, and extremities.³ Histology reveals the characteristic presence of round clear cells with abundant, pale cytoplasm located between the basal and suprabasal keratinocytes. On immunohistochemistry these cells are positive for antibodies against cytokeratin AE1/AE3, cytokeratin 7 (CK7), cytokeratin CAM5.2, carcinoembryonic antigen, epithelial membrane antigen, and gross cystic disease fluid protein.³ Both the origin and the role of these clear cells remain unclear.⁴ It is suggested that they are derived from secretory cells of the eccrine glands.⁵ It has also been proposed that clear cells are derived from Toker cells and may serve as precursor cells of Paget disease, although progression from CCP to Paget disease has not been demonstrated to date.²

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Table 1 Epidemiological, Clinical, and Histopathological Characteristics and Duration of Follow-up

Patient	Age	Sex	Age at Onset	Lesion type	Lesion Size	Lesion Number	Location	Histopathology	Immunohistochemistry	Treatment	Follow-up
1	11 y	F	12 mo	Asymptomatic hypopigmented macules	2–3 mm	>10	Axilla, mammary line, lower abdomen	Psoriasiform hyperplasia, few cells located in the basal layer containing abundant cytoplasm and a central nucleus	CK7 ⁺	Observation	Unchanged after 10 years
2	1 y	F	10 mo	Asymptomatic hypopigmented macules	2–3 mm	>10	Lower abdomen, pubic area	Few cells located in the basal layer containing abundant cytoplasm and a central nucleus	CK7 ⁺	Observation	Decrease in lesion size/number after 12 months
3	1 y	F	Birth	Slightly raised, asymptomatic macules and papules	2 mm	>10	Axilla, lower abdomen	Few, scattered cells located in the basal and suprabasal layers and containing abundant pale cytoplasm	CK7 ⁺	Observation	Slight increase in lesion size/number after 18 months
4	1 y, 11 mo	F	7 mo	Asymptomatic hypopigmented macules	2–3 mm	4	Lower abdomen, pubic area	Round cells located between basal and suprabasal keratinocytes and containing abundant cytoplasm	CK7 ⁺	Observation	None
5	7 y, 10 mo	M	4 mo	Asymptomatic hypopigmented macules and papules	2–3 mm	>10	Axilla, lower abdomen, buttocks, lumbar region	Acanthosis, few cells located in the basal and suprabasal layers containing abundant pale cytoplasm	CK7 ⁺	Observation	No changes after 12 months
6	2 y, 9 mo	F	20 mo	Asymptomatic hypopigmented papules	2 mm	>10	Lower abdomen, pubic area	Clear cells located in the basal and suprabasal layers	CK7 ⁺ EMA ⁺	Observation	No changes after 9 months
7	9 y, 10 mo	F	9 mo	Asymptomatic hypopigmented macules	2–3 mm	>10	Lower abdomen, pubic area	Few large cells located between basal keratinocytes and containing clear cytoplasm and a round nucleus	CK7 ⁺ , EMA ⁺ , CEA ⁺	Observation	No changes after 12 months
8	6 y, 10 mo	F	12 mo	Asymptomatic hypopigmented macules and papules	2 mm	>10	Legs	Clear cells forming a gland-like structure in the basal layer	Mucin ⁺ , PAS ⁺	Observation	Decrease in lesion size/number after 18 months

Abbreviations: CEA, carcinoembryonic antigen; CK7, cytokeratin 7; EMA, epithelial membrane antigen; F, female; M, male; PAS, periodic acid-Schiff.



Figure 1 Male aged 19 months with macules and papules (2–3 mm) on the lower abdomen and pubic area.

CCP is asymptomatic, and does not require treatment. In most cases lesions disappear with time.³

Case Descriptions

A search for patients diagnosed with CCP in 3 pediatric hospitals in Peru and Argentina between 2010 and 2016 identified 8 cases: 7 females and 1 male. Lesions had appeared before 2 years of age in all cases, and were present from birth in one case (Table 1). All patients had hypopigmented lesions (2–3 mm in diameter). The majority of lesions were macules, followed by papules, and most were located on the lower abdomen and pubic area (Fig. 1). Other affected areas included the axillae, mammary line, buttocks, lumbar region, and legs.

