



Treatment of Morbihan disease

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Morbihan disease (MD) is a rare condition that involves rosaceous lymphedema or erythematous lymphedema of the middle and upper thirds of the face. It typically affects the periorbital region, forehead, glabella, nose, and cheeks. The etiology of MD remains unclear, and its diagnosis is challenging. MD often tends to be unresponsive to therapies commonly used to treat rosacea, including corticosteroids, isotretinoin, and antibiotics. Surgical treatments have therefore been attempted, but most cases showed unsatisfactory responses. These problems could have resulted from an incorrect recognition and interpretation of the pathophysiology of MD and inaccurate planning of the operation, resulting in recurrence or exacerbation of edema.

Abbreviations: MD, Morbihan disease; LVA, lymphaticovenular anastomosis.

Keywords: Edema / Eyelids / Lymphedema

INTRODUCTION

Morbihan disease (MD) is a rare disease, with an unclear place in the nosography. MD was first described in 1957 by the French dermatologist Degos, and is characterized by rosaceous lymphedema or erythematous edema of the middle and upper thirds of the face [1]. It typically affects the periorbital region, glabella, nose, and cheeks. In MD, edema and erythema are not usually associated with pain or pruritis, but they can cause significant changes in the periorbital contour, leading to visual impairment and cosmetic disfigurement. Although MD is generally considered to be a chronic form of rosacea, it may exist in the absence of other features of rosacea, and thus may represent a separate disease process. The difficulty of diagnosing MD is that there are many similar diseases, e.g., myxomas, lympho-

mas, granulomas, and rhinophymas, which causes confusion [2-5]. In addition, there is no standard treatment modality and regimen. In this study, we present an overview of the pathophysiology, diagnosis, and treatment of MD.

CLINICAL FEATURES

MD is characterized by a chronic and recurrent pattern of erythema and symmetrical non-pitting facial edema, mainly in the middle and upper thirds of the face (cheek, nose, glabella, periorbital region, and forehead) with accentuation of the periorbital region (Fig. 1). The onset of MD is usually slow, with intermittent reversible swelling eventually becoming permanent swelling and thickening of the skin. Other features of rosacea may be present, such as telangiectasia, papules, and pustules [6]. This process can lead to distortion of the facial contour and vision impairment due to narrowed visual field, and can cause psychosocial stress.

PATHOPHYSIOLOGY

The pathogenesis of MD remains uncertain. In MD, lymphede-

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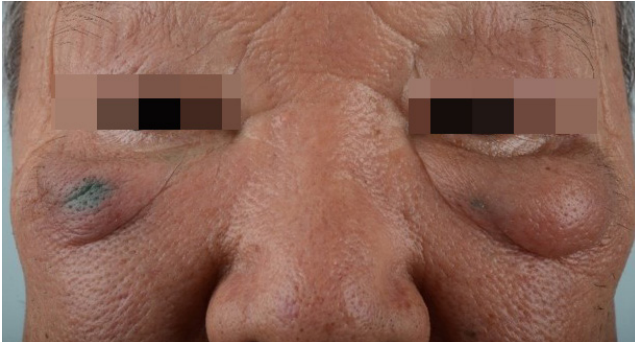


Fig. 1. A 70-year-old Morbihan disease patient. Chronic and non-pitting edema at periorbital region.

ma follows the loss of lymphatic vessel wall integrity and the leakage of intraluminal fluid through the lymphatic vessel wall [7]. The cause is unknown, but possible pathogenic factors have been suggested, such as local dysregulation of lymphatic vessels, lymphatic obstruction by granulomas and histiocytes, and chronic inflammation due to acne, rosacea, or contact dermatitis that destroys supporting connective tissue around dermal lymphatic vessels [8]. Jansen and Plewig [9] reported infiltration of mast cells around the lymphatic collecting vessels, and suspected that mast cells might play an important role in the pathogenesis of MD. However, mast cell infiltration is not a consistent histologic finding in MD. These obscure aspects of the pathophysiology add to the difficulty of diagnosing and treating MD.

DIAGNOSIS

There are no biochemical or histopathological findings specific to MD. Therefore, for the differential diagnosis, biopsy of the affected tissue is recommended. The histopathologic findings of MD include perivascular dermal edema, lymphohistiocytic infiltration, mast cell infiltration to the periadnexal tissues of the lymph vessels, dilation of lymphatic vessels, non-caseating granulomas, and sebaceous gland hyperplasia [10]. The differential diagnosis for MD includes other granulomatous and inflammatory facial conditions [11]. Diseases to consider include sarcoidosis, orofacial granulomatosis, systemic lupus erythematosus, foreign body granuloma, scleroedema of Buschke, lupus vulgaris, and cutaneous pseudolymphoma. Patch tests can exclude allergic contact dermatitis. Functional evaluation of the facial lymphedema using indocyanine green lymphography is helpful for evaluating lymphatic function and planning lymphatic surgery (Fig. 2) [12].

