Interleukin-1 receptor antagonist (anakinra) for Schnitzler syndrome

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Schnitzler syndrome is a rare autoinflammatory disease, which is defined by the presence of two major criteria: chronic urticaria and monoclonal immunoglobulin M (IgM) or immunoglobulin G gammopathy, in combination with at least two additional minor criteria: recurrent fever, leukocytosis and/or elevated C-reactive protein (CRP), objective signs of abnormal bone remodelling and a neutrophilic infiltrate in skin biopsy. We report on a 68-year-old female patient with a 10-year medical history of chronic urticaria, recurrent fever, severe arthralgia and increased CRP. Over the years, multiple diagnostic investigations were performed without conclusive findings, and therapeutic attempts with anti-histamines and several immunosuppressive agents had failed. The decision to initiate monotherapy with interleukin-1 (IL-1) receptor antagonist was based on immunohistochemical detection of the abundance of IL-1β positive cells in the patient's skin biopsy. After starting treatment with anakinra, disappearance of symptoms could be observed within 24 h. Discontinuation of the treatment resulted in a rapid relapse of the symptoms. Finally, already after the initiation of therapy with anakinra, the suspected diagnosis of Schnitzler syndrome could be confirmed by detection of IgM-gammopathy that was initially absent.