

other authors have proposed it as a fibroproliferative hamartoma that simulates the nail matrix histology.^[2] Onychomatricoma has a female to male ratio of 2.16:1; mean age of presentation is 51 years and occurs more frequently in Caucasian patients. Less frequently, it occurs in African Americans and exceptionally in Mexican patients. Its predominant location is the fingernails (75%). Predisposing factors such as trauma or onychomycosis have been suggested.

Pathology shows a wide histological spectrum as recently described by Perrin *et al.*,^[3] but typically consists of fibroepithelial tumor composed of a proximal pedunculated base and a distal zone with multiple projections. There is a two-layered stroma with a collagenous and fibroblastic superficial coat and a deep core with less cellularity and thicker collagen; a V-shaped hyperkeratogenous zone can be observed. Beyond the lunula, the nail plate is thickened and burrowed with cavities containing serous fluid. Immunohistochemistry for cytokeratin 5 and 14 are positive, along with K17, K6, K16, and K75 in most cases, suggesting a differentiation towards the nail bed and the nail isthmus.^[4] Recently CD34 was also reported to be positively expressed.^[3]

Magnetic resonance shows tumoral core in the matrix with invaginations into the funnel-shaped nail plate; the center has a low intensity signal and peripheral and axial images show the holes in the nail plate and the tumoral digitations.^[5]

Differential diagnosis for onychomatricoma include fibrokeratoma of the nail matrix, glomus tumor, squamous cell carcinoma, Bowen's disease, viral warts and unguis fibroma. This last diagnosis was the clinical impression in both patients described above; however, it can be ruled out by the hyperplastic and onychogenic nature of the epithelium. Complete surgical excision is the first line of treatment. Recurrence and malignant transformation have not been reported, although dysplasia may be present.^[3]

Onychomatricoma is an infrequent subungual tumor that primarily occurs in Caucasians. Only two cases have been previously reported in Mexican patients. Treatment should be individualized but complete surgical excision remains as the gold standard. Because onychomatricoma can easily mimic other unguis diseases, physicians and particularly dermatologists should be acquainted with the proper diagnosis and treatment of this adnexal tumor.

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Localized scleroderma *unius lateri* and *Borrelia burgdoferi* infection

Sir,
Localized scleroderma (named also morphea) is an inflammatory autoimmune skin sclerosis of unknown etiology. A causative role of *Borrelia burgdoferi* infection has been proposed.^[1] The *B. burgdoferi* infection can be assessed with serological test as well as polymerase chain reaction (PCR) on blood and skin sample.^[2] Here we report the case of a Moroccan 27-year-old male, living in Italy since 10 years. In October 2010, he

presented to the neurologist for progressive weakness and hypotrophy of his left limbs, which started 4 years before [Figure 1a]. For the presence of scleroatrophic plaques on the affected areas and the left side of the trunk, he was referred to the dermatologist. Physical examination revealed multiple oval and round circumscribed areas of deep induration of the skin on the left upper and lower limbs, left half of the trunk and on the interscapular region. Sclerodermic plaques were painful, severely atrophic and hyperpigmented, adherent to the underneath tissues, with ill-defined borders, brownish color and no evidence of telangiectasia [Figures 1b and 1c]. Neither erythema nor lilac ring was present. Since their appearance, they progressively increased in number and became more painful, mainly in the last 6 months. Hematochemical investigations including C3 and C4 complement factor levels, antidouble strand DNA antibodies, antibodies to extractable nuclear antigens (RNP, Sm, SS/A, SS/B), topoisomerase I (Scl/70), antisynthetasis (Jo1), IgG phospholipid units (GPL), IgM phospholipid units (MPL), C1q, antinuclear antibodies (ANAs), Weil-Felix test and *B. burgdoferi* serology (ELISA assay), were performed. The results were within the normal limits except for ANAs titer (4.7 index, normal values: 0.00 – 1.50 index; ANA-IIF: Positive for speckled/nucleolar type, medium positivity) and *B. burgdoferi* IgM (LYMEM 136.0 AU/ml; normal values: 0.0-22.0). The patient did not remember any bite in the previous months. A skin biopsy of a recent lesion on the distal third of the left thigh was performed. The histological evaluation confirmed the diagnosis of localized scleroderma [Figure 2]. Direct immunofluorescence (DIF) analysis was negative. Nailfold capillaroscopy did not show microvascular abnormalities. PCR analyses for *Borrelia* detection,^[2] performed on DNA obtained from peripheral blood and skin biopsy samples, were negative. The patient was then treated with benzathine penicillin intramuscular 12,000,000 IU per day for 20 days, and after 1 month, a 20 days cycle was repeated. No other medication was prescribed. After 4 months he showed a clear improvement of the symptoms and serological test for *B. burgdoferi* resulted negative. Physical examination showed that the plaques were softer and less adherent to the underneath tissues than before the benzathine penicillin treatment [Figure 3].

