Efficacy of thalidomide in refractory adult Still’s disease: A new case report

Sirs,

Adult Still’s disease is a febrile disorder of unknown etiology which affects young adults during the second or third decade, presenting either as a primary disease or as a relapse of childhood onset Still’s disease (1). Clinical features are characterized by an initial pharyngitis with prolonged fever, polyarthralgia and maculopapular rash; liver and pericardium involvement are commonly observed (2). The biological data comprise a major increase in all inflammatory proteins and leukocytosis usually >20,000 cells/mm³. The poor prognosis of Still’s disease is determined by the long time course of its evolution, the frequent relapses, and serious complications [thrombotic thrombocytopenic purpura (3), amyloidosis (4)]. Levels of tumor necrosis factor α (TNFα), like the levels of other inflammatory markers (C reactive protein, ferritine), increase of Still’s disease and therefore some.

Corticotherapy (prednisone) was started at the age of 150 mg/day. One month later the disease was in total remission, with a CRP level at 2 mg/L despite a reduction in the daily prednisone dosage to 30 mg/day. Nine months after the beginning of thalidomide treatment, remission was maintained with a CRP level at 9 mg/L, and a prednisone dosage of 15 mg/day. Throughout the follow-up the normal levels of other inflammatory markers and anti-angiogenesis properties of thalidomide has increased interest in this molecule, especially in the field of inflammatory and immune-mediated diseases. Thalidomide is able to inhibit TNFα production in human monocytes by stimulating mRNA degradation in a dose-dependent fashion and to reduce the half-life of TNFα mRNA by 50% (11). Stanbe et al. (12) reported the efficacy of thalidomide in a 44-year-old woman with adult onset Still’s disease refractory to other treatments. The patient was therefore given thalidomide (200 mg/day, then 100 mg/day). Improvement was observed after 2 weeks of treatment (thalidomide + prednisone) and the disease was in remission during a 6-month follow-up period. During the study, the percentage of monocytes producing TNFα decreased from 42.7% before thalidomide to 1.4% during the treatment.

Our results confirm the efficacy of thalidomide in adult Still’s disease, even at a dosage of 150 mg/day. This efficacy is particularly remarkable when one considers the cost-effectiveness of this drug in comparison to other anti-TNFα drugs. Moreover, no significant side effects were observed. In conclusion, thalidomide seems to offer a new approach for the treatment inflammatory diseases linked to the overproduction of TNFα. Refractory adult Still’s disease might be a new indication for thalidomide as long as careful monitoring of peripheral nerve conduction with electromyograms every 6 months is conducted to detect asymptomatic neuropathy. In fertile women the use of thalidomide should be authorized only in women taking the birth control pill.

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References