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Letters to the Editor

Periorbital lupus profundus successfully treated with oral tacrolimus plus low dose prednisolone

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Dear Editor,

We report a case of lupus profundus presented with periorbital edema successfully treated with oral tacrolimus plus low dose prednisolone.

A 47-year-old woman was referred to our dermatological clinic because of bilateral periorbital edema. The patient had a past medical history of rheumatoid arthritis (RA) for 3 years. She had lupus panniculitis on the right cheek in 1998. Clinical examination revealed erythematous firm indurations on the bilateral periorbital areas (Fig.1a). 4-mm sized light red plaques were observed on her forehead. Sclerodactyly and nail fold bleedings were observed on her fingers. Histological examination of the eyelid showed patchy infiltration of lymphocytes in the subcutaneous tissue (Figure 1b, c). Normal architecture of fat lobules was lost and most adipocytes were replaced by fibrous tissue. Vacuolar degeneration of epidermal basal cells and lymphocytic infiltration around the dermal blood vessels were noted. The histological examination of the forehead lesion revealed epidermal and dermal changes similar to those in the periorbital lesion. Laboratory findings revealed that antinuclear antibodies were positive at 1:640 with a speckled and nucleolar pattern. Anti-ds-DNA antibody and anti-Sm antibody were negative and anti-Scl70 antibody was detected (211 U/ml). The blood counts, C3, C4

were within normal limits. Rheumatoid factor, C-reactive protein were elevated 93.6 IU/ml (normal < 25 IU/ml), 3.04 mg/dl (< 0.3), respectively. Abnormal finding in systemic organs was not detected. We diagnosed her as having periorbital lupus profundus and DLE on the forehead.

The lesions flattened after the initiation of oral prednisolone (30 mg/day). However, her blood pressure increased, and tapering of prednisolone worsened her eyelid edema. Because of the accompanying systemic sclerosis, the increase of prednisolone was hesitated, and oral tacrolimus (3 mg/day) was added. The periorbital lesions improved dramatically and prednisolone was tapered to 10 mg/day (Figure 1d). No serious side-effect of tacrolimus was noted during the course.

To the best of our knowledge, this is the first case of periorbital lupus profundus successfully treated with tacrolimus.

Lupus panniculitis has been treated with corticosteroids, cyclosporine A, dapsone or hydroxychloroquine. Oral corticosteroids and/or hydroxychloroquine are usually effective for eyelid edema diagnosed as DLE, lupus profundus and systemic lupus erythematosus (SLE)¹⁻³. Tacrolimus is well known as an effective medication for RA⁴. We chose oral tacrolimus because her activity of RA was high, and high-dose prednisolone and cyclosporine A were hesitated because of complications of high blood

pressure.

Eyelid edema is not common in SLE in general. Its incidence as the initial manifestation of SLE is 0.1%, and its overall incidence is 4.8% ⁴⁾. Serarslan et al¹⁾ reported periorbital edema and erythema as symptoms of DLE. We speculate that periorbital panniculitis might have been underrecognized in the past, because of the difficulty to take deep skin biopsy from these areas and the epidermal and dermal changes in lupus profundus can be interpreted as DLE.

Taken together, our report suggests that periorbital edema may occur as a manifestation of lupus profundus, and oral tacrolimus might be used as a second-line treatment.

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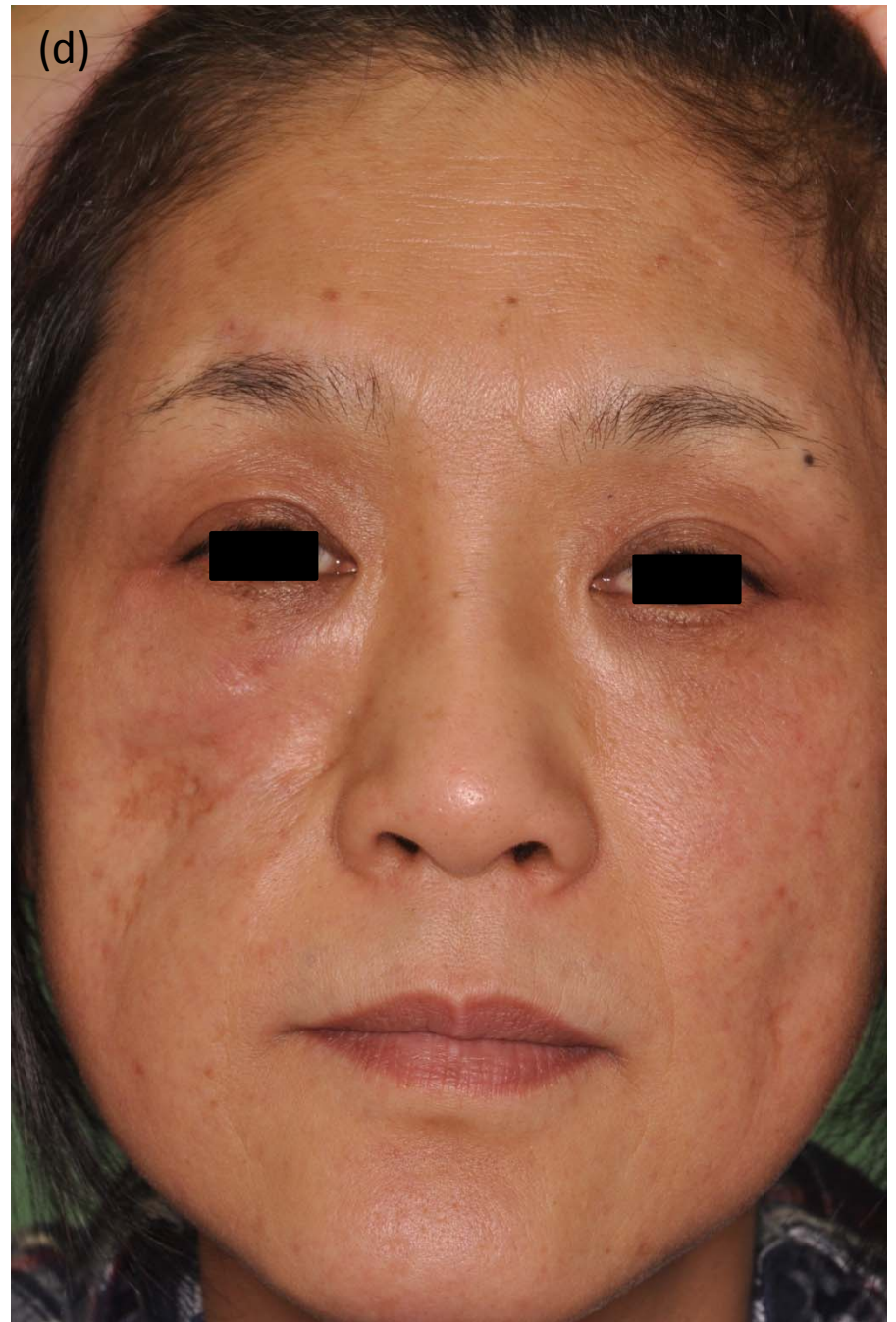
Figure legends

