This patient with swollen, deformed hand joints and an elevated erythrocyte sedimentation rate likely has untreated inflammatory polyarthritis. The presence of concurrent neutropenia and splenomegaly raise strong suspicion for Felty syndrome, an uncommon but serious complication of long-standing, erosive rheumatoid arthritis (RA).

Felty syndrome is marked by the formation of autoantibodies against neutrophil components and granulocyte colony-stimulating factor, leading to neutropenia (ie, absolute neutrophil count <2,000/mm3) and an increased risk of recurrent bacterial infection (particularly of the skin and sinuses). Neutrophils coated with antibodies are also trapped in the spleen, which usually results in splenomegaly. Most patients also have extraarticular manifestations of RA such as lymphadenopathy, rheumatoid nodules, and/or necrotizing skin lesions.

The diagnosis is made based on clinical features but is supported by the presence of high-titer rheumatoid factor and anticitrullinated peptide antibodies (both of which are usually elevated in RA). Most patients are also HLA-DR4 positive (indicating a genetic susceptibility). Other causes of neutropenia should be ruled out with bone marrow biopsy and peripheral smear prior to establishing the diagnosis. Symptoms generally improve with treatment of the underlying RA.