Hoffmann Syndrome: Presentation in Hypothyroidism

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Hoffmann syndrome is muscle hypertrophy with muscle weakness complicating hypothyroidism. In this communication, a case of Hoffmann syndrome complicating primary hypothyroidism (due to Hashimoto's thyroiditis) in a male is described.

A forty-three-year-old male presented with 6 years history of lower limb weakness and dyspnea for the past 1 month. Central nervous system examination revealed a patient with calf muscle hypertrophy (Figure 1) and generalized hypotonia with muscle power of 3/5, with the proximal group of muscles affected more than the distal muscles. Deep tendon reflexes were normal except for ankle jerks, which revealed delayed relaxation, the pseudomyotonic reflex. On investigating, the patient had anaemia, hypertriglyceridemia and hypercholesterolemia. Thyroid profile revealed a T3 level of 19 ng/dl (normal 60–200 ng/dl), a T4 of 1.9 μg/dl (normal 4.5–12 μg/dl) and a Thyroid stimulating hormone (TSH) level of 150 μI U/ml (normal 0.3–5.5 μI U/ml). FNAC of the thyroid swelling (goitre) revealed features of Hashimoto's thyroiditis. Creatine phosphokinase revealed a value of 1780 U/l (normal <140 U/l). Serology for anti-microsomal antibody was positive in high titres. Chest roentgenogram was normal and electrocardiogram revealed low voltage complexes. Echocardiogram revealed dilated cardiomyopathy with an ejection fraction of 42%. Electromyographic (EMG) evaluation showed mild spontaneous activity, polyphasic myopathic motor unit potentials (MUAPs) with small amplitude and duration in proximal muscles consistent with hypothyroidism. The nerve conduction studies (NCS) of the nerves of both extremities were normal with prolonged F wave latencies in the common peroneal nerves bilaterally.

Although, muscular symptoms are common in hypothyroid patients (varying from myalgia, weakness, stiffness, cramps and easy fatigability in 30–80% of patients), muscular hypertrophy with muscle stiffness is reported in less than 10% of the patients.[1,2] Hoffmann's syndrome is a specific, rare form of hypothyroid myopathy, which causes proximal weakness and hypertrophy of muscles. The neurological manifestations of hypothyroidism usually occur later and is unusual to see it as the initial symptoms.[1]

Calf muscle hypertrophy accompanies a wide variety of diseases like Duchenne and Becker muscle dystrophy, infiltration by sarcoid granulomas, amyloid deposits and focal myositis. The muscle involvement in hypothyroidism is caused by changes in muscle fibres from fast twitching type II to slow twitching type-I fibres. There is hypertrophy of the muscles due to accumulation of glycosaminoglycans. Gastrocnemius is almost always involved as our patient.

The thigh, arm and forearm muscles may also be involved. It is usually associated with delayed muscle relaxation, producing the pseudomyotonic reflex. The rate-limiting step in muscle relaxation is reuptake of calcium by the sarcoplasmic reticulam, which is dependent on calcium ATPase content (SERCA-1) of the muscle fibre. Calcium ATPase activity of fast twitch variety of muscle fibre is decreased in hypothyroidism producing delayed relaxation, the pseudomyotonic reflex.

EMG findings in hypothyroid myopathy show fibrillations, positive sharp waves and complex repetitive discharges (CRD). Our patient had mild neuropathy on NCS, which can also be observed in patients with hypothyroidism.[3,4] With thyroxine replacement not only the serum levels of enzymes but also the enlargement of the muscles and the symptoms of weakness returned to normal in 3 months in our patient. (Figure 4) The puffiness of face and eyelid oedema resolved after 3 months of thyroxine replacement (Figures 2 and 3). Repeat echocardiogram showed an ejection fraction of 55% with complete reversal of the dilated cardiomyopathy. The EMG and NCS done after 6 months revealed reversal to normal.

This rare clinical presentation of Hoffmann syndrome in hypothyroidism due to Hashimoto's thyroiditis is highlighted in order to focus the attention on the occurrence of this rare complication in the absence of overt manifestations of hypothyroidism.

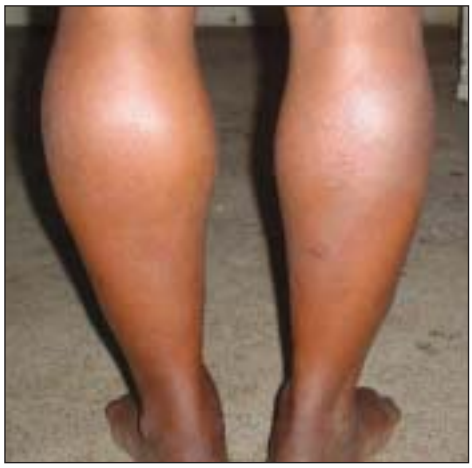

Figure 1: Photograph of the patient shows prominent muscle hypertrophy involving both the gastrocnemius muscles

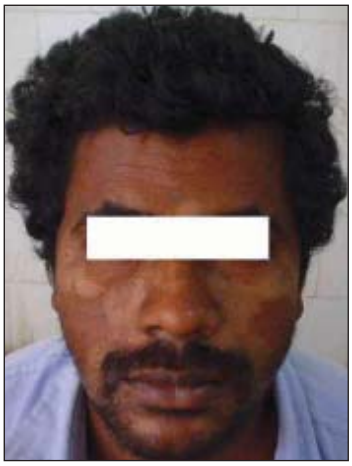

Figure 3: Patient after treatment with regression of the puffiness of face and oedema of eyelids

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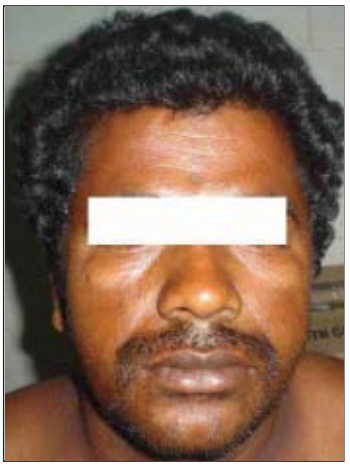

Figure 2: Patient with puffy face and oedematous eyelids

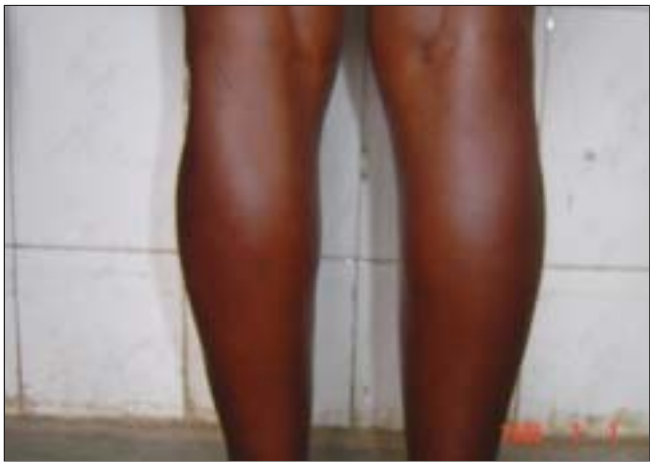

Figure 4: Resolution of calf muscle hypertrophy after 6 months of treatment

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