



Figure 3 Periungual telangiectasias observed with capillaroscopy.

diseases such as rheumatoid arthritis, systemic lupus erythematosus, Sjögren syndrome, and dermatomyositis.² Dermatomyositis (specifically, juvenile dermatomyositis) is the autoimmune disease most frequently related to the development of lipodystrophy.³ The prevalence of lipodystrophy in juvenile dermatomyositis varies among studies from 10% to 40%. In the only study performed with a large number of patients, acquired lipodystrophy was observed in 8% of patients with juvenile dermatomyositis, while only 1 of the 692 adult patients with dermatomyositis had lipodystrophy.⁴ To date, only 2 cases of lipodystrophy associated with adult-onset dermatomyositis have been published.^{5,6} The reasons for this lack of association are unknown.

One study has shown that a number of manifestations of dermatomyositis, such as joint rigidity, muscle atrophy, panniculitis, and calcinosis, could be related to the development of lipodystrophy.⁴ Of all these signs, our patient had only joint rigidity in the hands.

The etiology and pathogenesis of lipodystrophy associated with autoimmune disease remain unclear. Patients with partial lipodystrophy often have low C3 levels, which may trigger the alternative complement pathway and promote development of the disease.²

The majority of patients described in the literature developed lipodystrophy years after the onset of dermatomyositis, as a late complication of severe, chronic disease;

our patient, however, developed symptoms of dermatomyositis and lipodystrophy practically simultaneously. In contrast to what occurs in most cases of partial lipodystrophy, there were no metabolic abnormalities in our patient. We were also unable to demonstrate etiologic or pathogenic mechanisms associated with immunologic abnormalities such as low levels of C3. In agreement with reports from similar cases in the literature,^{5,6} treatment with oral prednisone and mycophenolate mofetil did not improve the facial lipoatrophy in our patient.

We have presented a very rare case of adult lipoatrophy simultaneously associated with dermatomyositis.

Based on published data, it would be prudent to closely monitor all patients with dermatomyositis who develop a loss of subcutaneous fat in order to progressively rule out abnormalities in the metabolism of lipids and carbohydrates.

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Photodynamic Therapy With Methyl-aminolevulinate can be Useful in the Management of *Scytalidium* Infections

La terapia fotodinámica con metilaminolevulinato puede ser útil en el manejo de las infecciones por *Scytalidium*

To the Editor,

Scytalidium spp., recently renamed *Neoscytalidium* spp., are keratinophilic molds that cause superficial disease

(skin, nails) which is indistinguishable from and sometimes occurs concomitantly with dermatophyte infections.¹ While infection with these fungi, and in particular *Scytalidium dimidiatum*, is fairly common in tropical climates, it is rare in Spain. The clinical and microbiological diagnosis of such an infection is a challenge, especially because the growth of these fungi is suppressed by the antimicrobial component of the media routinely used for the isolation of dermatophytes. There is, at present, no effective oral or topical treatment for skin and nail infections caused by *Scytalidium*.^{2,3}

Antimicrobial photodynamic therapy (PDT) is an emerging treatment for infections. This process involves the application of a photosensitizer that binds to the microor-



Figure 1 Onychomycosis of the first and second toes of right foot caused by *Scytalidium dimidiatum* infection, before photodynamic therapy.

ganisms. When excited by an appropriate light source in the presence of oxygen the photosensitizer produces reactive oxygen species that induce cell death, either by apoptosis or necrosis.⁴ Our group had recently demonstrated the usefulness of methyl-aminolevulinate PDT in the treatment of onychomycosis caused by non-dermatophyte molds⁵ and yeasts.⁶

A 61-year-old healthy woman from Cameroon, resident in Spain for more than 5 years, presented with a very long history of dystrophic toenails. She had been diagnosed with onychomycosis caused by *Aspergillus* spp. and treated with several topical antifungal agents with no improvement. On examination, she presented thickened, opaque nail plates with a yellow or brown discoloration and cracked surfaces (Fig. 1). Significant scaling was observed around the nails, between the toes, and on the soles. Her fingernails were unaffected.

Nail clippings and periungual skin swabs confirmed *S. dimidiatum* on microscopy and cycloheximide-free agar mycological culture. No dermatophytes were found, but colonies of *Aspergillus* spp. were seen on Sabouraud plates from 1 of the 2 affected toenails.

Bearing in mind the failure of previous antifungal treatments and the lack of any effective treatment for this infection, and encouraged by our good results in onychomycosis,^{5,6} proposed this treatment to the patient.

