



An Unusual Neurological Syndrome of Hypothyroidism

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Abstract

Neurological manifestations of hypothyroidism are not usually seen early in the course of the disease, particularly muscle disease. Entrapment neuropathy is more common and may be a presenting symptom. Hypothyroidism presenting as muscle stiffness and pseudohypertrophy in adults is known as Hoffman's syndrome which is supported by presence of elevated CPK and EMG features of myopathy, neuropathy or mixed pattern. With hormone replacement therapy symptoms and CPK levels normalize promptly. We report a case of hypothyroidism with extreme muscle pain, calf muscle hypertrophy as initial symptoms.

Introduction

The symptoms of hypothyroid myopathy are proximal muscle weakness, muscle cramps, pains, myxedema on muscle percussion, delayed relaxation of deep tendon jerks and rarely, hypertrophy.^[4,8]

Hoffman's syndrome is a form of hypothyroid myopathy seen in less than 8% of hypothyroid patients particularly the hypertrophy and stiffness form.^[4,7] In children with congenital hypothyroidism, a similar syndrome of muscle hypertrophy known as Kocher-Debre-Semelaigne syndrome.^[6]

Case Report

A 40-year-old man presented with history of puffiness of face, extreme cramps and pain over upper and lower limbs particularly deltoid area, calf muscle and thighs, fatigability, over somnolence. There were no sensory symptoms. He did not give any specific symptoms of cold in

tolerance, clinically his pulse rate was 70 beats /min, BP of 130/70 mm Hg, facial puffiness with prominence of both parotid regions.

He had bilateral calf muscle hypertrophy with normal power and delayed relaxation of ankle jerks. His higher functions, cranial nerves were normal. Cardiovascular examination was normal.



Figure 1: Facial Puffiness



Figure 2: Left calf muscle hypertrophy



Figure 5: Bilateral calf muscle hypertrophy

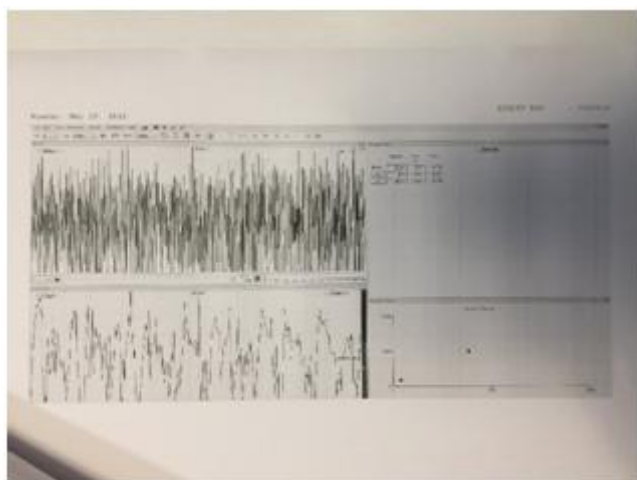
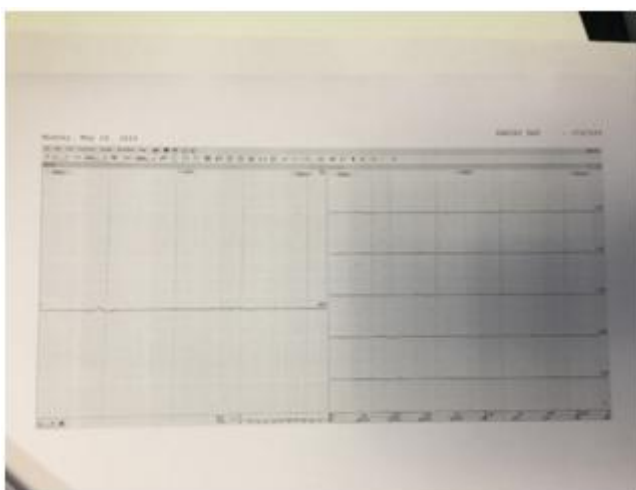


Figure 2 & 4 : Electromyography report showing myopathic motor unit potentials

The laboratory investigations revealed normal WBC and RBC count, blood sugars, electrolytes, ECG, X-ray, USG and ECHO.

His uric acid was 9.3mg/dL, total cholesterol was 300mg/dL, LDL 132 mg/dL and triglycerides 1016 mg/dL (fasting blood sample).

The LFTs showed AST 60 IU, ALT 81 IU and GGTP 136 IU (He consumes ethanol once a week).

He has severe obstructive pulmonary impairment in the pulmonary function tests.

His TSH was 147.2 μ IU /mL (Normal value – 0.4-4.2 μ IU/mL),

T3 – 0.8 pg/dL (Normal 2.2.-4.2 pg/dL),

T4 – 0.3 ng/dL (Normal 0.9-1.7 ng/dL), and TPO Antibodies were negative.

Total CPK levels were 942 U/L (normal up to 240 U/L).

Electromyography showed myopathic MUAPs (Motor Unit Action Potentials). Muscle biopsy was not done as patient refused to give consent.

Discussion

Hoffman’s syndrome was first described by Hoffman in 1897 in an adult after thyroidectomy.

[1] The first presentation with muscle stiffness, myopathy and pseudohypertrophy is very unusual

[4]. Primary hypothyroidism is the most common cause of thyroid insufficiency. Since TPO

Antibodies were negative, Hashimoto's thyroiditis was ruled out. The etiology of pseudo hypertrophy in Hoffman's syndrome remains obscure though increase in connective tissue levels has been postulated. Abnormalities in oxidative enzymatic activity and hypertrophy due to accumulation of glycosaminoglycans,^[2,4] and changes in muscle fibres from fast twitching Type II to slow twitching type I fibres have all been postulated as possible mechanisms^[6] The common groups of muscles affected are arm and leg muscles.^[4] Following treatment with thyroxine, hypertrophy and weakness recede well.^[9] CPK levels are elevated in thyroid myopathy (10-100 times greater) and is still the best biochemical marker of myopathy^[5]. But the fall of enzyme levels with the treatment happens slowly and may take weeks to months.

Our patient presented with predominant muscle pain and stiffness and hypertrophy of calf muscles with 5-fold elevation of CPK levels and EMG showing myopathic motor unit end potentials.^[3] He has been started on L-thyroxine 100 µg /day.

In view of the rarity of presentation with muscle pain and stiffness, awareness about this syndrome leads to early correct diagnosis.

Conclusion

Hoffman's syndrome has good prognosis with symptoms remissions seen as early as 2-3 months. On very rare occasions, worsening of symptoms in some at the beginning of treatment may occur, probably caused by rise in metabolic demand induced by thyroxine. In these cases, the use of steroids during some time of the treatment as membrane-stabilizing effect can be beneficial.^[10]

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