A young lady with swelling and stiffness of calf muscles

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INTRODUCTION

Hypothyroidism is a “Pandora’s Box”. It can present in a myriad of ways – with classical signs in some, subtle signs in some and unusually in some patients. It causes metabolic changes in several systems of the body including the muscles as well. We report a case of Hoffman’s syndrome, a rare form of hypothyroid myopathy, which causes proximal weakness and pseudohypertrophy of muscles.

CASE REPORT

A 20-year-old lady came to OPD with complaints of swelling and stiffness of both calf muscles which started 15 days back. She complained of cramping in both calf muscles and difficulty in getting up from squatting position and climbing stairs. She did not complain of cold intolerance, weight gain, increased sleep, lethargy, constipation, hoarseness of voice or menstrual disturbance. On examination, her pulse rate was 62/min and BP was 120/90 mm of Hg. There was an apparent hypertrophy of bilateral calf muscles (gastrocnemius) [Figure 1], dry coarse skin and mild diffuse goiter. She had proximal muscle weakness in both lower limbs with delayed relaxation of ankle jerk. There was no calf muscle tenderness. Other systems were unremarkable.

Investigations revealed T3 0.25 ng/ml (normal range: 0.60–1.81 ng/ml), T4 1.12 µg/dl (normal range: 4.5–12 µg/dl), TSH 150.2 µIU/ml (normal range: 0.3–5.5 µIU/ml) suggestive of primary hypothyroidism. Creatine kinase was elevated (742 U/l) (normal <140 U/l). Complete hemogram, random blood sugar (RBS), urea, creatinine, serum electrolytes, liver function tests (LFT) and fasting lipid profile were unremarkable. ECG and chest X-ray did not reveal any abnormality.

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ABSTRACT

Hypothyroidism causes a variety of changes in the body. Though uncommon, hypothyroidism can present as myopathy. Hoffman’s syndrome is a specific, rare form of hypothyroid myopathy, which causes proximal weakness and pseudohypertrophy of muscles.

Key words: Hoffman’s syndrome, hypothyroidism, myopathy

Figure 1: Pseudohypertrophy of calf muscles

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The patient was diagnosed to have primary hypothyroidism complicated by hypothyroid myopathy – Hoffman’s syndrome.

She was started on Thyroxine 50 µg od that was increased to 100 µg od after 2 weeks. The patient had significant improvement in terms of biochemical parameters and clinically as well after 3 months of treatment.

**Discussion**

Hypothyroidism causes a variety of changes in the body. Deficiency of thyroid hormone interferes with metabolic function by slowing it down or decreasing it. These metabolic changes occur in the muscles also.[1,2] Pain on muscle exertion is due to defective carbohydrate metabolism. Delayed muscle contraction and relaxation may be due to a change in the distribution from fast-twitch fibers to slow-twitch fibers.[1,2] A reduction in muscle mitochondrial oxidative capacity and beta-adrenergic receptors, as well as the induction of an insulin-resistant state, may result in these changes.[1,2] Neuromuscular symptoms are present in 30–80% of patients with hypothyroidism. Patients may have muscle cramping, proximal symmetrical muscle weakness, muscle stiffness, and exercise intolerance.[1,2] Slowness of muscle relaxation and of muscle contraction is noted in hypothyroid myopathy. Delayed relaxation of deep tendon reflexes is due to impaired calcium sequestration by sarcoplasmic reticulum, which prolongs twitch duration.[3] Deep tendon reflexes are delayed in approximately 85% of patients with hypothyroidism. Mounding of the muscle after light percussion (i.e., myoedema) occurs in one-third of patients with hypothyroidism. Calf muscle hypertrophy with weakness is seen in Duchenne and Becker muscle dystrophy, focal myositis, sarcoid granulomas and amyloid deposits in muscles. Although muscular symptoms are common in hypothyroid patients (varying from myalgia, weakness, stiffness, cramps and easy fatigability in 30–80% of the patients), muscular hypertrophy with muscle stiffness is reported in less than 10% of the patients.[4,5] Pseudohypertrophy of the muscles results from accumulation of glycosaminoglycans.[5] Gastrocnemius is almost always involved as it was in our case. Muscle enlargement, stiffness and cramping, proximal muscle weakness are a constellation of findings seen in these patients.

Hoffman’s syndrome is a specific, rare form of hypothyroid myopathy, which causes proximal weakness and pseudohypertrophy of muscles. In adults with hypothyroidism, these findings are known as Hoffman’s syndrome. In children with cretinism, they are called Kocher-Debré-Sémélaigne syndrome.[6]

We are reporting this case of Hoffman’s syndrome for rarity of this clinical entity.

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