



RHEUMATOID NODULES, A RARE PRESENTATION IN MIXED CONNECTIVE TISSUE DISEASE: A CASE REPORT

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BACKGROUND

Mixed Connective Tissue Disease (MCTD) is a rare systemic autoimmune disease characterized by signs and symptoms of overlapping of at least two connective tissue diseases including systemic lupus erythematosus (SLE), systemic sclerosis, polymyositis, dermatomyositis and rheumatoid arthritis, along with the presence of a distinct antibody, anti-ribonucleoprotein U1 (RNP). Most authors describe MCTD as an independent entity, while some believe that it may represent an early stage of a definitive connective tissue disease, for example SLE, or overlap syndrome. The presence of rheumatoid nodules is associated with more severe disease phenotypes.

CASE REPORT

A 32-year-old male patient presented polyarthralgias on metacarpophalangeal, wrists, shoulders, elbows, heels, associated with mild local edema, with morning stiffness > 1 hour. In subsequent months, the following findings were added: Raynaud's phenomenon (RP), diffuse hand edema, muscle weakness and dysphagia. The investigations were carried out with complementary tests that confirmed the diagnosis of MCTD: anti-RNP reagent, elevated muscle enzymes and inflammatory tests, positive ANA and rheumatoid factor, delayed esophageal emptying (by esophageal transit scintigraphy), abnormal periungual capillaroscopy (microangiopathy with SD-pattern). Initiated methotrexate (MTX), nifedipine and domperidone, with improvement of muscle weakness and RP, however, as a result of gastrointestinal adverse event, MTX was replaced by azathioprine (AZA), achieving disease remission. Four years after diagnosis, subcentimetric nodulations appeared on the palmar face of some distal phalanges. The biopsy of the lesions was compatible with rheumatoid nodules (fibroconnective tissue with fibroblast proliferation, foci of fibrinoid necrosis and histiocytic palisade).

CONCLUSION

Despite the positivity of the rheumatoid factor and the presence of joint complaints, we can not diagnose the patient as having a overlap syndrome, in view of the fact that synovitis itself had not been demonstrated (neither clinically nor ultrasound or by MRI of the hands). The relevance of the case is due to the atypical evolution of the patient with the presence of rheumatoid nodules, common extra-articular manifestation of rheumatoid arthritis but rare in MCTD. In addition, the nodules usually arise on the extensor surfaces of the joints and at points of pressure of the extremities, unlike the case in question, in which the nodules affected the palmar face of distal phalanges.