



BMH Med. J. 2018;5(3):85-87 **Case Report**

A Rare, Yet Treatable Cause Of Muscle Weakness

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Abstract

Muscle hypertrophy and weakness are common symptoms of muscular disease which usually show poor response to treatment and poor prognosis most of the time. But there are potentially treatable conditions which can mimic muscular disease signifying the importance of proper clinical evaluation of myopathic symptoms to recognise treatable conditions at the earliest. In this case we are discussing about an adult male who presented with myopathic symptoms which significantly improved with specific management of the underlying condition.

Key words: muscle weakness, muscle hypertrophy, hypothyroidism

Introduction

Myopathic symptoms like fatigue, muscle weakness, and cramps are common and can be due to variety of causes. Hypothyroidism can present with wide range of symptoms including myopathic symptoms. Myopathic manifestations in hypothyroidism includes fatigue, exertional pain, slow movement, diminished deep tendon reflexes, stiffness, myalgia, myoedema, proximal weakness, and cramps [1]. Pseudo-muscular hypertrophy is a rare manifestation seen in patients with hypothyroidism.

Case history

A middle aged male patient presented with history of progressive difficulty in squatting and difficulty in getting up from sitting position for the last 2 years without any sensory, bowel, bladder or cranial nerve symptoms. He also gives history of generalised oedema including facial puffiness and bilateral oedema of the legs. There is no history of any other significant symptoms or major illnesses in the past. On examination he was obese with BMI of 31, pale looking with hoarse voice, coarse skin, macroglossia and bilateral non pitting pedal oedema. His pulse rate was 52/ minute and BP of 120/76 mm of Hg. Neurological examination revealed weakness of all four limbs with predominant weakness of the proximal muscle groups, generalized hyporeflexia and delayed

relaxation of deep tendon reflexes. There was also bilateral calf muscle hypertrophy (**Figure 1**). Routine blood investigation including complete blood count, renal function test and liver function test were within normal limits. ECG showed sinus bradycardia. His lipid profile showed high total cholesterol, and CPK was elevated. Thyroid function test showed TSH $>100\mu\text{U}/\text{ml}$ (normal 0.27 - 4.94 $\mu\text{IU}/\text{mL}$) with very low T3 and T4 suggestive of severe hypothyroidism. A provisional diagnosis of Hoffman syndrome, that is hypothyroidism associated myopathy and pseudohypertrophy of muscles was made and he was started on 100 microgram of thyroxin and kept under follow up. Within the first three months itself he showed significant improvement in his muscle weakness and there was partial regression of muscle hypertrophy.



Figure 1: Bilateral calf muscle hypertrophy

Discussion

Myopathic symptoms are seen in 30 to 80% of patients with hypothyroidism [2]. The common myopathic symptoms in hypothyroidism include proximal muscle weakness, cramps, myoedema, delayed relaxation of deep tendon reflexes and rarely pseudo hypertrophy of the muscles [3]. The different variants of myopathies described in hypothyroidism includes Hoffmann syndrome, Kocher-Debre-Semelaigine syndrome, atopic myopathy, myasthenia syndrome and polymyositis like syndrome [4,5]. Pseudo hypertrophy is seen usually in those who are having severe long standing hypothyroidism.

Muscle pseudo hypertrophy associated with hypothyroidism is called Hoffman syndrome in adults and Kocher-Debre-Semelaigine syndrome in children. Such manifestations are seen in less than

10% of hypothyroid individuals [5]. The manifestations in Hoffman syndrome include proximal muscle weakness, stiffness and pseudo hypertrophy of skeletal muscles especially calf muscles. Other muscles involved are thigh, forearm muscles and tongue [2]. The pseudo hypertrophy of the calf muscle is due to accumulation of glycosaminoglycan, increase in the connective tissue and increase in the muscle fibre size and number [5]. Hoffmann syndrome was first described by John Hoffman in 1897 in a patient who developed stiffness and difficulty in relaxation following thyroidectomy [5]. In Hoffman syndrome the fast twitch type 2 muscles are changed into slow twitch type 1 muscles and there is alteration of oxidative muscle enzyme activity with decreased calcium ATPase activity resulting in delayed relaxation of the muscles [5]. Investigation shows mild to moderate increase in CPK level in 70 to 90% of the cases. But this level does not correlate with the muscle symptoms or severity of the disease. AST (Aspartate Amino Transferase), LDH (Lactate Dehydrogenase) and aldolase are also found to be high. Electrophysiological studies show myogenic, neurogenic or mixed pattern and nerve conduction studies may show entrapment neuropathy or axonal sensorimotor polyneuropathy. Muscle biopsy shows fibre necrosis, atrophy or hypertrophy with increase in the number of nuclei and increase in connective tissue.

Because of the presence of muscle pseudo hypertrophy and myopathic symptoms it has to be differentiated from other myopathies like Becker's Muscular dystrophy, Duchenne Muscular dystrophy, focal myositis, amyloidosis, myotonia and polymyositis. Hoffman syndrome has favourable prognosis and usually subsides with treatment of hypothyroidism [5]. CPK level will become normal but it may take months or years. Clinicians should be familiar with Hoffmann syndrome because it is a treatable cause of myopathy associated with good prognosis.

Conclusion

This case highlights the importance of identification of potentially treatable conditions which can mimic myopathy. Hypothyroidism can present with myopathic symptoms and pseudo-muscular hypertrophy, which is a potentially treatable condition.

References

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