

Morbihan disease and extrafacial lupus miliaris disseminatus faciei: a case report

Kenzen Kou,^{ab} Keishi Chin,^b Setsuko Matsukura,^b Takeshi Sasaki,^c Akinori Nozawa,^c Michiko Aihara,^a Takeshi Kambara^b

From the ^aDepartment of Environmental Immuno-Dermatology, Yokohama City University Graduate School of Medicine; ^bDepartment of Dermatology and ^cDepartment of Pathology, Yokohama City Medical Center, Yokohama, Japan

Correspondence: Dr. Kenzen Kou · Department of Environmental Immuno-Dermatology, Yokohama City University Graduate School of Medicine and Department of Dermatology Yokohama City Medical Center, Yokohama, Japan · T: +81-45-787-2675; F: +81-45-786-0243 kenzenkou@yeah.net

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Lupus miliaris disseminatus faciei (LMDF) is characterized by discrete dome-shaped papules on the bilateral face and neck. We report the first case of LMDF with a widespread distribution of extrafacial papules and concomitant bilateral eyelid edema. A unique case of a 30-year-old man presented with nonpruritic, painless, papular eruptions (3-5 mm in diameter) involving the bilateral trunk and upper extremities, along with bilateral eyelid edema. There was no facial eruption involved. The outbreak initially involved the back, and gradually spread to the chest and wrists. The histologic examination of the eyelids revealed histiocytic epithelioid cells, dermal intercellular edema, and an expanded vascular space, which led to the diagnosis of Morbihan disease. The immunohistochemical staining of the truncal eruption revealed nodular inflammatory changes involving the middle to lower dermis around the follicles, containing histiocytes, multinucleated giant cells, and small lymphocytes. This led to the diagnosis of LMDF. The truncal papular eruption ultimately improved with roxithromycin treatment, but the eyelid edema required surgical treatment.

Morbihan disease is a very rare condition, which manifests as persistent upper facial lymphedema, often involving the eyelids.¹ The underlying pathogenic mechanisms are unclear; however, a link with rosacea has been suggested. There have been very few case reports of Morbihan disease published to date.¹⁻⁴

Lupus miliaris disseminatus faciei (LMDF) is an epithelioid cell granuloma of the dermis that can progress to central necrosis.⁵⁻⁷ Although the pathogenesis of LMDF is unclear, a link with rosacea has also been suggested. Infiltrates comprising lymphocytes, histiocytes, and neutrophils are hallmarks of the condition.⁶ LMDF is characterized by discrete brown/red to yellow dome-shaped papules on the face, including eyelids.^{8-10,11}

We herein describe a unique case of Morbihan disease (with bilateral eyelid edema) and extensive truncal LMDF.

CASE

A 30-year-old man with a history of hypertension pre-

sented with eyelid edema (**Figure 1a**) and worsening truncal lesions. The rash comprised 3 to 5 mm diameter, painless, non-pruritic, papules, that initially involved the back and gradually spread to the chest, abdomen, and extremities. Chest x-ray and chest computed tomographic findings were normal, as were liver enzyme levels. An anti-nuclear antibody test was negative. The patient was initially diagnosed with folliculitis and received treatment with minocycline 100 mg daily. The symptoms appeared to improve after 5 months of treatment. However, abnormal liver enzyme levels led to the termination of minocycline treatment. Subsequently, oral dapsone 50 mg daily was prescribed for 2 months; however, the patient's symptoms did not improve. Skin biopsies of the edematous eyelids and truncal eruption were performed.

The histologic examination of the eyelid biopsy revealed histiocytic epithelioid cell, dermal intercellular edema, and an expanded vascular space (**Figure 2**). Alcian blue staining was negative and D2-40 staining specific for lymphatic endothelial cells was positive, and

there was evidence of lymphangiectasia and lymphatic obstruction in the eyelid tissue. Acid-fast bacilli and fungi stains of the biopsy were negative. The histologic examination of the truncal eruption revealed nodular inflammatory changes, involving the middle to lower dermis around the follicles, containing histiocytes, multinucleated giant cells, and small lymphocytes. The necrosis of epitheloid cell granulomas in the center of the

lesions was also noted. Based on the histologic findings, eyelid edema was diagnosed as Morbihan disease, while the truncal lesions were diagnosed as LMDF.

Oral roxithromycin 300 mg daily was prescribed and the truncal eruptions gradually resolved within 1 year. However, the eyelid edema showed minimal improvement. The subsequent surgical treatment of eyelid edema resulted in significant symptomatic improvement (Figure 1b).

DISCUSSION

To our knowledge, this unique presentation of Morbihan disease (with bilateral eyelid edema) and extrafacial LMDF has not previously been described.

The histologic examination of eyelid tissue revealed histiocytic epitheloid cell, dermal intercellular edema, and an expandable vascular space accompanied by lymphatic blockage. These characteristics are similar to those reported in a previous case of persistent eyelid edema,¹ which was subsequently diagnosed as Morbihan disease. This very rare condition, also known as “rosacea lymphedema” and “solid persistent facial edema,” is a complication of rosacea, which does not tend to regress spontaneously. The pathogenesis of Morbihan disease remains uncertain,¹² and thus, it is a diagnosis of exclusion after ruling out other conditions known to cause eyelid swelling. Other case reports of this disease have described dilated blood vessels, perifollicular fibrosis, and perivascular and perifollicular infiltration of histiocytes, lymphocytes, and neutrophils. Hence, we are confident the eyelid edema in our patient was due to Morbihan disease.

