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Merkelsson-Rosenthal Syndrome: Is Psoriasis a New Component of the Syndrome?

Merkelsson-Rosenthal Sendromu: Psöriazis, Sendromun Yeni Bir Komponenti mi?

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Dear Editor,

Psoriasis is a relatively common erythematous-scaly skin disease with unknown etiology. Scrotal tongue, also known as fissured tongue, and geographic tongue prevalence of psoriasis patients is considered to be higher than the healthy population (1). Scrotal tongue and psoriasis were found to co-exist with a rate of 22.6% to 47.5% in different case series (2). While psoriasis and geographic tongue may co-exist on a genetic predisposition, researchers have failed to prove the existence of a genetic association between scrotal tongue and psoriasis (1).

Merkelsson-Rosenthal syndrome (MRS) is a rare clinical picture with unknown etiopathogenesis. MRS is characterised by the triad of recurrent orofacial edema, recurrent facial paralysis, and scrotal tongue. However, finding all three symptoms in the same patient is very uncommon and this triad is only seen in the 25% of these patients while the rest of the patients lack one or two of the symptoms (3). In case of orofacial edema, granulomatous inflammation non-caseified is histologically very common though histopathological findings are not diagnostic and the absence of granulomatous inflammation does not exclude the diagnosis of MRS. In such cases, diagnosis is based on clinical findings.

The etiology of MRS is not exactly known but, so far researchers have blamed contact hypersensitivity, infectious, and genetic factors.

A 21 years old male patient who had been followed for psoriasis for approximately 7 months was admitted with edema in the upper lip. The patient described the edema as swelling of the lip with intervals of a couple of months and lasting for about 20 days. During this period, the patient did not have any accompanying skin lesions and there was no decline in the edema. Having

detected scrotal tongue on oral mucosa examination, we diagnosed the patient with oligosymptomatic Merkelsson-Rosenthal syndrome. Our patient had no history of any facial paralysis. His family history was unremarkable as far as MRS was concerned. The patient was admitted with localised typical erythematous scaly psoriatic lesions on the extensor surface of the extremities approximately 7 months ago and received clinical diagnosis of psoriasis for which the patient was then given topical treatment and asked to attend the follow-up examinations. The psoriatic lesions were under control due to an ointment containing combined topical calcipotriene and betamethasone. The last follow-up examination showed no active psoriatic lesions. We offered a treatment of 32 mg/day of methylprednisolone (for 3 days) and antihistamines (5mg/day levocetirizine) for the lip edema. The lip edema distinctly decreased within 3 weeks.

There are only a few cases in the literature with MRS and psoriasis at the same time. Halevy et al. have detected psoriasis in 5, palmar psoriasiform lesion in 1, and psoriatic arthritis in 1 of the 12 MRS patients. Of these patients, only 42% presented with all of the triad of findings. Histologically, granulomatous inflammation was present in 23% of these cases (4). Gaudio et al. have identified MRS in a psoriatic arthritis patient receiving etanercept treatment (5). The presence of scrotal tongue, a component of MRS, in association with psoriasis is well defined in the literature (1, 2). Halevy et al.'s study have identified psoriatic picture in 7 of the 12 MRS patients (58.3%) which strongly supports a possible association between MRS and psoriasis (4). To our knowledge, our case is the first reported case of MRS with psoriasis in Turkey.

In the light of the literature, we believe psoriasis may be a newly defined component of MRS. It is clear that there is a need for prospective studies with large case series for a better understanding of the potential relationship between MRS and psoriasis on an etiopathogenetic basis.

With respects.

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