A PICTOGRAM OF A YOUNG MALE WITH THE OPENING DIFFICULTY OF HIS GRIP

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An 18 years male visited the Neurology outpatient department of Dhaka Medical College with the opening difficulty of the grip for the last five years. It improved with the repetitive opening and closing of the grip was painless and did not increase with exposure to the cold. He had muscular hypertrophy in the different groups of the muscle. There was no difficulty in opening and closing the eyelids. He did not have any family history of such type of illness.

Difficulty in opening the grip-Myotonia

Hypertrophy of the calf muscles

Hypertrophy of the supraspinatus, infraspinatus, & paraspinal muscles

Questions:

What are the findings shown in the pictures? What is the clinical diagnosis of the patient? What is the treatment of the condition?

The patient had Myotonia with a warm-up phenomenon and hypertrophy of the muscles.

Provisional Diagnosis: Myotonia congenita

Differential diagnosis:

- Myotonic dystrophy
- Paramyotonia Congenita
- Potassium-aggravated Myotonia

Investigation findings:

Serum electrolyte was within normal. The EMG revealed a dive-bombers sound with a burst of repetitive action potential with a frequency of 60 Hz and 60 μV amplitude.

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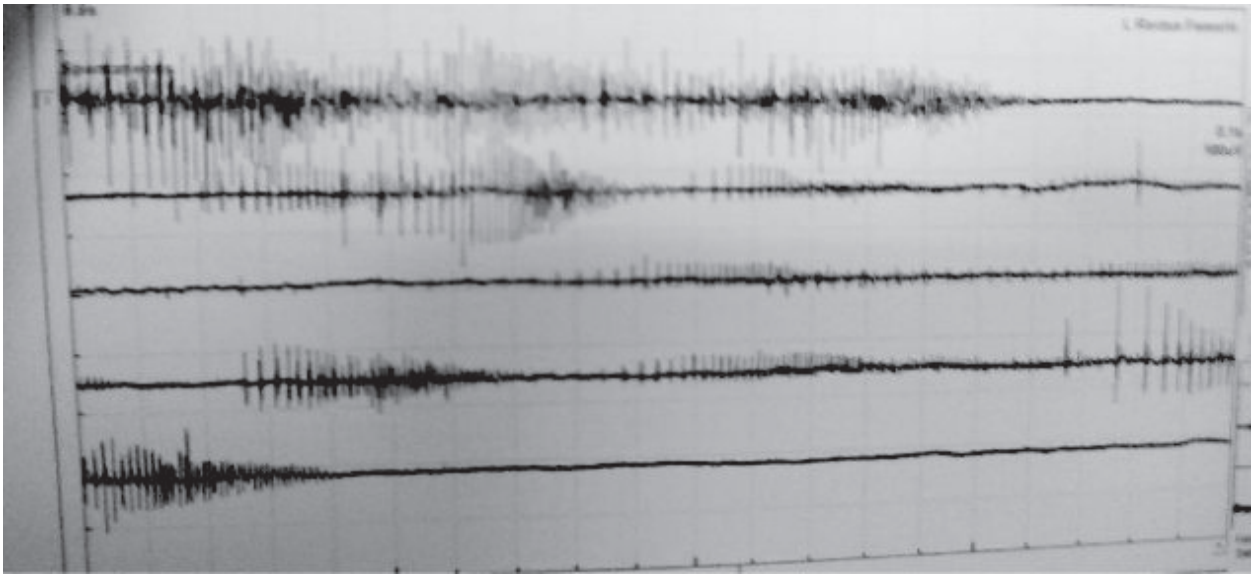

Case discussion:

The patient had grip myotonia both clinically and in EMG. We could differentiate it from Myotonic dystrophy by the warm-up phenomenon, absence of orbicularis oculi involvement, and muscular hypertrophy. In addition to these, the absent cold-induced exaggeration differentiates it from the Paramyotonia congenita. The potassium-exaggerated myotonia is usually painful and has prominent involvement of orbicularis oculi muscles. Their absence, in this case, helps in differentiation.

The Myotonia Congenita has two modes of inheritance. The Thomsen myotonia, an autosomal recessive form present in the first decade, and the autosomal dominant Becker myotonia, usually in the second decade. As our patient presented in the second decade, it suggests the Becker Myotonia.

We treated the patient tab phenytoin 100 mg twice daily with a good result in the subsequent follow-up.