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CASE REPORT

Morbihan Disease May Induce Nonspecific Inflammatory of Extra-Facial Region: A Case Report and Literature Review

FeiFang Deng[1,2], Peng Wang[2]

[1]Women and Children's Health Care Hospital of Luohu, Shenzhen, People's Republic of China; [2]Department of Dermatology, Affiliated Nanshan Hospital of Shenzhen University (Shenzhen Nanshan People's Hospital), Shenzhen, Guangdong, People's Republic of China

Correspondence: Peng Wang, Affiliated Nanshan Hospital of Shenzhen University (Shenzhen Nanshan People's Hospital), Shenzhen, 518000, People's Republic of China, Tel +861382313270 4, Email wangpeng02@126.com

Abstract: This case describes a 29-year-old man with Morbihan disease, characterized by a 5-year history of recurrent facial erythema, papules, and pustules, followed by persistent right face edema. Initially misdiagnosed as lupus erythematosus, he received multiple treatments with only transient relief. Clinical manifestations combined with pathological findings confirmed a diagnosis of Morbihan disease associated with rosacea. Notably, the patient developed erythema and papules on the inner thighs, along with small papules on the trunk and upper limbs for 3 months. A biopsy revealed nonspecific inflammation. Since the morphology of these lesions differed from any known skin disease and no other cause could be identified, we believe this may be a manifestation of the disease. Extra-facial manifestation not previously reported. Various therapies were attempted: doxycycline and azelaic acid improved erythema; baricitinib was ineffective; while omalizumab and intense pulsed light (IPL) achieved partial improvement in erythema and edema. Long-term isotretinoin therapy provided sustained remission without relapse during tapering, confirming its efficacy and safety. This case expands the clinical spectrum of Morbihan disease by documenting extra-facial involvement and reported isotretinoin as an effective long-term therapy, with omalizumab and IPL as promising adjuncts. Furthermore, the literature review (2004–2025) summarizes the clinical features, pathology, and treatment responses of reported cases, showing a male-to-female ratio of 2.81:1. Lymphatic or vascular dilation was significantly correlated with a favorable outcome. Conversely, dermal edema and granuloma were inversely associated with treatment efficacy, indicating poorer prognosis. However, based on uncontrolled data, these findings are strictly hypothesis-generating and do not establish a definitive treatment hierarchy.

Keywords: case report, Morbihan disease, literature review

Introduction

Morbihan disease is an uncommon and challenging disorder, characterized by chronic and persistent erythema and edema predominantly affecting the central and upper facial regions, including the zygomatic area, nose, glabella, eyelids, and forehead.[1,2] First described by Robert Degos in 1957, the disease is named after the French region where the initial case was observed.[3] Although frequently associated with rosacea, approximately 20%–30% of cases arise in patients without any prior history of the condition.[4] Given the extreme rarity of this condition, reliable incidence or prevalence data in China are currently unavailable. Furthermore, to our knowledge, no specific medications have been definitively reported to induce Morbihan disease.

The pathophysiology of Morbihan disease is thought to involve a complex interplay of chronic inflammation and lymphatic dysfunction. It is generally regarded as an end-stage complication of recurrent vasodilation associated with rosacea, where a loss of vascular integrity leads to fluid extravasation. This chronic inflammatory state eventually causes lymphatic vessel injury and remodeling, impairing drainage and resulting in lymphedema.[1,5] Diagnosis is mainly clinical and histopathological findings, requiring careful exclusion of mimicking conditions. Differential diagnoses typically include angioedema (which typically resolves within 24–48 hours),[6] and classic rosacea (which is primarily characterized

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by papules and pustules),[5] as well as systemic lupus erythematosus, sarcoidosis, and orofacial granulomatosis. Importantly, Morbihan disease does not undergo spontaneous remission, responds poorly to therapy, and lacks established effective treatment options, making timely recognition particularly important.[5,7] Additionally, the nonspecific cutaneous manifestations of this disease outside the facial region have not been reported in the literature. Here, we describe the case of a 29-year-old Chinese male with a 5-year history of recurrent facial erythema, edema, and papulopustular lesions. He was initially misdiagnosed lupus erythematosus before being ultimately confirmed to have Morbihan disease associated with rosacea. He tried various medications, ultimately achieving control with isotretinoin. This case also presented unique extra-facial involvement, highlighting nonspecific inflammation in the extra-facial region and the efficacy and safety of long-term low-dose isotretinoin treatment.

