

## Case Report

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# Hoffmann's Syndrome a Presenting Manifestation of Hypothyroidism

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## Abstract

Hypothyroidism associated with different neuromuscular manifestations. Rarely patient with hypothyroidism may develop muscular pseudohypertrophy a condition called Hoffmann's syndrome in adults. We report a patient present mainly with myalgia, cramps, calf pseudohypertrophy and persistently elevated Creatine Kinase (CK) with absence of other systemic symptoms of hypothyroidism. His laboratory works revealed severe hypothyroidism. Treatment with L-thyroxine resulted into disappearance of symptoms, normalization of CK and return of calf size to normal. Hypothyroidism should be considered when evaluating patients with neuromuscular complaints even in absence of systemic symptoms.

**Keywords:** Hoffmann's syndrome; Hypothyroid; Myopathy; Case report

## Introduction

Hypothyroidism is one of the most prevalent endocrine disorders. Different neuromuscular complications can develop with hypothyroidism. Nonspecific neuromuscular complaints like myalgia, muscle cramps and mild elevation of Creatine Kinase (CK) are the most common myopathic features related to hypothyroidism. Other myopathic features included proximal muscle weakness and myoedema. Rarely hypothyroid myopathy may associate with muscular pseudohypertrophy a condition called Hoffmann syndrome in adults and Kocher-Debre-Semelaigne syndromes in children.

We report a case of hypothyroidism presented solely with myopathic feature and calf pseudohypertrophy in absence of other systemic symptoms with resolution of symptoms with L-thyroxine treatment.

## Case Presentation

A 54 year old male patient was evaluated in the neurology clinic for persistently elevated Creatine Kinase. He was assessed initially at the cardiology clinic for syncope episode 6 months prior to his neurology evaluation. His cardiac work up including electrocardiogram, 24 holter monitor and 2D echocardiogram were unremarkable. He was diagnosed with vasovagal syncope. In laboratory works he was found to have high cholesterol level for which he was started on atorvastatin 20mg once daily. No base line CK was done. One month after starting atorvastatin, liver function test and CK were checked. His CK level was found at 1208 U/L (35-232 U/L). The diagnosis of statin induced myopathy was made and statin was discontinued. Repeat CK in two months time showed CK level of 1800 U/L.

When he was seen in neurology clinic, the patient gave 2 years history of easily fatigability, exercise intolerance and calf cramps following minimal exertion. He also reported significant increase in his calf muscle size over the last 2 years. He has no muscle weakness and no myotonia. No history of cold intolerance, weight gain or change in bowel habit. No family history of muscle disease, muscle hypertrophy or hyperkemia.

He is known case of type II diabetes on oral hypoglycaemic and hypertensive on angiotensin converting enzymes inhibitors.

Physical examinations revealed normal blood pressure and heart rate. He has coarse facial features and hoarse voice. No thyroid enlargement. His motor examination showed enlarged tender calves bilaterally. No muscle weakness and he have normal tendon reflexes.

Nerve conduction study showed evidence of bilateral carpal tunnel syndrome and normal lower limb study normal electromyography.

Blood works revealed CK 1800 U/L, aspartate aminotransferase (AST) 59 U/L (15-37 U/L), lactate dehydrogenase (LDH) 325 U/L (85-227 U/L).

Thyroid function test showed thyroid stimulating hormone (TSH) of  $10^3$  mIU/L (0.4-4.2 mIU/L), T3 0.33 pg/ml (2.01-4.2 pg/ml), T4 0.43 pg/ml (0.9-2.2 pg/L).

The patient diagnosed with Hoffman syndrome. Treatment with L-thyroxine was started. In 2 month follow up the patient reported significant improvement in his exercise tolerance. Decrease in his calf size and disappearance of muscle cramps.

His repeat blood works showed normalization of his CK and TSH.

## Discussion

Persistently elevated CK is one of the common referral causes to the neuromuscular clinics. Although CK is predominantly reflect muscle related disorders, elevated CK can be seen in non neuromuscular causes as well in non myopathic neuromuscular disorders. Before attributing the high CK to myopathic cause one should rule out non neuromuscular causes that include hypothyroidism, drugs related high CK, toxin, alcohol and trauma. Mild elevation of serum CK also seen in neurogenic muscle atrophy and can be seen with Amyotrophic Lateral Sclerosis (ALS) [1]. Patient with hypothyroidism commonly present with mildly elevated serum CK. Severe elevation of serum CK and rhabdomyolysis with association with hypothyroidism have been [2]. There is no relation between the level of serum CK and the degree of weakness [3,4]. The cause of elevated CK in hypothyroidism is not very well understood. Muscle degeneration is thought to be the cause of elevated CK. Other possible cause includes decrease CK clearance [2]. With L-thyroxine treatment the serum CK return to normal.

Hoffmann's syndrome is a rare type of hypothyroid related

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myopathy. It was described first at 1897 by Hoffmann [5]. The clinical features of Hoffmann's syndrome are variable and included myelgia, muscle cramps, and varying degree of weakness. However, the cardinal feature is an increase in muscle size. Myotonia has been also reported in association with Hoffmann's syndrome [6].

Electrophysiological studies can be normal or can show nonspecific myopathic feature. Our patient had a normal EMG studies and that may reflect absence of motor weakness on his exam.

The mechanism of increase in muscle size in hypothyroidism is not well understood. A different mechanism is thought to be the cause of muscle hypertrophy that include increase in muscle size, increase in the number of muscle fibre and increase in connective tissue.

Muscle biopsy in patient with hypothyroid myopathy showed predominance of type II fibre [7], presence of Core like structure [7,8] and abnormal glycogen accumulation [9]. In the study by Ono in 1987 none of the cases of Hoffman's syndrome showed individual muscle hypertrophy [7]. However, Modi reported that the presence of cores correlate with the severity of hypothyroid, muscle hypertrophy, cramps and the duration of hypothyroid status [8].

In patient presenting with elevated CK assessment of thyroid function test is necessary to rule out hypothyroidism even in absence of systemic symptoms.

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