

In conclusion, BCPHTPCN is a recently described tumor that shows a preference for the perioral region, and whose pathology findings are a hybrid between perineurioma and cellular neurothekeoma. Immunohistochemistry is variable, but important findings include positivity for S100A6, MiTF, NKI/C3, PGP9.5, EMA, and NSE. In our case, we draw particular attention to the presence of numerous CD68⁺ and CD163⁺ cells between the tumor nests. This peculiar trait has been reported previously in 2 patients, but as a less intense feature.¹ Given the variability of the immunohistochemistry findings and, in general, the low levels of expression of typical neural markers in this neoplasm, we consider the clinical and morphological features to be key elements in reaching the diagnosis.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

Acknowledgments

The authors would like to thank the Dermatology Department and the technicians of the Pathology Department of Complejo Hospitalario de Navarra.

References

1. Requena L, Sitthinamsuwan P, Fried I, Kaddu S, Schirren C, Schärer L, et al. A benign cutaneous plexiform hybrid tumor of perineurioma and cellular neurothekeoma. *Am J Surg Pathol.* 2013;37:845–52.
2. Yamada S, Kitada S, Nabeshima A, Noguchi H, Sasaguri Y, Hisaoka M. Benign cutaneous plexiform hybrid tumor of perineurioma and cellular neurothekeoma arising from the nose. *Diagn Pathol.* 2013;8:165.
3. Linos K, Stuart L, Goncharuk V, Edgar M. Benign cutaneous biphasic hybrid tumor of perineurioma and cellular neurothekeoma: A case report expanding the clinical and histopathologic features of a recently described entity. *Am J Dermatopathol.* 2015;37:319–22.
4. Hornick J, Fletcher C. Cellular neurothekeoma: Detailed characterization in a series of 133 cases. *Am J Surg Pathol.* 2007;31:329–40.
5. Fetsch J, Laskin W, Hallman J, Lupton G, Miettinen M. Neurothekeoma: An analysis of 178 tumors with detailed immunohistochemical data and long-term patient follow up information. *Am J Surg Pathol.* 2007;31:1103–14.

C. Areán,^{a,*} A. Córdoba,^a L. Requena,^b M^a.L. Álvarez^a

^a Servicio de Anatomía Patológica, Complejo Hospitalario de Navarra, Pamplona, Navarra, Spain

^b Departamento de Dermatología, Fundación Jiménez Díaz, Universidad Autónoma, Madrid, Spain

* Corresponding author.

E-mail address: carolina.arean.cuns@navarra.es (C. Areán).

Majocchi Granuloma of the Breast: A Rare Clinical Entity



Granuloma de Majocchi: Una rara entidad

Dear Editor,

Deep penetration of the skin by dermatophytic agents may provoke a granulomatous inflammatory skin reaction. This was first described by Majocchi.¹ Majocchi granuloma is characterized clinically by papular, pustular, or nodular inflammatory lesions occurring typically on the limbs or face,² though isolated cases affecting the vulva or scrotum have been published.³ We report a unique presentation of Majocchi granuloma in the breast; there are no previous reports of involvement of this location.

A 28-year-old female presented with a slightly painful, raised reddish lesion that had developed in her left breast over the previous 3 months. She gave no history of trauma and did not describe constitutional symptoms. The lesion had increased in size after the repeated application of mud to the area over 2 months. Treatment with a potent topical corticosteroid (betamethasone valerate, 0.122% wt/wt) was then applied twice daily for a month.

Clinical examination revealed a single erythematous plaque with areas of scarring. A yellowish discharge emanated from multiple openings in an area beneath the left nipple measuring approximately 5 × 4 cm (Fig. 1). On

palpation the plaque was firm and indurated and was slightly tender; there was no underlying breast lump. Diascopy findings were nonspecific. Gram stain of the discharge showed the presence of gram-positive cocci and abundant neutrophils. No acid-fast bacilli (AFB) were seen on direct smear. Potassium hydroxide (KOH) mount and fungal culture of skin scrapings and of the discharge was negative. Ultrasonography of the breasts was normal, though a single lymph



Figure 1 Papular-pustular plaque on the left breast with central scarring and a purulent discharge visible at the periphery.

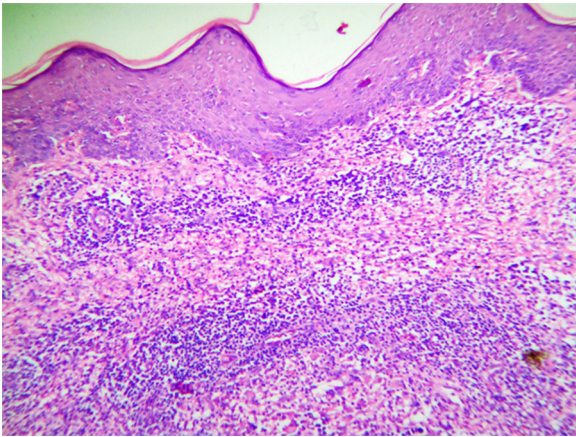


Figure 2 Acanthosis of the epidermis with a mixed cell inflammatory infiltrate in the superficial and mid dermis. Hematoxylin and eosin, original magnification $\times 100$.