Case Presentation

A 29-year-old man presented with a five-year history of recurrent facial erythema, papules, and pustules, along with persistent edema on the right side of the face. He was previously misdiagnosed with lupus erythematosus and treated with oral minocycline, hydroxychloroquine, and topical agents, which provided only temporary relief (Figure 1a).

Given the persistent facial swelling, Morbihan disease was suspected. The diagnosis was based on a combination of clinical, laboratory, and histopathological findings. A skin biopsy showed mild parakeratosis, and follicular plugging. The dermis exhibited superficial capillary dilatation accompanied by a predominantly lymphocytic infiltrate with occasional histiocytes in a perivascular and perifollicular distribution (Figure 1c). Mild dermal edema with no plasma cell infiltration or dermal fibrosis. Notably, while scattered, ill-formed granulomas were observed, they lacked the well-organized, non-caseating or non-necrotic structure characteristic of sarcoidosis or orofacial granulomatosis. Alcian blue staining showed mucoprotein

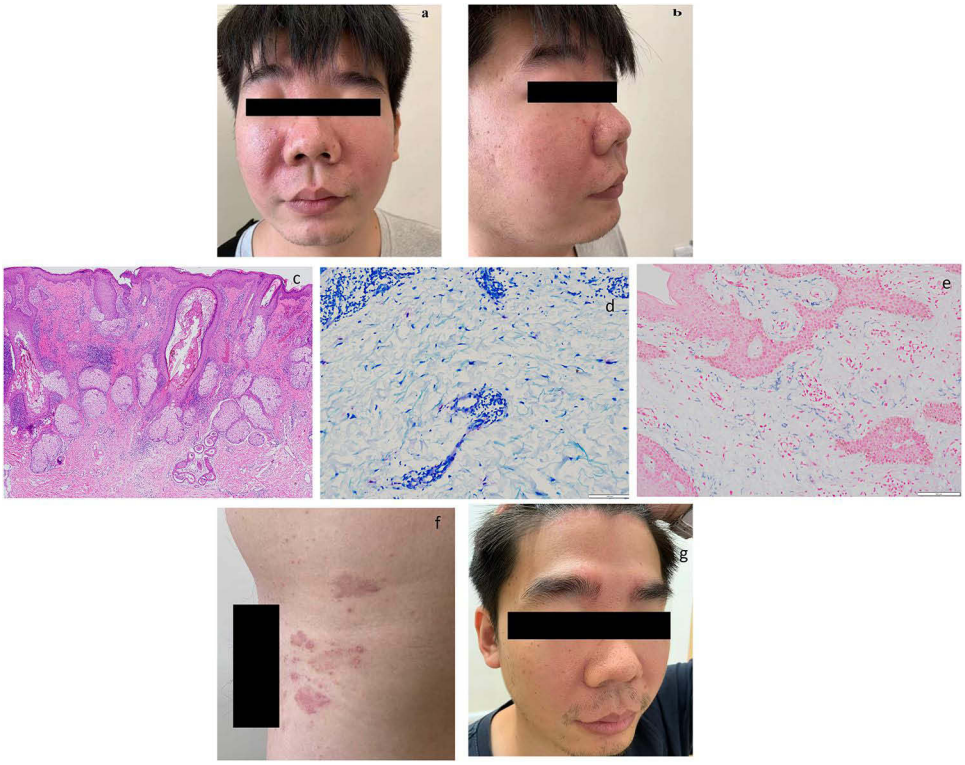

Figure 1 Clinical changes and histopathological finding in the patient. (**a**) Clinical presentation before treatment, showing marked erythema and severe swelling of both cheeks, the nose, the glabella, and the right upper eyelid, accompanied by multiple inflammatory papules and pustules. (**b**) Post-treatment improvement, with regression of papules and pustules and marked reduction of erythematous lesions. (**c**) Histopathology showing mild parakeratosis, follicular plugging dilatation of superficial dermal capillaries, and perivascular and perifollicular infiltration of inflammatory cells, predominantly composed of lymphocytes and histiocytes. Granulomas were observed. Mild dermal edema with no plasma cell infiltration or dermal fibrosis. (**d**) Alcian blue staining showing mucoprotein deposition. (**e**) Giemsa staining showing mast cell infiltration. (**f**) New erythema and papules developed on the proximal thighs. (**g**) Oral isotretinoin for two years achieved marked erythema resolution and stable mild edema.

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deposition (Figure 1d), and Giemsa staining detected mast cell infiltration (Figure 1e). While these pathological features were non-specific, they were consistent with the diagnosis of Morbihan disease in the appropriate clinical context.

