Morbihan Disease – An Old and Rare Entity Still Difficult to Treat

Morbihan disease (MD), also known as Morbihan syndrome, "solid persistent facial edema and erythema", "rosacea lymphedema", and "solid facial edema in acne", is a rare and often unrecognizable entity, that presents with a slow occurrence of persistent lymphoedema of the upper two-thirds of the face (1,2).

A 30-year-old woman presented to our Department with persistent, asymptomatic face edema and erythema lasting for 18 months. She was previously treated for rosacea with doxycycline (100 mg/day for four months) without improvement. Dermatological examination revealed erythematous, nonpitting, solid edema located on the mid-forehead, nose, and cheeks with sparse erythematous papules and pustules on the entire face including the chin and comedones, papules, and pustules on the back (Figure 1 and Figure 2). She was otherwise healthy and was not taking any medication. Laboratory tests with immunological tests and Quantiferon test together with MRI of the orbits, chest X-ray, chest high-resolution computed tomography, cranial X-ray, and abdominal ultrasound were all within normal limits. Histopathology revealed dermal edema, perivascular and peri-adnexal lymphohistiocytic infiltrate, and sebaceous gland hyperplasia. Based on the typical clinical picture, histopathological findings, and the exclusion of several differentials the diagnosis of MD was established. The patient was treated with oral isotretinoin (20 mg/day for eight months) without regression of solid edema and erythema on the face but with complete regression of acne on the trunk. She was started on oral corticosteroids (prednisolone, 20 mg/day for two months followed by reduction of the dose over three months), again with only slight short transient improvement and rapid relapse of facial erythema and edema. The patient refused any other suggested treatment. We treated our patient for a total 2 years and followed up for 5 years.

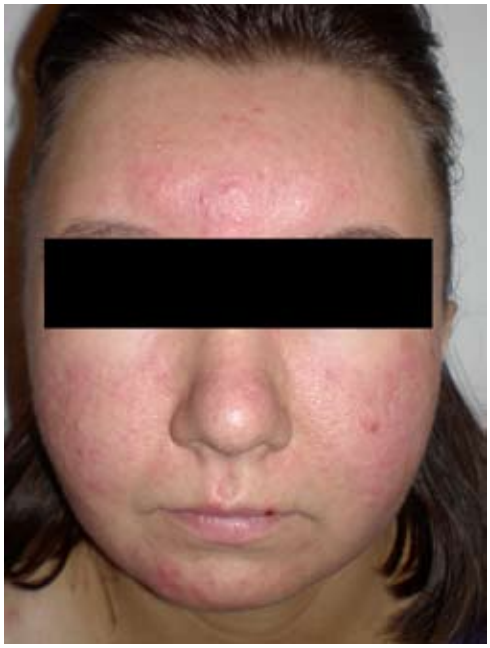

Figure 1. Erythematous, nonpitting, solid edema on the mid-forehead, nose, and cheeks with sparse erythematous papules and pustules.

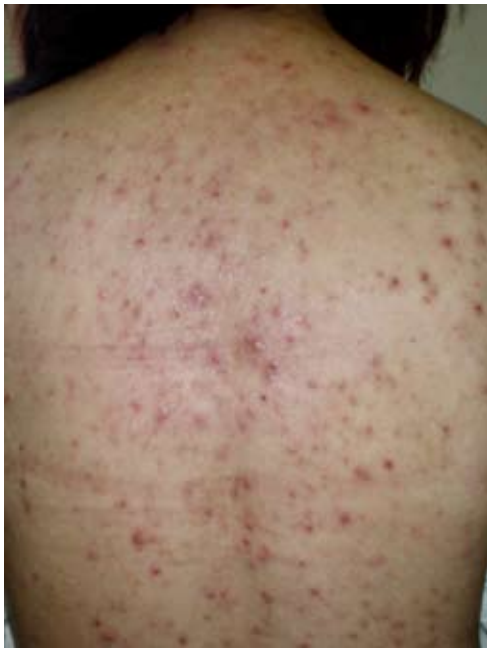

Figure 2. Comedones, papulopustular acne on the back.

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The pathogenesis of MD is still unknown. It is considered a clinical variety or a complication of rosacea or acne which does not tend to regress spontaneously. It is believed that chronic inflammation in patients with MD is due to acne or rosacea causing structural damage to blood and lymph vessels (1,3,4). However, cases of MD without previous history of rosacea and acne have been reported supporting the distinct disease theory (3,4). Edema and erythema are localized on the upper half of the face affecting the forehead, glabella, eyelids, nose, and cheeks. Although the symptoms may come and go, MD usually does not improve without treatment (5). Several therapeutic options have been reported, although there is no established standard treatment for MD. Reported therapy includes short-term oral isotretinoin (0.5 mg/kg/day), long-term oral isotretinoin (40-80 mg/day, 10-24 months), long-term doxycycline, combination of systemic corticosteroids and antibiotic (prednisolone 20 mg/day for 2 weeks and doxycycline 200 mg/day for 12 weeks), slow-releasing doxycycline monohydrate (40 mg/day for 6 months), long-term minocycline (50 mg/day for 4 months), and a combination of both oral retinoid and ketotifen (isotretinoin 0.7 mg/kg/day for 4 months, ketotifen 2 mg/day for 4 months) (1,2,6,8). The disease is frequently recalcitrant to therapy, and only several cases of successfully treated patients with MD have been reported (1,2,4, 6-8).

We presented a patient with characteristic features of MD, which is a persistent, cosmetically disturbing condition, unfortunately mostly refractory to therapeutic measures.

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