The initial clinical diagnosis was CCP in 6 patients and postinflammatory hypopigmentation in 2 patients. In all cases, histology revealed few scattered large cells with abundant clear cytoplasm located in the basal and suprabasal layers. Seven patients were positive for anti-CK7 immunostaining (Fig. 2). In 3 of these patients CK7⁺ cells were located next to the opening of the acrosyringium, while in Patient 8 the CK7⁺ cells formed a gland-like structure and were positive for periodic acid-Schiff (PAS) and Alcian blue stains.

None of the patients received treatment. Seven patients underwent clinical follow-up for 9 to 18 months. Information on the clinical evolution of the remaining patient (Patient 1) was provided by a family member. Lesions remained stable in 4 patients (after 9, 12, and 12 months, and 10 years of follow-up, respectively), improved in 2 patients (after 2 and 18 months), and worsened in 1 patient (after 18 months).

There are few reported cases of CCP,² and only 5 out of 39 reported cases involve non-Asian patients.^{6–8} The majority of patients in our series were female and most developed lesions within the first years of life, except for one patient who was diagnosed at 11 years of age. This profile is in line with that described in previous case reports, in which all patients have been children,² except for 1 adult case.⁹

All patients in our series were born to Latin American parents and presented multiple small hypochromic macules and/or papules in the lower abdomen and pubic area. Other affected areas included the axillae, mammary line, buttocks, and lumbar region. In one patient, lesions were

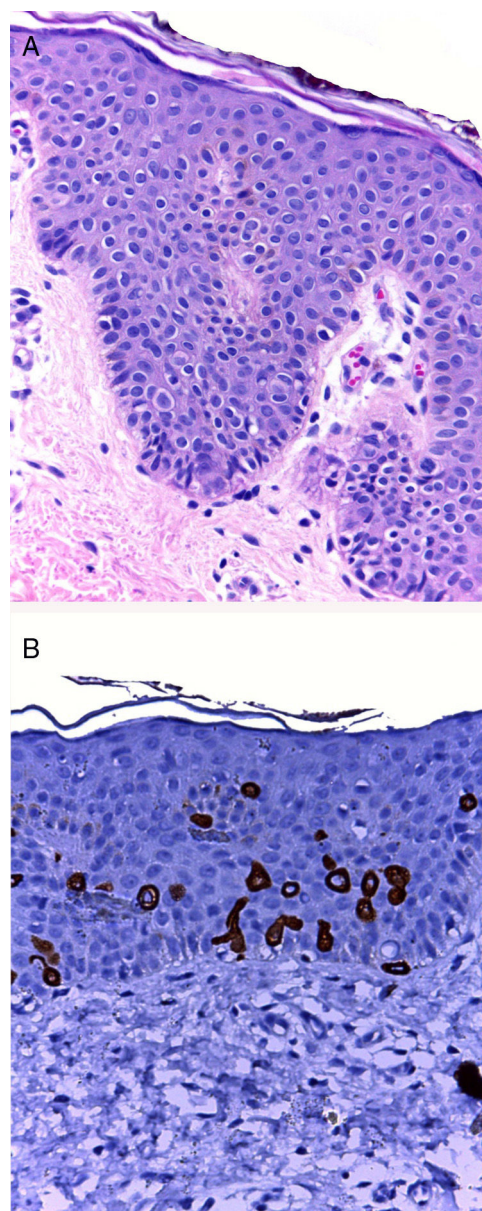


Figure 2 A, Few scattered round cells with abundant clear cytoplasm in the basal layer of the epidermis (hematoxylin-eosin, original magnification $\times 10$). No changes are evident in the dermis. B, Anti-cytokeratin 7 immunostaining reveals the presence of clear cells, located predominantly in the basal layer and also in the suprabasal layer.

located exclusively on the legs, which is not a commonly affected region.

