Important Results Derived from our 60 Year Experience on Familial Mediterranean Fever and Behçet Disease

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Abstract: FMF: is the first described and prototype of the auto inflammatory diseases. Important conclusions derived from our original cohort of 544 patients are: FMF is more prevalent in Turkey than in many other countries, juvenile onset and positive family history are common. Peripheral arthritis is one of the cardinal manifestations. Sacroiliitis and SpA are frequently associated, but HLA-B27 positivity is not significant. AA amyloidosis is quite high (11%), the 2 cases are associated with Behcet disease. Colchicine is drug of choice including amyloidosis. We published our diagnostic criteria for FMF in 1997. BD: first described by Turkish dermatologist Prof. Hulusi Behçet in 1937. Main conclusions of our original cohort of 624 cases are: BD: also is more prevalent in Turkey, more common in men (M:F 1.67), juvenile onset and familial occurrence are not rare. Non-oral aphthous beginning is observed 1/3 of the cases. Thrombophlebitis, pulmonary involvement and peripheral arthritis are quite frequent. Some cases fulfill the SpA criteria. We observed 9 cases of AA amyloidosis. Skin pathergy is most specific feature of BD. Vital organ involvement and some demographic factors play an important role on poor prognosis. We described our revised diagnostic criteria in Seoul 2000. We certainly accept the title of “Behçet Disease” only!

Key words: FMF, BD, Turkey