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Letter to the Editor

Rheumatoid arthritis revealed by polyadenopathy, diarrhea and digestive AA amyloidosis

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A 78-year-old woman was addressed for fatigue, anorexia, diarrhea, pitting legs "edema" (Fig. 1A) and a 5-kilogram weight loss in the past 18 months. Her medical history consisted of arterial hypertension, and total knee replacement for osteoarthritis at the age of 65. Blood explorations revealed aregenerative anemia (Hb = 94 g/L, mean corpuscular volume 92 μm^3) and an elevated C-reactive protein (59 mg/L). Liver function was normal (ASAT = 24 U/L; ALAT = 25 U/L, prothrombin time > 70%) as well as renal function (creatininemia 0.8 mg/dL), albuminemia (35 g/L), urinary sediment and proteinuria/creatininuria (< 0.02 g/mmol). Echocardiography revealed no heart dysfunction, troponinemia and blood NT-proBNP were normal. A chest abdomen and pelvis CT scan were performed showing several profound and peripheral supracentimetric lymph nodes (Fig. 1B). Only one axillar adenopathy (2 cm diameter) appeared hypermetabolic on PET-CT scan (SUV = 6.2) and was biopsied showing non-specific signs including follicular hyperplasia and polyclonal plasmocytosis (Fig. 1C). Digestive tract endoscopies were macroscopically normal,

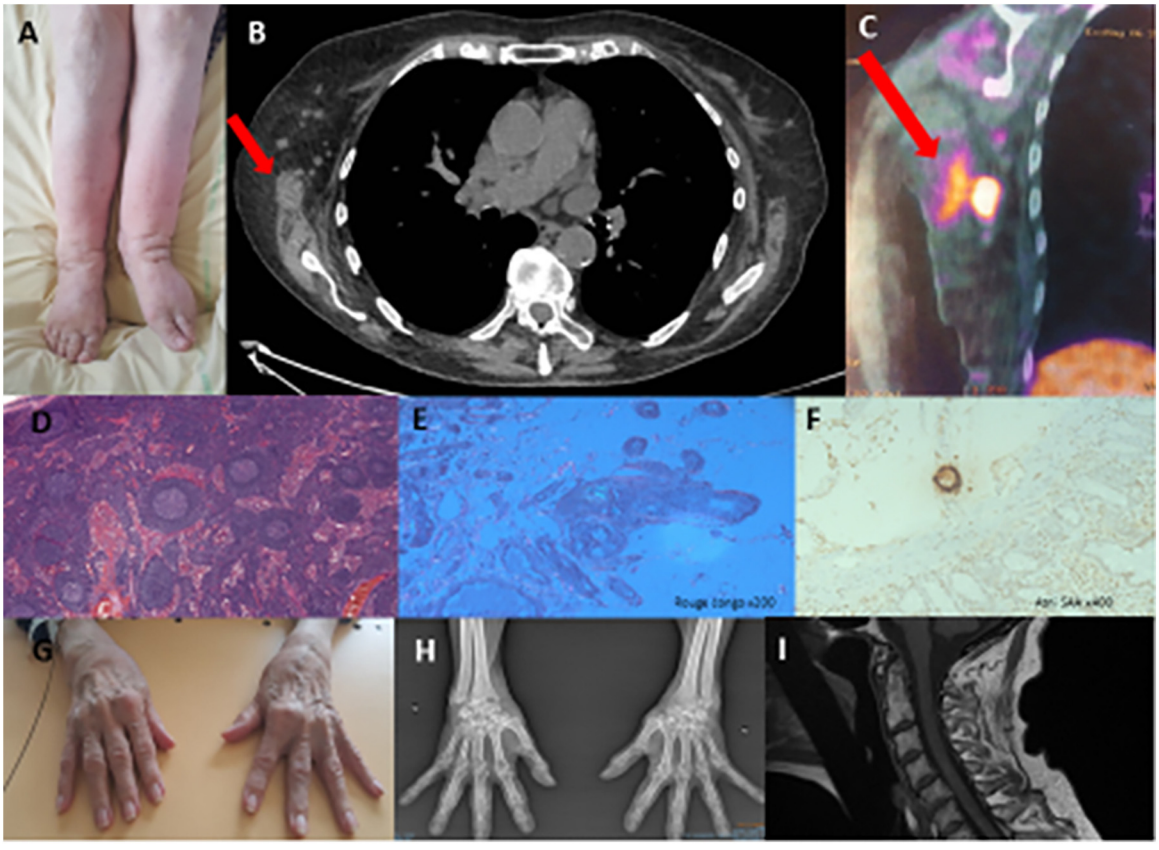

Fig. 1. Main clinical, radiological and pathological features of the patient. Panel A: legs edema first observed; panel B: axillar adenopathy on chest CT scan; panel C: biopsy of adenopathy showing follicular hyperplasia and plasmocytosis; panel D: Congo red staining of duodenal biopsies showing amyloid deposits; panel E: immunohistological staining of duodenal biopsies showing serum amyloid A protein deposits; panel F: aspect of hands and wrists; panel G: X-ray radiographs of hands and wrists showing bilateral severe carpitis and involvement of all metacarpophalangeal and proximal interphalangeal joints.

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duodenal biopsies showed vascular Congo red positive deposits (Fig. 1D). Therefore, the patient was referred to our center for amyloidosis typing. AL amyloidosis was suspected because of her age and the presence of polyadenopathy on CT scan, but no monoclonal gammopathy was detected, free light chains were normal in blood and urine. Finally, the immunohistological typing of the duodenal deposits showed AA amyloidosis (Fig. 1E). Bone marrow biopsy was normal. Clinically, we were struck by her wrists (Fig. 1F) which were seemingly swollen and we questioned the patient in order to look for classical AA amyloidosis causes [1,2]. She reported chronic daily wrists and cervical pains that she treated by self-medication with daily paracetamol. The wrists and hands X-ray showed bilateral severe carpitis and involvement of all metacarpophalangeal and proximal interphalangeal joints (Fig. 1G). Cervical MRI showed odontoid synovitis. The immunological blood tests showed a very high rate of anti-CCP antibodies (> 250 U/mL) as well as rheumatoid factor (320 U/mL). Rheumatoid arthritis (RA) was eventually diagnosed whereas the legs edema was attributed to lymphedema. Low dose corticosteroid therapy (10 mg prednisone per day) was started in association with methotrexate (15 mg per week) for RA and interleukin-6 receptor inhibitor tocilizumab for AA amyloidosis [3–5]. That resulted in a dramatic improvement of her clinical condition, the inflammatory syndrome resolved completely within one week (CRP = 0.4 mg/L and SAA protein < 6 mg/L compared to 123 mg/L before treatment). Anti-CCP antibody level decreased to 196 U/mL 24 months after initiation of treatment. In conclusion, this is a very rare presentation of rheumatoid arthritis revealed by digestive AA amyloidosis. Both the low level of pain and the presence of multiple lymphadenopathies are unusual in RA, though some series report a high rate of lymphadenopathy among patients with active RA [7]. It shows the wide range of clinical presentations in RA, including cases of predominant extra-rheumatological symptoms and emphasizes the conception of RA as the endpoint syndrome of still poorly understood pathophysiological features rather than as a single source disease. The absence of kidney involvement in AA amyloidosis is also uncommon [1,2,6]. AA amyloidosis can develop for several years before being symptomatic, and digestive amyloidosis is usually only responsible for diarrhea and malabsorption [8,9]. This case also illustrates the importance of precise history taking and clinical examination, as those were the key to reaching final diagnosis here after two years of explorations.

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Disclosure of interest

The authors declare that they have no competing interest.

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