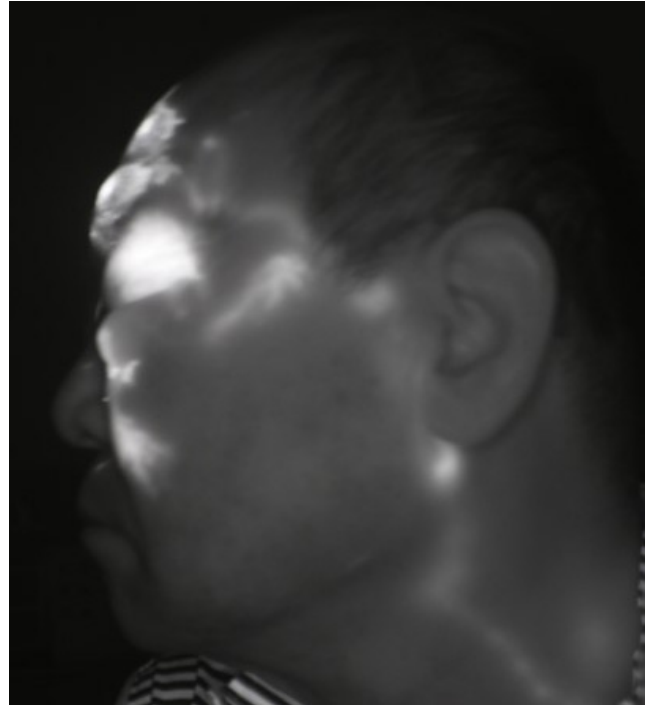


Fig. 2. Indocyanine green lymphography of Morbihan disease patient. A 70-year-old male biopsy proven Morbihan disease patient. Severe dermal backflow noted at periorbital region.

TREATMENT

The treatment of MD is frequently refractory and difficult. There is no definitive treatment modality and regimen.

Several reports have described medical treatments, including systemic corticosteroids, tetracycline, doxycycline, thalidomide, and isotretinoin, but most cases exhibit a chronic refractory waxing-and-waning course and inadequate treatment response [13]. The antibiotics tetracycline, minocycline, and doxycycline are effective for inflammatory rosacea that can be checked with histopathology, which shows extensive mast cell infiltration [14-16]. Shim et al. [17] reported successful treatment with doxycycline and colchicine over 7 months without recurrence during 14 months of follow-up. If there is an inadequate response to medical treatment, it is possible to attempt surgical treatment, such as eyelid reduction surgery, lymphatic drainage surgery, carbon dioxide laser treatment, and local steroid injections [18,19]. Simple excisional surgery is not effective and complete excision is not feasible for aesthetic and functional reasons. Excisional surgery poses a risk of recurrence or even exacerbation of edema. Standard compression therapy for lymphedema is not applicable to MD due to the function of the affected anatomic region. Furthermore, as an important surgical treatment for facial lymphedema after head and neck cancer treatment, lymphaticovenular anastomosis (LVA) is effective

[20]. Similar to extremity lymphedema, LVA is a curative physiological treatment for facial lymphedema in which a new drainage pathway is made by a lymphatic-vein bypass. Based on this evidence, lymphatic drainage surgery is applicable to the treatment of MD. For lymphatic bypass surgery for the treatment of periorbital edema, drainage can be achieved by anastomosis of a subdermal venule with a lymphatic collecting vessel in the preauricular region [12]. In addition to the lymphatic collecting vessels, there are lymph nodes in the preauricular region that can be shunted to venous drainage using a nodal-venous anastomosis to the facial vein. Both LVA and nodal-venous anastomosis can increase the outflow of lymph drainage from the periorbital region. This type of lymphatic drainage surgery can be performed under local anesthesia. Another advantage of these supermicrosurgical procedures is a lower potential for eyelid malposition than in blepharoplasty or excisional surgery. Another physiological option is vascularized lymph node transfer using a lymph node flap to the preauricular region. However, lymph node transfer for the facial region is too invasive to perform under local anesthesia and not acceptable from an aesthetic standpoint.

CONCLUSION

The diagnosis and treatment of MD are challenging. The diagnosis can be made by eliminating possibilities in the differential diagnosis that are similar in terms of clinical and histopathological features. Medical treatment includes antibiotics for inflammatory cases, but mostly yields unsatisfactory results. Surgical treatment options include excisional surgery and functional lymphatic surgery. Considering the psychosocial stress and social/professional impact of MD, efforts must be made to elucidate its pathophysiology in order to develop better treatments in the future.

NOTES

Conflict of interest

No potential conflict of interest relevant to this article was reported.

Patient consent

The patient provided written informed consent for the publication and use of his images.

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