node of 20×8 mm with a preserved hilum was observed in the left axilla. Chest X-ray was normal and the Mantoux skin test produced a wheal of 8×6 mm. Routine blood tests were within normal limits. The patient was treated with oral cefuroxime 500 mg twice a day plus topical 2% mupirocin cream but showed no improvement after 2 weeks of treatment. We performed skin biopsy based on a differential diagnosis of hidradenitis suppurativa, lupus vulgaris, or subcutaneous fungal infection. Hematoxylin and eosin staining of the sample revealed a mixed cell, granulomatous inflammatory reaction in the dermis (Fig. 2). Ziehl-Neelsen and periodic acid Schiff (PAS) stains to detect AFB and fungi were both negative. Mycobacterial culture of the tissue sample was negative, but fungal culture on Sabouraud dextrose agar with chloramphenicol produced growth of heaped up, folded violaceous colonies of waxy consistency after 3 weeks, suggestive of *Trichophyton violaceum*. Lactophenol cotton blue mount showed the presence of tangled, irregular branched hyphae with chlamydospores. With a final diagnosis of Majocchi's granuloma, the patient was treated with 250 mg of oral terbinafine once daily plus topical 2% sertaconazole cream twice daily. The lesion resolved completely within 8 weeks and there was no evidence of recurrence after 6 months of follow-up (Fig. 3).



Figure 3 Complete resolution of the plaque after finishing treatment.

In 1883, Majocchi¹ described a phenomenon in which dermatophytes, which usually remain limited to the stratum corneum, become more aggressive and invade the superficial dermis. Majocchi granuloma typically develops when a dermatophyte infection extends down a hair follicle in the setting of localized immunosuppression (most commonly a potent topical steroid) or systemic immunosuppression. The result is a granulomatous response in the dermis.² The commonly implicated organisms are *Trichophyton* species (*rubrum*, *mentagrophytes* complex, *violaceum*) as well as some nondermatophyte species such as *Aspergillus* and *Phoma*.⁴ Although historically *T. violaceum* has been the most commonly identified organism, Majocchi granuloma now a days is usually due to *T. rubrum*.

Two clinical forms of Majocchi granuloma, follicular and subcutaneous nodular, have been described.⁴ The follicular type usually develops after trauma, repeated shaving of hair-bearing areas, or topical corticosteroid treatment, and in cases of long-standing immunosuppression.^{5,6} The subcutaneous nodular type occurs in immunocompromised hosts. Our patient developed the follicular type, probably caused by endo-thrix-type infection due to anthropophilic *T. violaceum* secondary to the use of a potent topical steroid under occlusion, which could have led to deep penetration of the fungus. In modern medicine, systemic antifungals such as griseofulvin,⁷ itraconazole,⁸ and terbinafine⁹ are the mainstays of therapy as they are safe and effective. The duration of therapy should be of at least 4–8 weeks, and treatment should be continued until all lesions have cleared. In the reports in literature, nearly all lesions resolve without scarring within 6 weeks of starting antifungal. The response of Majocchi granuloma to oral terbinafine can be explained on the basis of its pharmacokinetics.¹⁰ Terbinafine is the preferred oral therapy for treating Majocchi granuloma not only for its superior efficacy in eliminating dermatophytes, but also because of its greater selectivity for the skin structures involved in Majocchi granuloma and fewer drug interactions than azole antifungals like itraconazole.⁹ We used terbinafine for its good antidermatophyte activity, adequate penetration into common sites of dermatophyte infection (stratum corneum and the hair follicle), lower rates of recurrence, low rate of drug interactions (its metabolism does not involve cytochrome P450), and its cost effectiveness when long-term therapy is warranted to prevent relapse.⁸ A diagnosis of Majocchi granuloma should always be considered in papular-pustular plaques, especially when the patient describes factors associated with local or systemic immunosuppression. Negative results of KOH examination or fungal culture of skin scrapings or of the purulent discharge do not exclude a diagnosis of Majocchi granuloma, which should be confirmed either by tissue culture or by PAS staining of histopathology samples. Although the detection of fungal spores or hyphae using special stains on histopathology samples will confirm the diagnosis, these structures may sometimes escape detection, as in our case.¹¹ Fungal spores and hyphae are usually detected within hairs or hair follicles and in dermal granulomas. Our inability to detect the fungal elements may have been due to the absence of hair follicles and the poorly defined granulomas in our biopsy specimen. To the best of our knowledge, this is the first reported case of Majocchi granuloma of the breast.

Conflict of interests

The authors declare no conflict of interest.