Histologically, clear cells can be confused with melanocytes, Tokel cells, or Langerhans cells.¹⁰ Diagnosis is primarily based on histology, although immunohistochemistry can be useful in doubtful cases. It is worth noting that in some cases the CK7⁺ cells were located close to the opening of the acrosyringium. Unusually, in Patient 8 these CK7⁺ cells formed a gland-like structure and were positive for Alcian blue and PAS, indicating the presence of mucin. These findings support the view that these cells have a glandular origin, as proposed by Kim and coworkers.⁵

The lesions usually increase in size and number during the first months and years of life, but gradually diminish without leaving sequelae within a mean period of 11 years after appearance.³ Although the follow-up of 7 patients was limited to 9 to 18 months, 2 patients (Patients 2 and 8) showed a decrease in lesion size and number during this time.

Familiarity with this entity is important given that hypopigmented lesions are common in pediatric patients. The main differential diagnosis is postinflammatory hypopigmentation. Other clinical diagnoses to consider include pityriasis versicolor, flat warts, and vitiligo.

This is the first reported series of CCP cases in Latin America. Identification of this rare entity in children is important to avoid erroneous diagnoses and unnecessary treatments.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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C. Apagüño,^{a,*} R. Pomar,^a J. Peceros,^a F. Velásquez,^a R. Ballona,^a R. Castro,^b R. Lipa,^c C. Chian,^d R. Cano,^e M. Larralde^e

^a Servicio de Dermatología Pediátrica, Instituto Nacional de Salud del Niño, Breña, Lima, Perú

^b Servicio de Dermatología Pediátrica, Instituto Nacional de Salud del Niño, San Borja, Lima, Perú

^c Servicio de Anatomía Patológica, Instituto Nacional de Salud del Niño, San Borja, Lima, Perú

^d Servicio de Anatomía Patológica, Hospital Nacional Arzobispo Loayza, Lima, Perú

^e Servicio de Dermatología Pediátrica, Hospital Ramos Mejía, Buenos Aires, Argentina

*Corresponding author.

E-mail address: consuelodejesus111@hotmail.com

(C. Apagüño).

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Ultrasound Characteristics of Lipoma of the Tongue[☆]



Características ecográficas de los lipomas linguales

To the Editor:

A 60-year-old man with a history of non-insulin-dependent diabetes mellitus and dyslipidemia consulted us for an asymptomatic lesion on his tongue. He reported that it had been present for approximately 2 years, growing at first but then becoming stable over the past few months.

Physical examination showed a rounded well-defined nodule on the right side of the anterior third portion of the tongue. The yellowish nodule measured approximately 6 mm at the longest axis, was reddish on the surface, flexible

to the touch, and not adherent to deep layers. Dermatologic ultrasound with an 18MHz probe showed an oval tumor that was slightly more hypoechoic than the adjacent tissues. The well-defined but not encapsulated tumor measured 7.5 × 2.5 mm on ultrasound and had no Doppler flow either on the surface or at the center. **Fig. 1** shows the full length of the nodule. Suspecting a benign, avascular tumor, we performed a 4-mm punch biopsy. The histopathologic

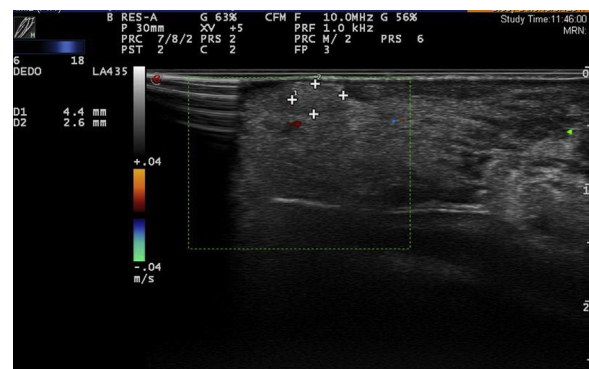


Figure 1 Ultrasound image of the full length of the lesion.

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