

complement levels and antibodies against SS-B, ds-DNA, and Sm were all within normal limits. A second biopsy taken from a lesion on the dorsum of the nose showed parakeratosis, follicular plugging, liquefaction of the basal layer, and focal cellular infiltration in the dermis (Fig. 1B). There was eosinophilic material in the upper dermis, which was positive with Congo red and Dylon staining (Fig. 1C) and anti-cytokeratin antibody 34 β E12 staining (Fig. 1D). The direct immunofluorescence study revealed IgM deposits at the dermal-epidermal junction, but no amyloid deposits were observed in another DLE specimen taken from the back of the ear. A detailed eye exam revealed decreased lacrimation, but saliva production measured by the Saxon test was normal.

Few papers have reported on secondary amyloid deposition in DLE, but it has been suggested that this phenomenon might be underreported.¹ Powell et al.¹ reported 3 cases of DLE on the head and neck with amyloid deposition. They also retrospectively examined 18 cases of DLE, and detected amyloid deposition in 1 case. In a Japanese series, Khan et al.² detected amyloid deposition in all 4 cases of hypertrophic DLE analyzed but in just 1 of 12 cases of nonhypertrophic DLE. They speculated that cutaneous amyloid material might have been deposited secondary to abnormalities in the basement membrane zone following repeated sunlight exposure for long periods. Our case was not hypertrophic-type DLE, but amyloid deposition was detected on the nose, which had an evident keratotic appearance. The fact that amyloid deposits were not detected in the DLE lesion on the back of the ear suggests that UV radiation may play a role in inducing basement membrane impairment, resulting in secondary amyloid deposition. The amyloid material also stained positively for 34 β E12, suggesting that the origin of the amyloid was degenerating epidermal keratinocytes, which may be precursors of cutaneous amyloid. The presence of lamina densa-like substances or disturbed keratinization in the basal layers might also be involved in amyloid production.³ Our patient also developed Sjögren syndrome, although she did not show any symptoms of dry mouth. Because systemic

sclerosis was ruled out, the presence of serum antientromere antibody was considered to be associated with Sjögren syndrome, although coexistence of this condition with DLE is rare.⁴ Anticentromere antibody has been detected in 10% of Japanese patients with primary Sjögren syndrome⁵; these patients had lower mononuclear cell infiltration in the minor salivary glands, possibly explaining the lower frequency of dry mouth in this subgroup of patients. This hallmark symptom of Sjögren syndrome was also absent in our patient. In conclusion, there have been few reports on secondary amyloid deposition in DLE, but there may be more cases, especially in hyperkeratotic lesions in sun-exposed areas.

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Milia en plaque on the posterior surface of both auricles following radiation therapy[☆]



Milia en Placa en la Cara Posterior de Ambos Pabellones Auriculares Secundaria a Radioterapia

To the Editor:

Milia are benign epidermal cysts that can present as an isolated finding or associated with other clinical alterations.

[☆] Please cite this article as: Pisauri AM, Alvarez-Gracia A, Ferrandiz-Foraster C, Bassas-Vila J. Milia en placa en la cara posterior de ambos pabellones auriculares secundaria a radioterapia. *Actas Dermosifiliogr.* 2016;107:156–158.

When found in groups on an erythematous base, the lesions are called milia en plaque.

We present the case of a 48-year-old man with history of adenocarcinoma of the lung with cerebral metastases for which he received palliative treatment with holocranial radiotherapy at a dose of 30Gy in 10 fractions. The radiation fields included the auricles of the ear (2 opposing lateral photon beams to the central nervous system); the total dose received by the auricles of the ear was calculated as between 20 and 25 Gy. The patient came to our outpatient clinic for asymptomatic lesions that had arisen on the posterior aspect of the auricles of both ears 3 months earlier. Since his youth he had occasionally presented isolated lesions of a similar appearance, but the multiple lesions had developed a month after the radiotherapy. On examination, multiple millimetric whitish papules with a shiny surface on an erythematous base were found in groups bilaterally on the auricles of the ears (Figs. 1 and 2). Punch biopsy revealed an infundibular follicular cyst full of orthokeratotic



Figure 1 Milia en plaque cysts on the posterior aspect of the auricles of both ears.

keratin (Fig. 3). Postradiotherapy milia en plaque was diagnosed based on the patient's past medical history and the clinical and histologic findings.

