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Letter to the Editor

Camptocormia with trigeminal involvement revealing myositis with anti-Ku antibodies

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Anti-Ku antibodies are positive mainly in autoimmune diseases. We report a case of inflammatory myositis with camptocormia and trigeminal signs, associated with the presence of with anti-Ku antibodies.

A 69-year-old woman presented perioral paresthesia, for two years. The patient developed a camptocormia progressively for six months. A perioral dysesthesia in the trigeminal areas was found. An isolate camptocormia was noticed without motor deficiency of the upper or lower limbs (Fig. 1). The rest of the neurological examination was normal. Laboratory analysis showed a creatine phosphokinase of 250 (normal < 170). Antinuclear antibodies were positive at 1/5000. Anti-Ku antibodies were positive and others antibodies against soluble nuclear antigens were negative. Anti-PL7, anti-PL12, anti-JO1, anti-Scl70, anti-MDA-5, anti-Mi2, anti-TIF1, anti-SRP and anti-PM/Scl were negative. An electromyography showed a myopathic signal on the face and limbs, especially on the arm. A lumbar punction was normal. A muscular MRI indicated a symmetric fat involution in thoracic paravertebral muscles, the gluteus maximus, the hamstring muscles and the adductors; a mild STIR signal was noticed on the right vastus medialis (Fig. 1). A lumbar muscular biopsy found a fat involution, an endomysial fibrosis and a slight muscle fiber necrosis. Lymphocyte proliferation areas were identified. A vastus medialis biopsy showed a perivascular and endomysial lymphocyte proliferation areas without inflammatory and without fibrosis. On immunohistochemistry tests, HLA Class I protein expression was increased on membrane and cytoplasm fibers. We conclude that this patient developed an inflammatory myositis with anti-Ku antibodies with trigeminal signs. A thoracic-abdominal CT scan eliminated a neoplasia. We treated this woman with human intravenous immunoglobulins 150 g/month during four months, prednisone during 12 months and mycophenolate mofetyl 1 g/day. The treatment stabilized camptocormia and improved trigeminal signs six month after the beginning of this treatment. The patient as in remission 12 months later.

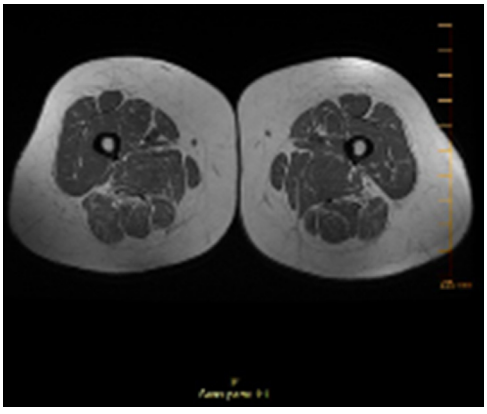

Fig. 1. Fat involution in muscles and a mild STIR signal on the right vastus medialis on MRI.

Anti-Ku antibodies are positive mainly in lupus, Sjögren's syndrome, polymyositis associated with systemic sclerosis and mixed connective tissue disease [1]. Anti-Ku antibodies positive are found 19% in lupus, 14% in systemic sclerosis, 9% in myositis, 20% in Sjögren's syndrome [2] Anti-Ku are often associated with muscular (40%) or articular symptoms (77–83%) [1,3,4]. Thirty-seven percent of anti-Ku antibodies are found in inflammatory myositis mainly in muscular-connective tissue diseases overlaps syndromes [3]. Clinical symptoms in myositis with anti-Ku antibodies are myalgia in all patients with proximal symmetric muscle weakness and axial involvement in 5 patients, and dysphagia [3]. Camptocormia is reported in 6 patients with anti-Ku [3,5]. Autoimmune diseases with anti-Ku antibodies are related most frequently with interstitial lung disease and peripheral neurologic manifestations [3]. Gryga et al. reported five patients with neurological signs with anti-Ku in connective tissue diseases: 3 develop a trigeminal or facial neuropathy, one patient has a trigeminal and facial neuropathy [6]. Rigolet et al. describe two patients with trigeminal signs in systemic scleroderma [3]. Another case of peripheral facial nerve paralysis with anti-Ku antibodies is reported in 2 patients [7]. In our patient, trigeminal symptoms preceded myositis.

Disclosure of interest

The authors declare that they have no competing interest.

References

[1] Belizna C, Henrion D, Beucher A, et al. Anti-Ku antibodies: clinical, genetic and diagnostic insights. Autoimmun Rev 2010;9:691–4.
[2] Yaneva M, Arnett FC. Antibodies against Ku protein in sera from patients with autoimmune diseases. Clin Exp Immunol 1989;76:366–72.
[3] Rigolet A, Musset L, Dubourg O, et al. Inflammatory myopathies with anti-Ku antibodies: a prognosis dependent on associated lung disease. Medicine 2012;91:95–102.
[4] Cavazzana I, Ceribelli A, Quinzanini M, et al. Prevalence and clinical associations of anti-Ku antibodies in systemic autoimmune diseases. Lupus 2008;17:727–33.

https://doi.org/10.1016/j.jbspin.2018.03.013

[5] Zenone T, Streichenberger N, Puget M. Camptocormia as a clinical manifestation of polymyositis/systemic sclerosis overlap myositis associated with anti-Ku. Rheumatol Int 2013;33:2411–5.
[6] Gryga K, Milewski M, Zółciński M, et al. Anti-Ku autoantibodies: series of 5 cases. Pol Arch Med Wewn 2009;119:95–7.
[7] Sutter R, Mengiardi B, Benz D, et al. Facial nerve palsy and anti-Ku autoantibodies. J Neurol 2012;259:361–3.

Sébastien De Almeida Chaves*
Guillaume Moulis
Gregory Pugnet
Laurent Sailler
Léonardo Astudillo
CHU Purpan Toulouse, place du Docteur-Baylac, 31059 Toulouse, France

* Corresponding author.
E-mail address: Seb.deac@hotmail.fr (S. De Almeida Chaves)

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