PSORIASIS

CHARACTERISTICS OF GENERALIZED PUSTULAR PSORIASIS IN THE SOUTH OF TUNISIA

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Introduction: Generalised pustular psoriasis (GPP) is a rare and severe form of psoriasis, with various clinical, epidemiological and genetic profiles.

Objectives: To review epidemiological,clinical and genetic features of GPP in Tunisia.

Methods :We conducted a retrospective studybetween 2012 and2018. The diagnosis of GPP was based on clinical and histological data. We performed a molecular analysis to identify variations on IL36RN gene.

Results: Over 7 years, we collected 17 new cases of GPP (2.4cases/year). The patients 'age ranged from 6 to 60 years with a sex ratio (M/F) of 1.12. Two familial cases were identified. The age of onset ranged from 1 to50 years with a mean age of 27 years. GPP was preceded by psoriasis vulgaris in 23.5%. Four cases (23.5%) begun as pustular psoriasis of pregnancy. Body surface affected varies between 20 and 100%.
Extracutaneous signs were seen in 10 cases (58.8%): articular signs(3 cases), liver test abnormalities (2 cases), conjunctivitis in one case and elevated inflammatory markers in 10 cases(58.8%).Molecular analyses were performed in 11 cases. In these patients, homozygous missense mutation (p.L27P) in IL36RN gene was identified in 3 patients originated from the same geographic area in the south of Tunisia. Heterozygous mutation was detected in 1 patient.In 64% of cases, no mutation in IL36RN gene was found. First line treatment was acitretin (0.5 to 1mg/kg/d) (58.8%). Local treatment with topical corticosteroids was recommended in 35.3% of cases. Methotrexate was used in 2 cases complicated by hematotoxicity in one case, so relayed by Infliximab. The course of the disease was usually good. However, relapses were often noted (35.3%) when treatment was stopped(17.6%) or when infection occurs (17.6%). One patient died because of renal failure.

Conclusions: We present a large case series of GPP.Our study showed phenotypic as well as genetic heterogeneity in our study population.