Milia en plaque is rare. It was first described in 1903 by Balzer and Fouquet, who reported clusters of milium cysts on the posterior aspect of the auricles of the ear. In 1978, Hubler et al.¹ called the condition milia en plaque.

This form of milia usually affects middle-aged adults and there is a slight female predominance (3 to 1). Milia en plaque presents clinically as clusters of yellowish-white papules on an erythematous base; the lesions are usually asymptomatic but pruritus may occur.^{2,3} Typical sites include the earlobes, preauricular and periocular² regions, the nose, and the limbs.⁴

Cysts can arise spontaneously (primary milia)⁵ or after different triggers (secondary milia), such as recurrent trauma, topical treatment with corticosteroids or 5-fluorouracil, cryotherapy,⁶ chemotherapy (6-mercaptopurine),⁴ or radiotherapy (as observed in our patient).⁷ Two cases of postradiotherapy milia en plaque have been reported in the literature; in both cases the lesions arose on normal skin within the radiation fields.^{7,8}



Figure 2 Multiple, millimetric whitish papules on an erythematous base on the posterior aspect of the auricle of the ear.

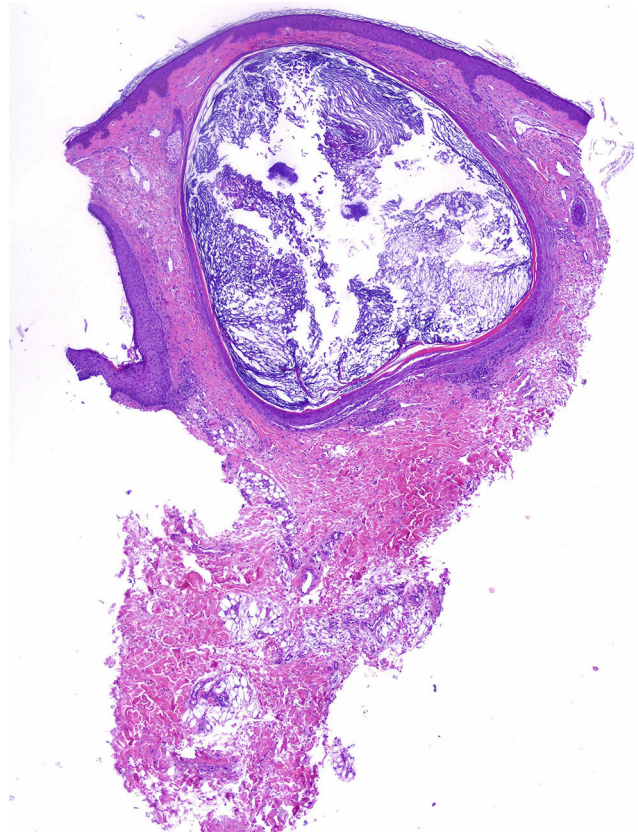


Figure 3 Cystic cavity in the superficial dermis. The cyst contains orthokeratotic keratin and is lined by a stratified squamous epithelium. Hematoxylin-eosin, original magnification x10.