Figure 1(a): Edema and erythema with firm indurations on the bilateral periorbital areas extending to the malar areas.

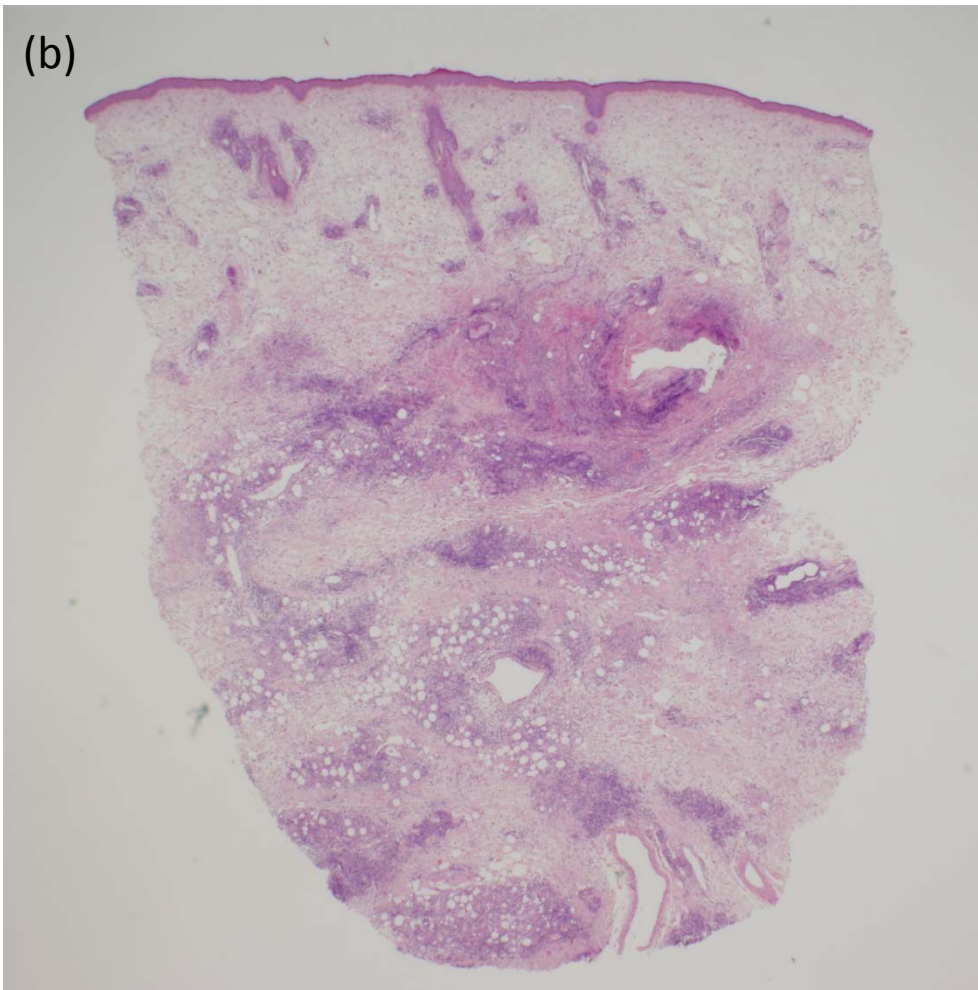
Figure 1(b): Histology of the right lower eyelid showed marked panniculitis.

Figure 1(c): The heavy lymphocytic infiltration in the subcutaneous tissue.

Figure 1(d): Lesions on the periorbital areas were improved dramatically with oral tacrolimus (3mg/day).



(b)



(c)