Extensive systemic evaluations were performed to exclude differential diagnoses: autoimmune and immunological tests (including antinuclear antibody [ANA] panel) were negative, and the minimal erythema dose (120 J/cm^2) was within normal range. Magnetic resonance imaging (MRI) of the thigh and creatine kinase levels showed no abnormalities, ruling out systemic lupus erythematosus and dermatomyositis. Furthermore, the absence of perioral involvement and facial nerve paralysis, combined with the lack of non-necrotic granulomas on biopsy, ruled out Melkersson-Rosenthal syndrome (MRS) and orofacial granulomatosis (OFG). Systemic sarcoidosis was also excluded based on the absence of typical non-caseating granulomas and normal pulmonary findings on chest imaging.

Consequently, the diagnosis of Morbihan disease was confirmed. Oral doxycycline (100 mg twice daily) administered for five months controlled papulopustular lesions but failed to resolve the edema. Subsequent treatments included topical azelaic acid, omalizumab for five months (partial response), baricitinib for one month (ineffective), and combined intense pulsed light (IPL) with yellow light therapy for three months (partial improvement). A repeated four-month course of doxycycline again produced limited benefit (Figure 1b). Subsequently, new erythema and papules developed on the proximal thighs, along with small papules on the trunk and limbs (Figure 1f).

Due to the lack of sustained response to previous therapies, doxycycline was discontinued, oral isotretinoin (20 mg twice daily) was initiated. Erythema and papules on the proximal thighs, along with small papules on the trunk and upper limbs, gradually faded, the rash flattened, and pruritus subsided. After three months of therapy, the erythema and papules on the proximal thighs and small papules on the trunk and upper limbs completely resolved, followed by marked improvement in facial erythema and a slight reduction in edema after four months. Continued treatment for two years achieved marked erythema resolution and stabilization of mild edema (Figure 1g). The dose was then tapered to 20 mg twice weekly for nine months and 20 mg once weekly for the past three months. On this low-dose maintenance regimen, the patient has remained stable without relapse. Adverse effects were limited to transient uric acid elevation and mild xerosis, both of which were manageable. As the dose was reduced, the adverse reactions gradually resolved.

Methods

Literature Search

This literature review was conducted and reported in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) guidelines. A comprehensive literature search was performed across the PubMed, Web of Science, and Science Direct databases from January 2004 to August 2025. The search strategy utilized the Boolean operator "OR" to combine the following keywords and their combinations: "Morbihan disease", OR "Morbihan syndrome", OR "solid persistent facial edema", OR "lymphedematous rosacea". Only articles published in the English were considered.