Neurological examination showed a moderate hypotrophy of the proximal muscles of the left arm, a mild hypotrophy of the proximal muscles of the left leg and a mild diffuse strength reduction, 4 out of 5 of the Medical Research Council scale (MRC scale) on



Figure 1: (a) Clinical feature in the study patient showing striking hypotrophy of the left limbs; (b, c) Hyperpigmented atrophic plaques over the left part of the trunk and left lower limb (arrows)

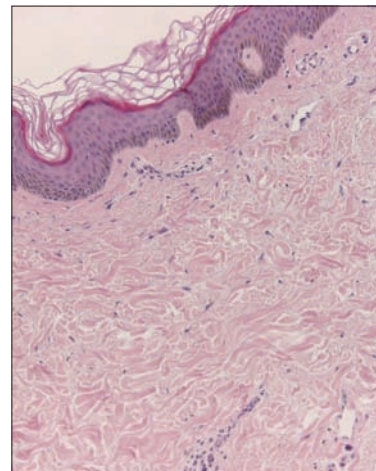


Figure 2: H and E-stained section of the punch biopsy specimen showing marked sclerosis of the dermis with disordered arrangement and parallelism of collagen fibers, lack of skin appendageal structures and mild chronic inflammatory infiltration (x220 magnification)

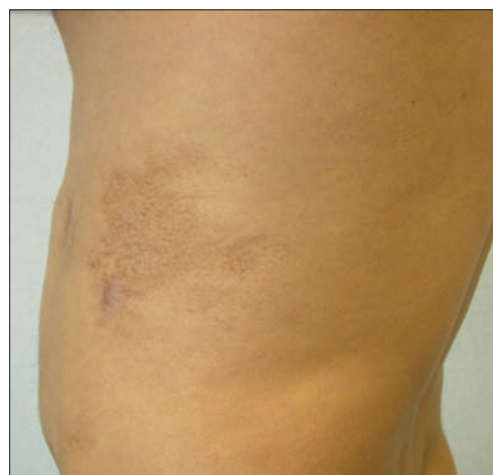


Figure 3: Clinical feature after benzathine-benzyl penicillin treatment: The plaques were softer, lighter in color and less adherent to the underneath tissues than before treatment

the same limbs. Remaining examination was entirely normal. The neuroimaging workup (brain MRI with MRA, cervical MRI), and the neurophysiological tests (motor-evoked potential, sensory-evoked potentials, electroneurography and electromyography) were normal.

This is an interesting case of a patient with a localized scleroderma and serological signs of *B. burgdoferi* infection that sought medical care for neurological symptoms. Yet, he had no central or peripheral nervous system lesion. The muscle wasting and weakness were secondary to the dermatological lesions.^[3] Nevertheless, *B. burgdoferi* detection by PCR was negative. This fact could be explained with the paucity of the bacteria, which did not reach the sensitivity of the analysis in this case. It is known, indeed that in the manifestation of long-standing infection of *B. burgdoferi*, the paucity of microorganisms could lead to a low detection rate by PCR, especially when the analysis is performed on archival biopsies.^[4,5] Negative results obtained by PCR did not prove the absence of the microorganism, for low sensitivity (45.2%) of the method.^[4]

Nevertheless penicillin provided good results in line with previous reports on the efficacy of this treatment for localized scleroderma.^[4,5]

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Multifocal dermatofibrosarcoma protuberans

Sir,

Dermatofibrosarcoma protuberans (DFSP) is a relatively uncommon soft tissue neoplasm with low to intermediate grade malignancy.^[1,2] It is a clinically distinctive, raised/atrophic or sclerotic cutaneous fibrous tumor that exhibits a pattern of slow, infiltrative growth. It has a marked tendency to recur locally after surgical excision^[3] with rare cases metastasizing to regional lymph nodes or distant sites.^[2] It constitutes less than 0.1% of all cutaneous malignancies and 1% of all soft tissue sarcomas.^[3] Most lesions occur over the trunk or proximal extremities and are focal in nature.^[1-3] Early diagnosis, wide excision and regular follow up are essential components of management of DFSP. Herein we report a case of multifocal DFSP.

A 59-year-old male presented with history of having concurrently developed 6 years back two asymptomatic swellings; a protuberant mass over left lumbar region and an ulcerated one on right mandible. The abdominal swelling, stated to be pea-sized at the onset, progressed gradually over the initial 5 years and relatively rapidly over the month preceding his presentation to us. Likewise the facial swelling too progressed from a millet-sized papule to a palm-sized plaque and ulcerated in the preceding month following the application of a caustic agent. Examination of the abdominal wall revealed a firm 5 × 5 cm, yellow to brown plaque with a central protuberance surmounted by an ulceration. The surrounding skin appeared retracted, shrivelled and hyperpigmented [Figure 1a]. The second-firm, erythematous, blanchable centrally encrusted 5 × 3-cm-sized plaque- seen over the right

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