Sirs, A 64-year-old white male visited our department with chronic proximal muscle weakness. His medical history consisted of type 2 diabetes, hyperlipidaemia treated with atorvastatin and morbid obesity. Shoulder and pelvic girdle weakness appeared one year ago and persisted after atorvastatin discontinuation. Musculoskeletal examination showed weak neck flexors, deltoid and gluteus muscles, with preserved distal muscle strength. Several thoracic līvid-red macules were present. No lymphadenopathies were present.

Laboratory tests showed a normal white blood cell count, mild thrombocytopenia (127,000/μL, reference 150,000-450,000/μL) and an erythrocyte sedimentation rate of 6 mm/hour. C-reactive protein was normal (1.0 mg/L, reference <5.0 mg/L). Creatine kinase (CK) was 1256 U/L (reference <190 U/L) with slightly elevated transaminases. Antinuclear antibodies, assessed by indirect immune fluorescence were negative. Anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase antibodies (anti-HMGCR-antibodies) were detected at high titer (132 SGU, reference <20 SGU). Electromyography of the left quadriceps muscle showed myogenic alterations (muscle membrane instability at rest and small short polyphasic action potentials at contraction). Pulmonary function tests were consistent with a restrictive pattern (TLC 58% of predicted; diffusion capacity Tlco 76% of predicted). High resolution computed tomography of the thorax revealed limited reticulations in basal areas suspect for non-specific interstitial pneumonia. Further screening showed multiple bilateral cervical and peri-clavicular lymph nodes, with preserved distal muscle strength. Several thoracic līvid-red macules were present. No lymphadenopathies were present.

Pathological examination of the quadriceps muscle biopsy revealed limited reticulations in basal areas and renal cancers (4, 5). We did not find any evidence of malignancy in the skin biopsy specimen (Fig 1F-H). Further screening showed multiple bilateral cervical and peri-clavicular lymph nodes, with low to absent metabolic activity. Bone marrow aspirate and biopsy were normal. The patient was diagnosed with two necrotising autoimmune myopathy (NAM) and an indolent B-cell non-Hodgkin’s lymphoma, type extranodal marginal zone lymphoma. In this subtype, a watchful waiting approach is recommended in the asymptomatic patient. However, hypothesising this NAM as a lymphoma-related paraneoplastic phenomenon, we considered B-cell targeting therapies as preferential therapeutic approach. He was treated with rituximab 375 mg/m² for 4 consecutive weeks and a tapering scheme of 48 mg methylprednisolone. This resulted in a significant decrease in CK-levels (247 U/L) and recovery of muscle strength.

In this case, one sole biopsy revealed the presence of an anti-HMGCR-positive NAM and a coexistent extranodal marginal zone lymphoma. To the best of our knowledge, this is the first report on statin-induced NAM in the context of lymphoma. This triggers the question whether the abnormal B-cell population could drive anti-HMGCR-antibody production. Unfortunately, at present, this remains an intriguing but unproven hypothesis.

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