Studies were included if they were original case reports or case series detailing patients with a clinical and/or histopathological diagnosis of Morbihan disease. Cases where facial edema was clearly attributable to other known etiologies (eg, systemic diseases, allergic reactions, or insect bites) were excluded. A "favorable outcome" or "improvement" was clinically defined as complete resolution or significant improvement of erythema and edema as assessed by the treating physician's global evaluation or as reported in the original case descriptions. Two authors (FFD and PW) independently screened the titles and abstracts of all retrieved records.

Data Extraction

Demographic data extracted included gender distribution and the median age at diagnosis. Clinical characteristics were systematically recorded, with particular attention to the anatomical sites of swelling and associated manifestations. Key histopathological findings were collected, focusing on the presence of dermal edema, lymphatic or vascular dilation, and the composition of the inflammatory infiltrate. Therapeutic interventions were also catalogued, including the number of patients receiving each treatment and the reported clinical response. Data analysis was performed using SPSS 26.0 software, with

Table 1 Summary of Demographics in Morbihan Disease Patients (n = 80)

Demographics	n (%)/Median (Q1, Q3)
Gender	
Males	59 (73.4%)
Females	21 (26.2%)
Average age at diagnosis (years)	55 (43, 62)
Location	
Upper third of the face	78 (97.5%)
Scrotum	1 (1.25%)
Ear	2 (2.50%)
Upper lip	1 (1.25%)
Chin	1 (1.25%)
Thigh	1 (1.25%)
Trunk and upper limb	1 (1.25%)
Clinical Manifestations	
Swelling of the upper third of the face	78 (97.5%)
Telangiectasia	4 (5.00%)
Scrotal swelling	1 (1.25%)
Upper lip swelling	1 (1.25%)
Ear swelling	2 (2.50%)
Chin edema	1 (1.25%)
Thigh erythema	1 (1.25%)
Trunk and upper limb papule	1 (1.25%)

Abbreviations: Q1, First Quartile; Q3, Third Quartile.

decimal places rounded to three significant figures. Categorical variables were compared using chi-square tests. Odds ratios (ORs) with 95% confidence interval were calculated, and p-values less than 0.05 were considered statistically significant.

Results

A total of 272 records were initially identified. After removal of duplicates, 98 records were excluded. Screening of titles and abstracts resulted in the exclusion of an additional 107 studies. Ultimately, 67 studies comprising 79 patients were included in the analysis (Supplementary Figure 1).

Including the present case, the total cohort consisted of 80 patients. There was a male predominance (59 males, 21 females; ratio 2.81:1), and the median age at diagnosis was 54 years (Table 1). The vast majority (78/80, 97.5%) presented with involvement of the upper and middle thirds of the face, all exhibiting persistent erythema and edema.

Histopathological examination was available in most cases and revealed features associated with therapeutic response (Supplementary Table 1). Lymphatic or vascular dilation (OR = 6.000, 95% CI: 1.568–22.962, P = 0.007) was significantly correlated with a favorable outcome. Conversely, dermal edema (OR = 0.167, 95% CI: 0.044–0.638, P = 0.007) and granuloma (OR = 0.321, 95% CI: 0.112–0.923, P = 0.037) were inversely associated with treatment efficacy, indicating poorer prognosis. These findings suggest that specific histopathological patterns may serve as predictive markers of clinical response. Treatment outcomes were reported for 78 patients (Supplementary Table 2). Of 24 patients treated with isotretinoin, 15 showed improvements. Among 19 patients receiving doxycycline, 13 improved. Systemic corticosteroids were effective in 6 of 16 cases. All four patients treated with omalizumab responded favorably.

Discussion

Morbihan disease is a rare and challenging condition that often presents with persistent erythema and edema of the upper face.[1,2] Diagnosis is difficult due to non-specific histopathological features and frequent overlap with other inflammatory dermatoses,[8,9] Therefore, careful clinical evaluation, exclusion of mimicking conditions, and supportive histopathology are essential for establishing the diagnosis.

