Reply to: "Hidradenitis suppurativa and Mediterranean fever gene mutations"

To the Editor: We appreciate the opportunity to reply to the letter by Ilgen et al regarding our case report, Potential role of serum amyloid A in hidradenitis suppurativa.[1]

Unfortunately, we did not consider the heterozygous Mediterranean fever gene (*MEFV*) mutations without familial Mediterranean fever (FMF) in our patient because this condition is not a prevalent disease in the white population.[2]

In the cases analyzed in our review, *MEFV* mutations without FMF in hidradenitis suppurativa (HS) with AA amyloidosis were investigated only by Ilgen et al.[3] Moreover, in the literature, we found a recently published case control study by Vural et al[4] that investigated heterozygous *MEFV* mutations in a cohort of 21 Turkish patients with severe HS without prior diagnosis of FMF. Among these patients, 2 had severe HS without any additional inflammatory disorders (acne, pyoderma gangrenosum, and arthritis). One was negative and the other one was positive for R202Q polymorphism, common in the population and not considered pathogenic.[4] Overall 38.1% of these patients with additional inflammatory diseases had heterozygous *MEFV* mutations. In consideration of prognostic importance of *MEFV* mutations in many inflammatory diseases other than HS, this topic needs to be further investigated.[5]

HS is certainly a disease with autoinflammatory features.[6] In 2017 we also adopted the Autoinflammatory Disease Damage Index in a cohort of 47 cases of severe Hurley III HS concluding that it "may be a promising tool to evaluate the long-term systemic outcome in HS."[7]

Other HS severity markers were investigated by a German team. In the retrospective study, they described a significant correlation between C-reactive protein levels and neutrophil count with disease severity according to the modified Hidradenitis Suppurativa Score.[8]

Further research is needed to clarify the pathogenesis, clinical phenotypes, and prognostic factors in HS to develop a tailored-based effective approach. In our opinion, the most important markers to evaluate in large cohorts of patients with severe HS could be Autoinflammatory Disease Damage Index items, C-reactive protein, neutrophil count, serum amyloid A, and heterozygous *MEFV* mutations.

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