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COMMON VARIABLE IMMUNODEFICIENCY MIMETIZING CASE OF SPONDILOARTROPATHY – CASE REPORT

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BACKGROUND

The Common Variable Immunodeficiency (CVI) is a set of diseases related to reduced serum levels of immunoglobulin, mainly, IgG4, with reduced antibody production, resulting in numerous symptoms, with: recurrent bacterial infections, pneumopathies, hepatosplenomegaly, lymphadenopathies, polyarthropathies and espondylarthropathies. Due to a large number of differential diagnoses, it is important to suspect primary immunodeficiency in infections with atypical behavior.

CASE REPORT

Male patient, 18 years old, one year ago presented complaints of neck pain, low back pain, pain in wrists, knees and ankles. In physical examination, presented pain on the palpation of sacroiliac and Achilles tendons, in addition to wrist arthritis. Lasegue and Patrick´s test were negatives, and Schober 10-14 cm. MRI revealed discreet bilateral sacroiliits, interstitial ligament edema from L3 to S1 and some vertebral erosions at the thoracic level, indicating possible spondyloarthropathy. In the laboratory tests: HLAB27, anti-CCP and rheumatoid factor negatives, antinuclear factor non-reactive. It was prescribed methotrexate e NSAIDs. In the evolution, persisted with arthritis, inflammatory low back pain and increased ESR and CRP, choosing to initiate immunobiological therapy with adalimumab. Four months later, presented pain in the hypochondrium and right iliac fossa, with CT of abdomen and cholangio-resonance demonstrating homogenous hepatoesplenomegaly, with normal hepatic markers. Has manifested pharyngitis with feverish spikes, recurrent respiratory infections requiring hospitalization, persisting with joint and lumbar pains. Due to non-clinical response and repetitive infectious pictures, the biological was suspends and further investigation was performed. New exams showed: ESR rate with CRP normal, anti-muscle smooth, anti-mitochondria, anti-LKM1 and ANCA non-reagents, protein electrophoresis without monoclonal peak. Performed immunoglobulin screening and immunological profile of the patient (IgG3, IgG4, IgGA, IgGA, IgM, IgD, CD4, CD8 and anti-pneumococcal antibody), was observed a significant deficiency of IgG4, with anti-pneumococcus, CD4 and CD8 low, defining a dysfunction of the autoimmune system, compatible CVI with predominance of IgG4. With the diagnosis, treatment with immunoglobulin was instituted every 28 days associated with prophylaxis with sulfamethoxazole/trimethoprim and amoxicillin. Currently, patient responds well to treatment and will perform the fifth dose of medication.

CONCLUSION

Because it represents a heterogeneous set of clinical manifestations, CVI with predominance of IgG4 generates difficulties and delayed diagnosis, providing a delayed treatment and worse prognosis. The case presented shows a young man with symptoms compatible with spondyloarthropathy, even using biological therapy. Due to the lack of clinical response and repetitive infections, an immunological profile was performed, closing the diagnosis of the disease.