ANCA positive polyarthritis revealing Whipple’s disease

Sirs, Whipple’s disease is a systemic disorder caused by *Tropheryma whipplei*, a telluric Gram positive bacterium. It is a rare disease with polymorphic clinical presentations. Diagnosis is often delayed and is suspected only after the appearance of digestive signs. We report here a case of ANCA positive polyarthritis revealing Whipple’s disease with systemic manifestations without digestive signs.

A 40-year-old Caucasian woman had a six-year history of peripheral chronic arthritis (metacarpophalangeal and proximal interphalangeal joints, wrists and elbows), cervical pain, myalgia, asthenia and night sweating. Laboratory findings revealed chronic inflammation: C-reactive protein level (116 mg/l) and increased erythrocyte sedimentation rate (60mm). Autoantibodies including anti-nuclear antibody, anti-DNAn, rheumatoid factor and anti-cyclic citrullinated peptide were negative, except the presence of anti-neutrophil cytoplasm antibodies (ANCA) at 1/1600, with cytoplasmic fluorescence without a specific pattern. Articular x-rays and bone scintigraphy were normal. The patient had intermittent symmetrical polyarthritis revealing Whipple’s disease. Treatment with doxycycline (200mg daily) + hydroxychloroquine (600mg daily) + methotrexate (15mg per week) was initiated, followed by a dramatic improvement of arthralgy, decreased the sweating and pleuresia within four days. The following treatment includes doxycycline (200mg daily) + hydroxychloroquine (600mg daily) for a total duration of 18 months.

The clinical diagnosis of Whipple disease is difficult. Classically, it appears with digestive symptoms associated with chronic diarrhea, abdominal pain and weight loss, migratory arthralgia and arthritis are present in 65% and 90% of cases, respectively. This disease predominantly affects men (87% of cases) at around fifty years of age (1-3). In spite of the multisiversal involvement that provides evidence of the evolution of the disease (pleuresia, ascites, polyadenopathy, melanoderma), digestive manifestations are absent, which highlights the originality of our case. Furthermore, the presence of ANCA is unusual. Diagnostic discussions lean especially towards vasculitis or connectivites (systemic lupus, rheumatoid polyarthritis), sarcoidosis and paraneoplastic syndromes.

In the case of suspected Whipple’s disease, a small bowel or duodenum biopsy must be performed in search of the PAS+ inclusions within the macrophages of the lamina propria. PCR assays targeting the 16S rRNA gene and 16S-23 intergenic regions of *T. whipplei* confirm the diagnosis (4). A search for the same elements on various tissues and body fluids according to the clinical presentation must be carried out (synovial fluid or tissue, adenopathy, cerebrospinal fluid, aqueous humor in the case of uveitis, cardiac valves).

Whipple’s disease is a rare disorder and the absence of gastrointestinal symptoms is an atypical clinical presentation. It must be considered when non-erosive polyarthritis associated with systemic symptoms simulating an autoimmunity disorder does not improve with usual immunosuppressive therapy.

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References