Acquired Neuromyotonia Precipitated by Thyroid Surgery and Associated with Antiacetylcholine Receptor Antibodies

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Dear Sir,

Receptors and ion channels of the neuromuscular junction are targets of several acquired immune-mediated diseases. Antibodies generated against acetylcholine receptors (AChR) result in myasthenia gravis (MG), while antivoltage-gated calcium channels are pathogenic in Lambert-Eaton myasthenic syndrome [1]. Since the description of Isaacs’ syndrome in 1961, several phenotypic variants of acquired peripheral nerve hyperexcitability syndromes have been described [2]. In neuromyotonia, failure to relax voluntary muscle contraction (‘pseudomyotonia’) occurs together with muscle twitching or cramps. Muscle stiffness, increased sweating and hypertrophic calf muscles can be detected in some patients [2]. The association with other autoimmune diseases, especially MG, and the paraneoplastic origin in some cases, e.g. presence of thymoma, suggests an autoimmune origin. Indeed, plasma exchange alleviates the symptoms and the IgG of patients can transfer the disease. Antibodies against voltage-gated potassium channels (VGKC) can be detected in the peripheral blood of the majority of patients [3–5]. Interestingly, antibodies generated against AChRs have also been described in some patients even in the absence of anti-VGKC antibodies, although their participation in the pathophysiology of the disease has not been investigated [6].

Case Report

The thyroid gland of a 62-year-old man was subtotally removed because of nodosal enlargement. Hyperthyroidosis was indicated by elevated free triiodothyromine, free thyroxine and decreased thyroid-stimulating hormone levels. Histology did not show any inflammation or tumor tissue, and he has been successfully treated with L-thyroxine. Two weeks after surgery, he realized that he could not relax his grip, and had generalized muscle stiffness. On neurological examination, signs of clinical pseudo-myotonia with delayed relaxation were observed without any indication of neuropathy, myopathy or involvement of the central nervous system. No clinical symptoms and signs of MG were observed. Simultaneous concentric needle electromyography on superficial digital flexor, deltoid and abductor pollicis brevis muscles showed continuous spontaneous motor unit activity at rest with repetitive double, triple and multiple bursts, compatible with neuromyotonic and myokimic discharges (Fig. 1a). Lidocaine blockade of the right median nerve did not modify the spontaneous discharges. The analysis of twenty motor units demonstrated normal duration of muscle potentials. Electromyographic recordings, including single-fiber electromyography and repetitive nerve stimulation, did not indicate any sign of MG, or Lambert-Eaton myasthenic syndrome. The sensory and motor conduction velocity, the compound muscle action potential, the sensory nerve action potential and F wave latency of the median nerve were normal. There was no indication of hereditary myotonia, periodic paralysis or any neurological disorder in his family or in his medical history. The hemogram, serum levels of thyroid and parathyroid hormones, calcium, phosphate, potassium, sodium, chloride, magnesium, creatinine, glucose, aldolase, lactate, liver function analysis, urinalysis were all normal. Creatinine kinase levels were slightly elevated (427 U/l). Total serum immunoglobulin, cryoglobulin, complement levels were in the normal range. A markedly elevated level of anti-AChR an-

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Anti-AchR-Positive Neuromyotonia after Thyroid Surgery

Fig. 1. Neuromyotonic discharges in deltoid (upper panel) and superficial digital flexor muscles (lower panel). Neuromyotonic discharges recorded 3 weeks after surgery (a) decreased after clinically effective treatment with mexiletine (b).
References