To our knowledge, this may be the first reported case of Morbihan disease presenting with nonspecific erythema and papules on the thighs, along with small papules on the trunk and upper limbs, extending beyond the typical facial region. After isotretinoin treatment, the lesions on the inner thighs as well as trunk and upper limbs resolved, and the erythema and papules on the face improved. Histopathological findings included dermal capillary dilation and perivascular lymphocytic infiltration. It is crucial to note that these are non-specific features shared with classic rosacea, rather than definitive diagnostic criteria. While occasional granulomas were identified, the well-formed, non-caseating granulomas typical of sarcoidosis or orofacial granulomatosis were absent. The epidermis was largely preserved with no evidence of plasma cell infiltration or fibrosis. HE staining indicated mild dermal edema. Alcian blue staining showed mucoprotein deposition and Giemsa staining revealed scattered mast cells.

Our pooled analysis suggests that certain histopathological features may have prognostic value. Lymphatic or vascular dilation was associated with more favorable treatment outcomes, whereas dermal edema correlated with poorer responses. However, it is crucial to emphasize that the ORs and prognostic associations derived from aggregated case reports are strictly exploratory. These statistical findings are heavily constrained by retrospective heterogeneity and inherent selection bias, though they highlight the potential of histopathological evaluation in anticipating therapeutic efficacy.

The pathophysiology of Morbihan disease remains complex and multifactorial. Current hypotheses primarily involve: (a) vascular dysfunction compromising wall integrity, leading to fluid extravasation; (b) an end-stage complication of recurrent vasodilation and chronic inflammation associated with rosacea; and (c) lymphatic system impairment, where chronic inflammation drives the damage and remodeling of lymphatic vessels, obstructing normal drainage.

Our review and the present case both underscore the limited efficacy of conventional therapies. The patient described here underwent treatment over a period of five years. Baricitinib proved ineffective. Doxycycline and isotretinoin provided partial and temporary control of inflammatory lesions, consistent with published data showing variable response rates.[5,10,11] Regarding omalizumab, while our pooled literature analysis showed a response in all three previously reported cases (n=3), this finding must be interpreted with extreme caution due to the extremely limited sample size. Physical modalities, such as IPL and yellow light therapy, offered some benefit, though the effect was incomplete.

While isotretinoin is considered a first-line treatment, it reportedly fails in approximately 20% of patients. However, in our case, the lesions including edema and erythema were well-controlled, and the patient has remained on long-term isotretinoin without any exacerbation during dose tapering. While we consider isotretinoin a highly valuable therapeutic option, its absolute superiority cannot be definitively established based on current uncontrolled data. Importantly, facial and periorbital edema remained the most refractory feature, illustrating the central therapeutic challenge in this disorder. These findings suggest that while antibiotics and retinoids may control rosacea-like inflammation, they may be insufficient for the lymphatic and vascular dysfunction underlying edema. The partial response to omalizumab supports an immune-mediated component, yet larger studies are needed to clarify its role.

Ultimately, therapeutic success may require multimodal strategies targeting both inflammatory and lymphatic pathways. Several methodological limitations must be acknowledged. First, the reliance on case reports and small series cannot conclude definitive treatment hierarchy. Additionally, restricting our literature search to English-language texts may introduce language bias. Future prospective studies are warranted to define prognostic histopathological markers and evaluate novel targeted therapies.

Conclusion

Isotretinoin proved effective in this unique case of Morbihan disease with extracutaneous manifestations; however, based on uncontrolled data, these findings are strictly hypothesis-generating and do not establish a definitive treatment hierarchy.

Data Sharing Statement

The original contributions presented in this case. Further inquiries can be directed to the corresponding author.

Ethics Declarations

All methods were carried out comply with the ethical standards of the Sixth Affiliated Hospital of Shenzhen University (Approval Number: LW-2025-023), as well as the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from the participant.

Consent for Publication

Written informed consent for the publication of this case report, including all clinical images and identifiable details, has been obtained from the patient.

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Disclosure

The authors report no conflicts of interest in this work.

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