Reversible Focal Neuromyotonia in SLE

Sir,

A 42-year-old woman with a 10 year history of Systemic lupus erythematosus [SLE] presented with constant painful spasms of her left 4th and 5th fingers of 2 weeks duration [Video 1]. These spasms persisted in sleep. She was on chronic immunosuppression with Mycophenolate mofetil 1 gm/day [MMF] and Prednisolone 20 mg/day, and was otherwise in remission. Her nerve conduction studies [NCS] were normal. EMG showed Neuromyotonia restricted to the left 4th lumbrical and abductor digiti Quintii muscles [Video 2]. EMG of other muscles were normal.

Focal neuromyotonia [isolated finger flexion] secondary to SLE was considered and she was started on Carbamazepine 200 mg tid. Within a week, her symptoms resolved and we could stop Carbamazepine for a month. An extensive blood work and MRI cervical spine were normal. Voltage gate potassium channel antibodies [VGKC] were also negative. She was continued on her MMF and Prednisolone. At her last follow-up 6 months later, she remained asymptomatic. A repeat NCS and EMG studies were normal.

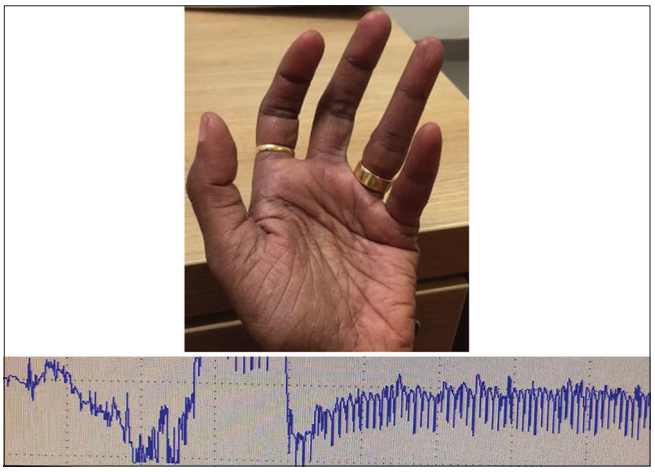

Figure 1: Photograph of left hand showing isolated finger flexion of the 4th and 5th fingers. Horizontal shows a run of neuromyotonia

Acquired Neuromyotonia is a condition that presents with generalized pain, muscle twitching [Myokimia and fasciculations], cramps, muscle stiffness and hyperhidrosis. Sensory symptoms such as pain and paraesthesia are present in a majority of patients. It goes by a number of synonyms such as 'Continuous muscle fiber activity [CMFA] syndrome, Isaacs-Merten Syndrome, Isaacs' Syndrome or 'Quantal squander'. 20% of patients have an associated thymoma and the disease can be associated with other peripheral neuropathies, autoimmune neuropathies or myasthenia gravis.

In addition to the peripheral nervous system [PNS] involvement, central nervous system [CNS] features can be present. These include insomnia, anxiety, hallucinations and behavioral changes. When the CNS manifestations predominate, the term Morvan's 'fibrillary chorea' or Morvan's syndrome is often used.[1] EMG shows continuous spontaneous activity such as doublets, triplets or multiplets [Neuromyotonic discharges]. These neuromyotonic discharges are high frequency motor unit discharges [high intraburst frequency] that begin

or end abruptly. Unlike myotonia, where the discharges have a waxing and waning quality of discharges a 'dive bomber' quality, these come to an abrupt end or wane off. Neuromyotonia is associated with a number of autoimmune causes and approximately 40% of the patients have associated VGKC antibodies. In addition, CSF examination can show raised proteins or Oligoclonal bands further supporting the autoimmune basis of the disease.

Neuromyotonia is often generated by axonal instability and not affected by general or spinal anaesthesia. However, they are abolished by neuromuscular blocking agents and diminished with nerve blocks depending on the site of generation. The pathogenesis of CMFA is postulated to be the down regulation of fast potassium channels, which prolongs depolarization and Sodium channel-blockers such as phenytoin, carbamazepine or mexiletine are often used due to their membrane-stabilizing properties. Other modalities such as IV methylprednisolone, Plasma exchange or IVIG have also been helpful.

Unlike generalized neuromyotonia, focal neuromyotonia [FMN] is rare. FMN can be seen after radiotherapy, radiculopathy, multifocal motor neuropathy, transposed muscle flaps, compression neuropathies and LGI-1 autoimmunity.[2,3] Ocular FMN has also been described after radiotherapy, cavernous sinus thrombosis, Graves' disease etc.[4] Idiopathic FMN or CMFA with isolated finger flexion has been described earlier.[3,5]

However, to the best of our knowledge, this is the first reported case of focal neuromyotonia in SLE, although generalized neuromyotonia has been noted in association with VGKC antibodies.[6] As of now, no direct causative relationship has been established with FMN and more work is needed on this front.

In our patient, FMN due to SLE caused muscle stiffness and cramps along with involuntary finger flexion. Fortunately, our patient displayed a prompt and sustained response to Carbamazepine.

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Conflicts of interest

There are no conflicts of interest.

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