

Cutaneous Epithelioid Angiomatous Nodule: A Recently Described Vascular Tumor

Nódulo angiomatoso epitelióide cutáneo. Un nuevo tumor vascular recientemente descrito

To the Editor:

Cutaneous epithelioid angiomatous nodule (CAEN) is a rare benign vascular proliferation recently described by Brenn and Fletcher.¹ It typically develops in adults as a solitary, small, reddish, fast-growing nodular lesion most frequently located on the trunk or limbs.² Histologically it consists of an infiltration of epithelioid cells, with abundant eosinophilic cytoplasm and vesicular nuclei with conspicuous nucleoli. Although the proliferation is predominantly solid, it is normal to encounter vascular



Figure 1 Reddish nodule about 5 mm in diameter and with an erythematous halo, located on the back.

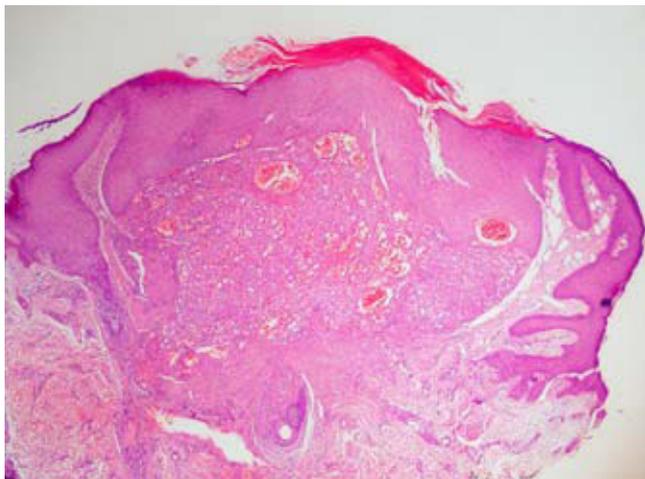


Figure 2 Well-defined unilobular proliferation of epithelioid cells located in the superficial dermis. Observe the accompanying epidermal hyperplasia.

lumens in scattered foci through the lesion. A peripheral lymphoplasmocytic infiltrate is observed in many cases.^{2,3} The differential diagnosis should include other tumors and vascular proliferations composed of epithelioid cells.² We describe the case of a 47-year-old woman with no personal history of interest, who consulted with a nodular lesion measuring 5 mm that had developed on her back over the previous 7 months. Examination revealed a small, reddish nodule surrounded by an erythematous halo (Figure 1). The lesion was surgically excised and histology was compatible with a diagnosis of CAEN (Figures 2 and 3).

Immunohistochemistry was positive for CD31 and CD34, and the lymph vessels of the tumor periphery stained intensely with D2-40.^{4,6} CAEN was first described in 2004 by Brenn and Fletcher following a study of 15 cases.¹ A further 17 cases have since been reported. Clinically CAEN presents as a rapidly growing (over weeks or months), usually solitary erythematous or violaceous nodule or papule, with a diameter ranging between 0.3 cm and 1.5 cm.⁴ The lesion usually develops in adults, with a wide age range of presentation, and is typically located on the trunk or extremities.³ Histology shows a solid, well-delimited proliferation, generally affecting the superficial dermis, and composed of epithelioid endothelial cells. Well-developed vessels are found in scattered foci through the lesion, and a lymphoplasmocytic infiltrate is present in the tumor periphery. A consistent finding is reactive hyperplasia of the epidermis, frequently delimiting the lesion peripherally.^{2,3} Some authors consider CAEN to be a specific entity, whereas other authors consider it to be a variant of epithelioid hemangioma, also called angiolymphoid hyperplasia with eosinophilia (ALHE).⁷ Nonetheless, as will be seen below, the clinical and histologic characteristics are different. Clinically, CAEN can easily be confused with a pyogenic granuloma; however, the latter has vessels of varying sizes grouped in a lobular pattern, and no epithelioid cells.

The differential diagnosis for CAEN should include all vascular proliferations composed of epithelioid cells, namely, ALHE, epithelioid angiosarcoma, epithelioid hemangioendothelioma,

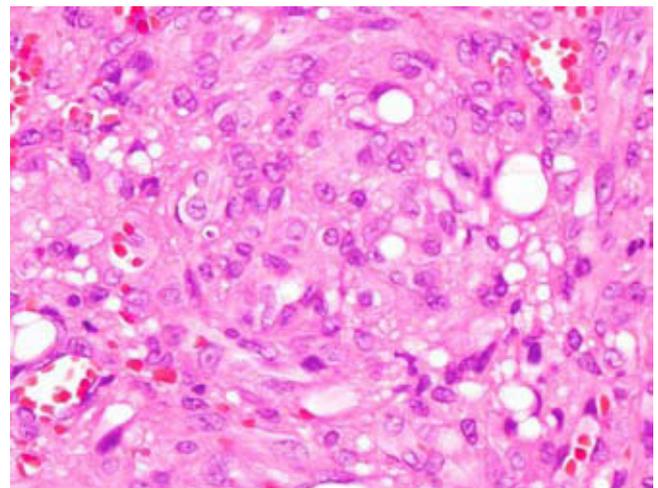


Figure 3 Detail of solid masses of epithelioid cells. Small vascular lumens.

