

Abstract

This is the first reported case of Schnitzler syndrome in South Asia. Schnitzler's syndrome is an acquired rare autoinflammatory disease characterized by a monoclonal gammopathy, chronic urticarial rash, and periodic fever with systemic inflammation. Around 300 cases were reported in about 25 countries and mostly in Caucasian descent. No cases have been reported in South Asia. We report a case of 64-year-old Sri Lankan woman presented with a 4-year history of chronic urticaria, periodic fever associated with constitutional symptoms with IgM monoclonal gammopathy. Diagnosis of Schnitzler's syndrome made based on Strasbourg diagnostic criteria. The patient was started on prednisolone and colchicine to which she has responded well.