For five days prior to PDT, a combination of 40% urea and 1% bifonazole (Mycospor Onicoset®) ointment was applied every night in occlusion to the nail plates. On the day of treatment, methyl-aminolevulinate (Metvix®) was applied to the nail plates and periungual skin, which were then covered with an occlusive dressing (Tegaderm®) and protected from light for 3 h, as previously described.⁷ When the dressing was removed, the nails were cleaned with 70% ethanol and irradiated using a 635 nm light emitting diode lamp (Aktilite®, 37 J/cm²). No side effects were observed during or after treatment. The same procedure was repeated 1 week later and every 2 weeks thereafter. Microbiological cultures became negative after the third session. A total of 4 sessions were administered.

Clinical improvement was noticed after 2 months. Four months later, the patient was clinically and microbiologically cured according to the standard criteria (Fig. 2). After 6 months of follow-up, cultures became positive in 1 of the 2 nails, but the nails remained clinically cured and there was no evidence under microscopy of nail penetration.



Figure 2 Nail plates showing significant clinical improvement 4 months after 4 sessions of methyl-aminolevulinate photodynamic therapy.

Infection with *S. dimidiatum* accounts for under 1% of cases of onychomycosis and seldom responds to amorolfine or terbinafine.² Recently, an intermittent posaconazole regimen has been proposed to treat superficial *S. dimidiatum* infection.³ However, all of these drugs are expensive and can have significant adverse effects. Moreover, there is little or no evidence of their effectiveness in this setting.

Fewer than 50 cases of onychomycosis treated with aminolevulinic acid or methyl-aminolevulinate PDT have been reported and most of these were caused by dermatophytes.⁵⁻⁸ Our group obtained good results with methyl-aminolevulinate, using one protocol based on the one previously reported by Piraccini et al.⁵⁻⁷ In localized mycosis, the therapeutic effect of PDT is twofold: the treatment directly kills the fungus and also reinforces the fungicidal effect by stimulating host immune cells, especially neutrophils.⁴

Although in previous cases^{5,6} we have successfully used a protocol with a 2-week interval between sessions, in this case we made some modifications. We reduced the interval between the first and second sessions to 1 week in an attempt to reduce the recovery capacity of the fungus. The results of *in vitro* experiments with dermatophytes suggest that reducing the interval between PDT sessions will improve the fungicidal effect.⁹

Microbiological diagnosis of onychomycosis is difficult; in fact up to 90% of the cases microbiologically diagnosed the first laboratory result may be negative.¹⁰ In our case, however, it is unlikely that the culture results after PDT were false negatives because no antimicrobial substances were present in the medium used. Another problem associated with onychomycosis in general, and *Scytalidium* infection in particular, is that recurrence is frequent. In this patient we observed microbiological reappearance of the fungus without clinical recurrence. A possible solution for recalcitrant cases might be periodic administration of PDT or a combination of PDT with an antifungal drug over a period of time.

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Urticaria-Like Reaction Secondary to Photodynamic Therapy in 2 Pediatric Patients[☆]

Reacción urticariforme secundaria a terapia fotodinámica en 2 pacientes pediátricos

To the Editor:

Photodynamic therapy (PDT) is a light therapy that involves the production of reactive oxygen species and free radicals to induce oxidation in biological tissues. The effect is achieved through the application of a photosensitizing agent, which is then activated by exposure to wavelength-specific light.¹ The most commonly used photosensitizer is methyl aminolevulinate (MAL). PDT is primarily used in the treatment of basal cell carcinoma, actinic keratosis, and Bowen disease, although it can also be used to treat certain non-neoplastic diseases. The most common side effects are pain and local inflammation; wheals are very rare.² We present the cases of 2 pediatric patients who developed urticarial reactions to PDT.

The first patient was an 11-year-old girl with nevoid basal cell carcinoma syndrome (Gorlin syndrome) who presented with the characteristic clinical manifestations, including frontal bossing, hypertelorism, small ears, syndactyly, palmar pits, and a large number of small, pedunculated, papular lesions on the neck, axillas, groin, and popliteal fossae. Histologic examination of these lesions revealed basal cell carcinoma. The lesions had previously been treated with cryotherapy, electrocoagulation, and imiquimod. PDT treatment was prescribed and administered on several lesions on the upper back and the neck according to the standard protocol: MAL 160 mg/g cream (Metvix) applied under occlusion for 3 hours followed by illumination using an Aktelite lamp at a dose of 37 J/cm². Three minutes after starting illumination, semicircular wheals appeared in the area being treated, and light exposure was stopped (Fig. 1A). The lesions disappeared after a few hours without treatment and did not recur. Some months later the patient underwent provocation testing with the MAL cream applied under occlusion for 3 hours followed by illumination. Wheals appeared in the illuminated area where the cream had been applied, but no lesions were observed in the area exposed to light without prior application of cream (Fig. 1B). The wheals disappeared spontaneously and there was no recurrence.

The second patient was a 4-year-old girl with no relevant past history or family history who presented with brownish, erythematous, maculopapular lesions in a blaschkoid distribution affecting the dorsum and anterior aspect of the

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