Follicular changes induced by chemotherapeutic agents have been described in the literature, especially with drugs that target the epidermal growth factor receptor. But the most common chemotherapy-induced changes described in the literature occur in the eccrine gland or duct, in the form of squamous syringometaplasia. We believe that chemotherapeutic agents were not relevant to the pathogenesis in our patient as no lesions were observed outside the fields of radiation.⁴

Histology reveals small cysts containing orthokeratotic keratin, located in the dermis. The cysts are lined by a squamous epithelium with a granular layer, and are accompanied by a mild mixed or lymphocytic perivascular infiltrate.^{2,4}

The pathogenesis of milia en plaque is unknown, although damage to the follicular infundibulum may be assumed in our case due to a direct effect of radiotherapy on the follicular epithelium. In other cases, numerous other factors that could in some way affect the follicular epithelium may be involved, including dermabrasion, 5-fluorouracil, acitretin, or solar damage.⁶

The differential diagnoses that should be considered include comedonal nevus, trichoadenoma of Nikolowski, steatocystoma multiplex, Favre-Racouchot disease, follicular mucinosis, and folliculotropic mycosis fungoides.⁹ Song JC et al.¹⁰ published a case in which skin metastases from a parotid gland carcinoma had a milia-like appearance. Skin metastases must be considered, though the absence

of atypia in the histopathology of the lesions in our patient excluded this diagnosis.

Treatment can be provided to patients with symptomatic lesions or for cosmetic reasons. Numerous treatments with satisfactory results have been described in the literature, including topical retinoids, cryotherapy, electrocoagulation, radiofrequency, carbon dioxide laser,² surgical excision, oral tetracyclines, and photodynamic therapy.^{3,9} Given the benign nature of the disease and the absence of any cosmetic issue, our patient was not a candidate for treatment.

In conclusion, milia en plaque is a rare but easily diagnosed disease. No cause is detected in the majority of cases. In our case, the recent history of exposure to radiotherapy, with a plausible temporal relationship, would suggest a causal relationship.

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Why Is Mohs Micrographic Surgery Underused in the Treatment of Dermatofibrosarcoma Protuberans in Children?☆



¿Por qué la cirugía micrográfica de Mohs está infrutilizada en el tratamiento del dermatofibrosarcoma protuberans infantil?

To the Editor:

Dermatofibrosarcoma protuberans (DFSP) is a slow-growing, intermediate-grade fibrohistiocytic tumor characterized by high rates of local recurrence but low metastatic potential. Few cases of congenital forms have been reported, but this is probably because early lesions can go unnoticed or be confused with other entities. There is evidence that recurrence rates are lower with Mohs micrographic surgery (MMS) than

with conventional surgery with wide margins.¹ Nonetheless, in 3 recently published pediatric series, wide excision was used in the vast majority of patients (41/43).²⁻⁴ We present a case of congenital giant-cell fibroblastoma, a histologic subtype of DFSP, which was diagnosed and treated in an 8-year-old boy. In this report, we highlight the advantages of MMS and the use of negative-pressure wound therapy (NPWT) in children with DFSP.

The patient was an 8-year-old boy with no relevant past history who was seen for a lesion in the right axilla that had grown and become progressively harder. The lesion, which was initially flat and reddish in color, had been present since birth. Examination revealed an indurated, mobile plaque measuring 4 × 2.5 cm. The plaque was pink but had a purple, slightly elevated area. The regional lymph nodes were not enlarged. Biopsy showed a poorly delimited tumorous lesion composed of spindle and oval cells that occupied practically the entire dermis and extended down into the subcutaneous tissue. Pseudovascular spaces surrounded by multinucleated giant cells were also visible. The spindle cells expressed CD34 and were negative for S-100 protein and smooth muscle actin (Fig. 1). A diagnosis of giant cell fibroblastoma was established, and extracutaneous invasion was ruled out by magnetic resonance imaging. The tumor was excised under general anesthesia. The technique chosen was slow MMS (Breuninger technique), with excision of 1-cm margins in the first stage (Fig. 2). This technique consists of the en

☆ Please cite this article as: Jubert E, del Pozo LJ, Saus C, Martín-Santiago A. ¿Por qué la cirugía micrográfica de Mohs está infrutilizada en el tratamiento del dermatofibrosarcoma protuberans infantil?. *Actas Dermosifiliogr*. 2016;107:158-162.