and bacillary angiomatosis.^{2,8} The differentiation between CAEN and ALHE is based on both clinical and histopathologic criteria. Clinically ALHE occurs on the head and neck, and often presents as a cluster of lesions, whereas CAEN occurs predominantly on the trunk, and typically occurs as a single lesion. Histologically ALHE affects the deep dermis and hypodermis, is multilobular, and is predominantly vasoformative, whereas CAEN tends to be situated in the superficial dermis, is unilobular, and is usually solid. Furthermore, ALHE has a greater inflammatory component, the eosinophil count is higher, and there is a more abundant stroma.² Epithelioid angiosarcoma should also be included in the differential diagnosis. Despite its more aggressive biological behavior, it shares certain morphological characteristics with CAEN that could lead to diagnostic error. Epithelioid angiosarcoma, however, is not so well circumscribed, and often presents with a large necrotic mass or smaller patches of necrosis. Moreover, nuclear atypia is conspicuous, with nuclear pleomorphism and atypical mitoses.⁹ Immunohistochemistry is useful because up to 50% of epithelioid angiosarcomas are cytokeratin-positive, whereas CAEN is cytokeratin-negative.² Epithelioid hemangioendothelioma is a low-grade angiosarcoma that may be included within the spectrum of vascular lesions with epithelioid characteristics. Compared to CAEN lesions, epithelioid hemangioendothelioma lesions are larger, less well defined, and deeper. Histologically, the vessels in epithelioid hemangioendothelioma are not well defined, and the growth has a fibromyxoid stroma⁹ similar to metastases from a carcinoma. Up to 25% of cases are cytokeratin-positive.² Bacillary angiomatosis typically affects immunosuppressed patients—for example, those with the human immunodeficiency virus²—and presents as multiple eruptive vascular lesions. Histologically, it has a lobular pattern with a conspicuous proliferation of capillaries separated by connective tissue septa. The endothelial cells lining the vessels are large, with an epithelioid appearance, and they protrude into the vascular lumens. Bacillary angiomatosis is further characterized by a neutrophil infiltration, and aggregates of granular material corresponding to bacterial colonies (*Bartonella henselae*).³ Although the pathogenesis of CAEN is unknown, its rapid development, the presence of epidermal hyperplasia, and the inflammatory response would suggest it is a reactive disorder.⁴

References

1. Brenn T, Fletcher CD. Cutaneous epithelioid angiomatous nodule: a distinct lesion in the morphologic spectrum of epithelioid vascular tumors. *Am J Dermatopathol*. 2004;26:14-21.
2. Requena Caballero C, Nicolau MJ, Haro R, Martorell A, Sanmartín O, Llombart B. Proliferative cutaneous epithelioid angiomatous nodule. *Actas Dermosifiliogr*. 2009;100:137-41.
3. Solís-García E, Segura-Sánchez JM, Rodríguez-Enríquez B, Sánchez-Sánchez-Vizcaíno F, González-Serrano MT. Nódulo angiomatoso epitelióide cutáneo. ¿Variante superficial y extrafacial de hemangioma epitelióide? *Rev Esp Patol*. 2006;39:59-62.
4. Álvarez-Argüelles-Cabrera H, Guimera-Martín-Neda F, Carrasco JL, García-Castro MC, Hernández-León CN, Díaz-Flores L. Cutaneous epithelioid angiomatous nodule. *J Eur Acad Dermatol Venereol*. 2008;22:1383-5.
5. Zamecnik M. Relationship between cutaneous epithelioid angiomatous nodule and epithelioid hemangioma. *Am J Dermatopathol*. 2004;26:351-2.
6. Fernández-Flores A. D2-40 and cutaneous epithelioid angiomatous nodule. *Am J Dermatopathol*. 2008;30:302-4.
7. Al-Daraji WI, Prescott RJ, Abdellaoui A, Khan MM, Kulkarni K, Youssef MM. Cutaneous epithelioid angiomatous nodule: different views or interpretations in the analysis of ten new cases. *Dermatol Online J*. 2009;15:2.
8. Fernández-Flores A, Montero MG, Renedo G. Cutaneous epithelioid angiomatous nodule of the external ear. *Am J Dermatopathol*. 2005;27:175-6.
9. Sangüeza OP, Walsh SN, Sheehan DJ, Orland AF, Llombart B, Requena L. Cutaneous epithelioid angiomatous nodule: a case series and proposed classification. *Am J Dermatopathol*. 2008;30:16-20.

J.M. Segura-Palacios,^{a,*} E. Gómez-Moyano,^a
A. Hiraldo-Gamero,^a A. Sanz-Trelles^b

^a*Servicio de Dermatología y Anatomía Patológica, Complejo Hospitalario Carlos Haya, Málaga, Spain*

^b*Servicio de Anatomía Patológica, Complejo Hospitalario Carlos Haya, Málaga, Spain*

*Corresponding author.

E-mail address: juanmanuelo_1983@hotmail.com
(J.M. Segura-Palacios).

Dermoscopy in Furuncular Myiasis

Dermatoscopia en la miasis forunculoide

To the Editor:

We report the case of a 40-year-old Peruvian man who came to our clinic because of the appearance, 2 to 3 weeks earlier, of papular lesions in the left pectoral

region. The lesions had persisted despite drainage and subsequent treatment with erythromycin, ciprofloxacin, and cloxacillin for 10 days. They had appeared a few days before the patient returned from a 21-day stay in Minas Gerais, Brazil. He stated that there were initially 6 to 7 lesions that he described as insect bites, and that, during the week prior to consultation, there had been soreness and stinging in the 2 persistent lesions. Physical examination revealed 2 indurated papules that were 0.5 and 2 cm in diameter in the left pectoral region;