Similar to previous reports, we found that treatment with minocycline failed to resolve eyelid edema. Moreover, roxithromycin also resulted in minimal im-



Figure 1. Patient with bilateral eyelid edema (a) before treatment and (b) after surgical treatment, showing the resolution of eyelid edema.

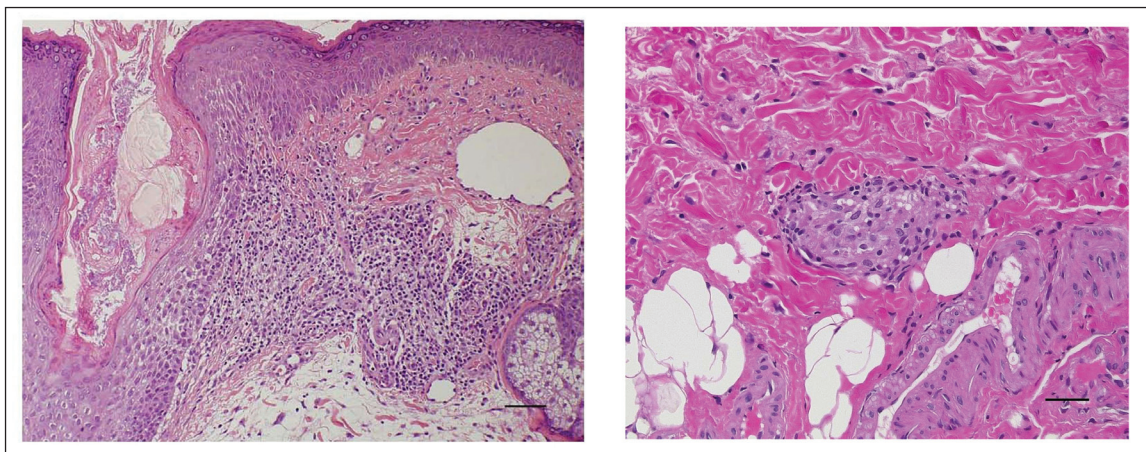


Figure 2. Histological images of Eedematous lower eyelid tissue histological images showing histiocytic epitheloid cell and dermal intercellular edema surrounding the follicles, with visibly dilated vascular space (100 μm at 200× and 400× magnification, respectively).

provement. Eventually, surgical drainage was performed and definitive resolution of symptoms, similar to that associated with the surgical management of periorbital lymphedema, was noted. Hence, surgical drainage would appear to be an effective treatment for eyelid edema in Morbihan disease. It must be noted, however, that we did not treat our patient with prednisolone, a treatment found to be effective in a previous case.¹

Contrary to its name, LMDF is not associated with systemic lupus erythematosus.⁹ However, the condition has been linked to rosacea,⁵ and the etiology of LMDF is likely multifactorial.⁷ One proposed mechanism involves a granulomatous reaction to ruptured epidermal cysts or hair follicle destruction.¹³ Depending on the contribution of these factors in a given case, the clinical and histopathological presentation of LMDF may vary considerably. Although LMDF with extrafacial manifestations are common, LMDF without concurrent facial involvement is rare.¹¹ In our case, the patient presented with widespread papular eruptions over the trunk, arms, and wrists, without typical facial manifestations. Therefore, our case is thought to be particularly noteworthy.

Rapidly diagnosing our patient was challenging because of the controversial pathogenesis of LMDF. The disease shows caseating and noncaseating epithelial cell granulomas that can resemble other granulomatous disorders.⁵ Granulomas in LMDF tend to appear in association with pilosebaceous units as per rosacea.¹¹ Unlike rosacea, however, LMDF can involve facial (eyelid, up-

per lip) and extrafacial locations,⁵ and usually spontaneously regresses over time. Therefore, LMDF appears to be a polyetiologic rosacea-like syndrome with unique presentation.⁵ Esteves et al reported successfully treating a patient with LMDF using oral minocycline and deflazacort,¹⁴ while Liao et al also successfully treated a patient with LMDF of the eyelids with minocycline.⁹ Conversely, in another report, a patient with LMDF of the neck and chest continued to worsen with minocycline and doxycycline treatment.¹¹ Our patient did respond to minocycline; however, this regimen had to be discontinued due to liver toxicity. The persistent LMDF eruption ultimately responded to roxithromycin treatment.

In summary, we have described a rare case of synchronous Morbihan disease and extrafacial LMDF. We hope this case will remind physicians of the existence of these conditions. Our case also underscores the necessity for careful follow-up of patients with LMDF because this condition is often difficult to diagnose and treat, and may mask other medical conditions. As both the persistent eyelid edema attributed to Morbihan disease and the extrafacial eruption attributed to LMDF share characteristics within the spectrum of rosacea, it is possible that this unique pattern of dermatological manifestations may share a similar pathogenic mechanism.

Conflict of interest

The authors declare that there is no conflict of interest in this article.

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