



INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

MELKERSSON-ROSENTHAL SYNDROME: A RETROSPECTIVE STUDY OF 35 PATIENTS SHOWING TREATMENT OUTCOMES, AND INDICATING LINGUA PLICATA RATHER THAN OROFACIAL SWELLING AS THE INITIAL SIGN

E Ozkaya⁽¹⁾ - S Kiliç⁽²⁾ - T Ö Akbulut⁽³⁾ - N Büyükbabani⁽⁴⁾

Istanbul University, Istanbul Faculty Medicine, Department Of Dermatology And Venereology, Istanbul, Turkey⁽¹⁾ - Istanbul University, Istanbul Faculty Of Medicine, Department Of Dermatology And Venereology, Istanbul, Turkey⁽²⁾ - University Of Health Sciences, Haseki Research And Training Hospital, Department Of Dermatology And Venereology, Istanbul, Turkey⁽³⁾ - Istanbul University, Istanbul Faculty Of Medicine, Department Of Pathology, Istanbul, Turkey⁽⁴⁾

Introduction: Melkersson-Rosenthal syndrome (MRS) is characterized by the triad of recurrent facial paralysis (FP), lingua plicata, and orofacial swelling; the latter as the most commonly reported initial sign. A standard management protocol is lacking.

Objective: To assess the clinical features and treatment outcomes in patients with MRS

Material and Methods: A retrospective, cross-sectional study on 35 patients diagnosed with MRS in our clinic between 2002-2018

Results: The male/female ratio was 1:4.8. The mean age at onset of the first symptomatic sign was 27.1 years. The referral diagnosis was episodic/persistent angioedema in all patients. The classical triad was present in 18 patients (51.4%). 31 patients had lingua plicata as the initial but asymptomatic sign, and 22 had a history of 1-10 attacks of FP. Histopathology showed granulomatous inflammation in 22 patients. First-line therapy was started with systemic corticosteroid (SC) (n=26), or intralesional corticosteroid (IC) (n=1). Second- or third-line therapy options were IC (n=5), systemic dapsone 50 mg/day (n=3), or clofazimine 100 mg/day (n=6). Six patients showed complete remission of their orofacial edema (4 with SC, 1 with dapsone, 1 without treatment). They had no relapse for a mean period of 3.4 years. 21 patients had partial remission, and three did not respond to any therapy.

Conclusion: The percent of patients presenting with a complete triad of MRS was much greater than in previous reports (8-25%). Lingua plicata seems to be the initial but often





overlooked sign of MRS. The coexistence of lingua plicata and history of FP permits a cost-effective approach to orofacial edema, preventing patients from unnecessary investigations with regard to systemic granulomatous conditions such as Crohn's disease or sarcoidosis. The spontaneous regression rate of orofacial edema is low. SC/IC seems to be the appropriate first-line treatment. Dapsone may be effective in selected patients, but clofazimine seems not promising.