References

1. Majocchi D. Separa una nuova tricofizia (granuloma tricofitico), studi clinici micologici [A new Trichophyton granuloma: clinical and mycological studies]. *Bull R Accad Roma*. 1883; 9220-3.
2. Kurian A, Haber RM. Tinea corporis gladiatorum presenting as a majocchi granuloma. *ISRN Dermatol*. 2011;2011:767589.
3. Ilkit M, Durdu M, Karakaş M. Majocchi's granuloma: a symptom complex caused by fungal pathogens. *Med Mycol*. 2012;50:449-57.
4. Smith KJ, Neafie RC, Skelton HG, Barrett TL, Graham JH, Lupton GP. Majocchi's granuloma. *J Cutan Pathol*. 1991;18: 28-35.
5. Steiner UC, Trüeb RM, Schad K, Kamarashev J, Koch S, French LE, et al. Trichophyton rubrum-induced Majocchi's granuloma in a heart transplant recipient. A therapeutic challenge. *J Dermatol Case Rep*. 2012;6:70-2.
6. Elgart ML. Tinea incognito: an update on Majocchi granuloma. *Dermatol Clin*. 1996;14:51-5.
7. Novick NL, Tapia L, Bottone EJ. Invasive Trichophyton rubrum infection in an immunocompromised host. Case report and review of the literature. *Am J Med*. 1987;82:321-5.
8. Gupta AK, Groen K, Woestenborghs R, de Doncker P. Itraconazole pulse therapy is effective in the treatment of Majocchi's granuloma: a clinical and pharmacokinetic evaluation and implications for possible effectiveness in tinea capitis. *Clin Exp Dermatol*. 1998;23:103-8.
9. Gupta AK, Prussick R, Sibbald RG, Knowles SR. Terbinafine in the treatment of Majocchi's granuloma. *Int J Dermatol*. 1995;34:489.
10. Faergemann J, Zehender H, Jones H, Maibach I. Terbinafine levels in serum, stratum corneum, dermis-epidermis (without stratum corneum), hair, sebum and eccrine sweat. *Acta Derm Venereol*. 1991;71:322-6.
11. Gupta S, Kumar B, Radotra BD, Rai R. Majocchi's granuloma trichophyticum in an immunocompromised patient. *Int J Dermatol*. 2000;39:140-1.

U. Khanna,* T. Kumar Dhali, P. D'Souza, S. Chowdhry

Department of Dermatology, ESIPGIMS Basaidarapur, New Delhi, 15, India

* Corresponding author.

E-mail address: urmi23khanna@gmail.com (U. Khanna).

Secondary Syphilis Mimicking Lichen Planus in a Patient With HIV Coinfection[☆]



Sífilis secundaria simulando liquen plano en el paciente con infección por VIH

To the Editor:

Syphilis rates have been on the rise in recent years. In Spain, this sexually transmitted infection mainly affects men who have sex with men (MSM), many of whom are coinfecting with human immunodeficiency virus (HIV).¹ The natural history of both syphilis and HIV infection is affected by the coexistence of these diseases. The clinical manifestations of syphilis can progress more rapidly in patients with concomitant HIV infection, and aggressive and atypical forms of syphilis are also more common in this population.²

We report the case of 2 MSM with HIV infection who visited their dermatologist with genital lesions. The first man, aged 35 years, reported slightly pruritic lesions on the genitals that had appeared a month and a half earlier. He had applied topical corticosteroids, but there had been no improvement. Good immunologic control of his HIV infection had been maintained since 2009 without antiretroviral therapy. The physical examination revealed erythematous plaques with a lichenoid appearance and a tendency to coalesce on the dorsal aspect of the penis (Fig. 1 A and B). Blood test results brought by the patient, which included

biochemistry, a complete blood count, and coagulation studies, were unremarkable. Skin biopsy revealed lichenoid dermatitis with effacement of the basal layer, exocytosis of neutrophils, and a band-like lymphoplasmacytic infiltrate (Fig. 2A). Numerous plasma cells were also observed on the wall and around the vessels of the dermis (Fig. 2B). Immunohistochemical staining for *Treponema pallidum* was positive (Fig. 2C). Syphilis serology was requested; the nontreponemal rapid plasma reagin (RPR) test showed antibody titers of 1/128 and the treponemal antibody test was positive.

The second man, aged 29 years, had been diagnosed with HIV infection in 2011 and had achieved good immunologic control without pharmacologic treatment. He presented with mildly pruritic scrotal lesions of 1 month's duration. The lesions had been diagnosed as eczema and treated with topical corticosteroids, but there had been no improvement. The physical examination showed numerous pink plaques with a lichenoid appearance on the scrotum and at the base of the penis (Fig. 3). Laboratory tests, including biochemistry, a complete blood count, and coagulation studies, showed no alterations, and serology for syphilis was also negative. Histologic examination of a skin biopsy sample showed lichenoid dermatitis and immunohistochemical staining was positive for *T pallidum*. The RPR test was positive (titer, 1/64), as were the enzyme-linked immunosorbent assay and hemagglutination results.

Both patients were diagnosed with secondary syphilis mimicking lichen planus, with exclusive genital involvement. The clinical outcome was satisfactory in both cases, with complete resolution of lesions following treatment with intramuscular benzathine penicillin G 2.4 million units and a decline in RPR titers.

Cutaneous manifestations of secondary syphilis appear 3 to 12 weeks after the onset of the primary chancre,

[☆] Please cite this article as: Jiménez-Gómez N. Sífilis secundaria simulando liquen plano en el paciente con infección por VIH. *Actas Dermosifiliogr*. 2016;107:612-614.