Teaching Video NeuroImages: Regional myokymia

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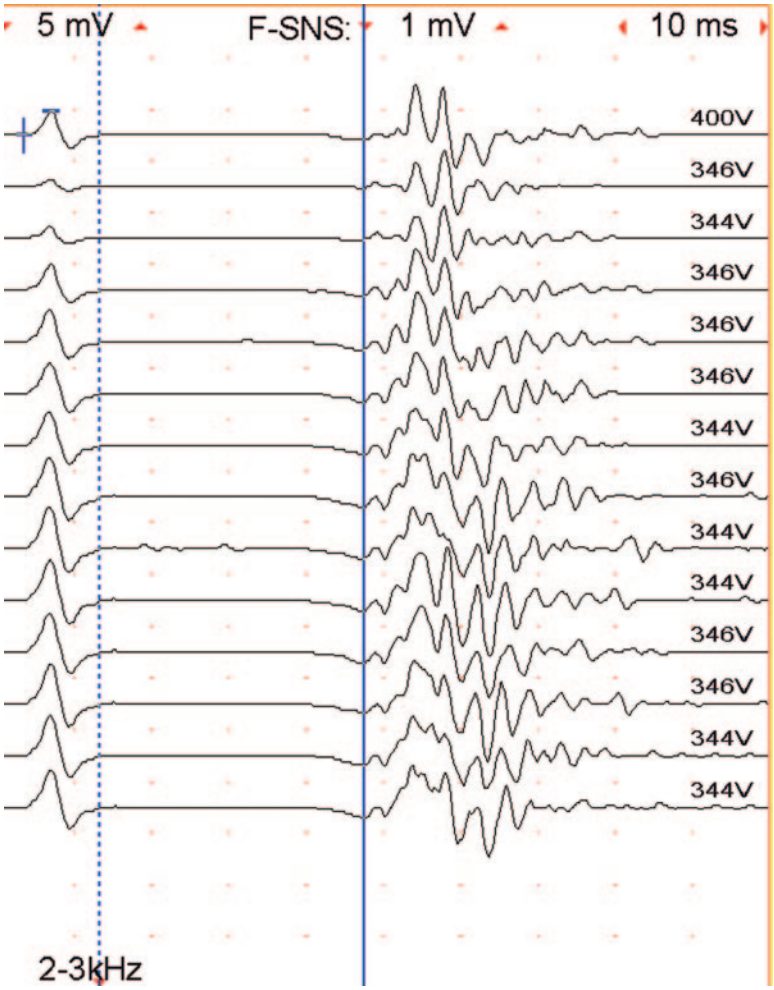
Figure 1 Tibial nerve conduction studies showing consistent F-wave hyperexcitability



A 26-year-old man presented with involuntary muscle twitching of the bilateral lower extremities. The continuous muscle rippling was noted in bilateral calves, posterior thighs, and buttocks (video). Nerve conduction studies revealed F-wave hyperexcitability (figure 1). EMG recording showed doublet, triplet, and multiplet motor unit discharges (figure 2). The clinical presentation and electrophysiologic findings suggested a diagnosis of Isaac syndrome, although symptoms are typically regional rather than generalized. This patient was successfully treated with phenytoin. Isaac syndrome was first described in 1961,[1] and is characterized by peripheral nerve hyperexcitability which manifests clinically as generalized muscle twitching, stiffness, pseudomyotonia, and cramping. Some patients have muscle hypertrophy and hyperhydrosis. Antibody to voltage-gated potassium channels can be detected in 40% of patients.

Supplemental data at www.neurology.org

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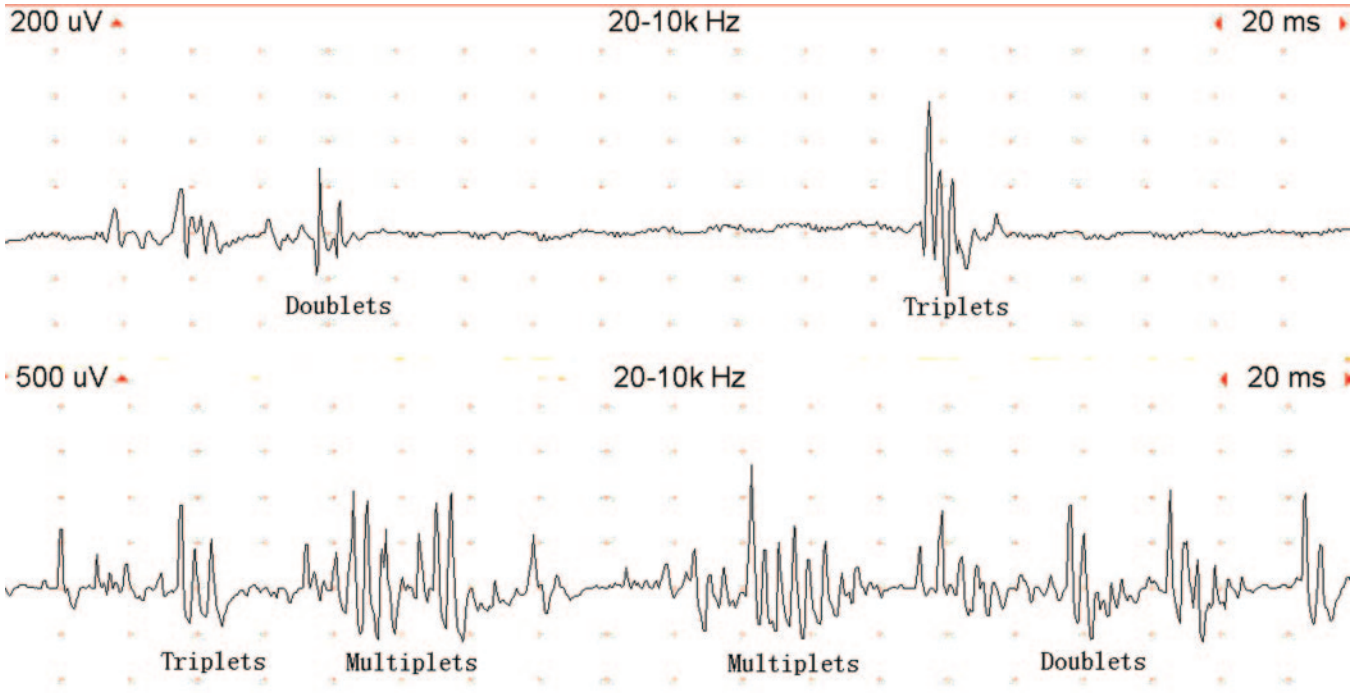
Figure 2 Spontaneous continuous irregularly occurring doublet, triplet, and multiplet single and partial motor unit discharges in medial gastrocnemius



Associated autoimmune disorders and other autoantibodies are found in approximately 50%, most notably antiacetylcholine receptor antibodies, indicating coexistent myasthenia gravis in about 20%. Typical EMG findings include continuous single motor unit discharges occurring as doublets, triplets, and multiplet single unit discharges firing at a high intraburst frequency (150–300 Hz; neuromyotonic discharges), and at lower frequencies (less than 60 Hz; myokymic discharges). The symptoms often respond well to anticonvulsants such as phenytoin and carbamazepine.[2]

REFERENCES

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2. Maddison P. Neuromyotonia. J Clin Neurophysiol 2006; 117:2118–2127.

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