Original Article

A Rare Case Of Hoffman Syndrome Presented As Hypothyroid Myopathy

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Abstract

Hypothyroidism's neurological manifestations are uncommon as first symptoms, and they usually appear later in the disease's course. Muscle hypertrophy is a very uncommon occurrence in hypothyroid people 1. Hoffmann's syndrome is a kind of hypothyroidism that manifests as muscle stiffness and pseudohypertrophy in adults. In most cases of hypothyroid myopathy, laboratory tests reveal elevated levels of muscle enzyme. The electro physiological investigation may reveal signs that indicate myopathy, neuropathy, or a combined pattern of myopathy and neuropathy. Hormone replacement therapy 7 restores symptoms and normalises enzyme levels in the blood. Now, presenting a case of young age male presenting with hypothyroidism having features of calf muscle hypertrtrophy

Key Words : Hoffmann syndrome; pseudo hypertrophy of muscles;anti - thyroid peroxidase antibodies.

INTRODUCTION

Hypothyroidism is a common endocrine disorder characterised by fatigue, weight gain, cold sensitivity, constipation, melancholy, mental sluggishness, and muscle cramps. Myopathic alterations have been recorded in 30-80% of hypothyroidism patients, whereas muscular hypertrophy with muscle stiffness has been described in less than 10% of cases 4. Hoffmann's condition is a rare manifestation of hypothyroid-associated myopathy. Proximal muscle weakness, hypertrophies in the extremities, stiffness, muscle cramps, and spontaneous muscle soreness are all symptoms of this illness, which is linked to an increase in muscle enzymes. During hypothyroidism, these observations might be detected at any moment. The prognosis for Hofmann's syndrome is excellent. It has a very good response to hormone replacement therapy 5. As a result, in individuals with myopathy-like symptoms, the differential diagnosis should includeThe fact that the myopathy could be a sign of hypothyroidism can help with diagnosis and treatment. Hoffman's syndrome is an uncommon form of hypothyroid myopathy characterised by proximal muscle weakness and pseudohypertrophy 2. It was initially identified in an adult who had muscle stiffness and difficulties relaxing muscles following thyroid surgery in 1897.

CASE REPORT

History

A 20yr old male presented to opd with complaints of swelling of both calf muscles for 1 year(fig 1a & 1b) and pain in the both calf muscles after walking for long distance for 7 days and pain in the both calf muscles while walking on stairs upwards for 7days Pt is a known case of hypothyroidism and on irregular treatment

Fig1a

Fig 1b

Calf Muscle Hypertrophy Fig 1a (Posterior View)&Fig 1b(Lateral View)

ON EXAMINATION: patient is conscious;coherent,afebrile Pallor-absent; icterus- absent; clubbing-absent;cyanosis-absent;Lymphadenopathy -absent;pedal edema- absent Swelling is present in bilateral calf muscle region, no tenderness, no local rise of temperature

Systemic examination:

CVS: s1;s2 heard; no murmurs
RS :Bilateral air entry present; no added sounds
P/A: soft, Non tender, No organomegaly
CNS: NO focal neurological deficit

INVESTIGATIONS

Pt admitted in the ward and evaluated for certain investigations
Free T3: 2.43pg/ml; Free T4:0.54ng/dl, TSH:158.50miu/ml(increased)
Creatinine kinase -258u/L(increased)
Anti microsomal/thyroperoxidase antibody-125IU/ml(increased)
Anti thyroglobulin antibody- 63.7IU/ml(increased)
Others - Hb-17.4; Rbc count - 6.04M, S.uric acid -7.8mg/dl
Neurology opinion obtained in view of calf muscle hypertrophy; pt was advised to get Nerve conduction studies of 4 limbs and Electromyelogram & Neurology surgery opinion . Nerve conduction study of lower limbs are within normal limitsEMG of Gastronemius Rt and lt lateral head showed myogenic patternNeurosurgery opinion obtained ; pt was advised muscle biopsy.Muscle biopsy was suggestive of hypothyroid myopathy and showed swollen and pale muscle fibres with focal loss of striation.ANA PROFILE AND ENA PANEL (anti -RNP,anti- sm;anti-ssa(RO);anti-ssb(LA);anti-jo1;;scl70) found to be negative.B/L arterial & venous Doppler shows normal study
Pt is being treated with Thyronorm 200mcg for hypothyroidism
Pt is being treated with zyloric 100mg for hyperuricaemia
T3;T4 ;TSH,CPK values as follows comparison of before treatment to after treatment.
Calf muscle circumference also improved with in 5 months after treating with thyroxine(35cm to 31cm)

	T3 (ng/dL)	T4 (µg/dL)	TSH (µIU/mL)	CPK (IU/L)
Normal	70-204	3.2-12.6	0.4-4.2	24-195
Before treatment	8.6	0.2	429.51	5360
After 3 months	34.2	2.8	265.32	2356
After 5 months	86.2	5.6	34.66	271

TSH = Thyroid stimulating hormone, CPK = creatine phosphokinase

DISCUSSION

Hoffmann's Syndrome was originally identified in an adult who acquired muscle stiffness and difficulties relaxing muscles following thyroid surgery in 1897 5. Myopathy with pseudohypertrophy of muscles is a very rare and unique first manifestation. Thyroid insufficiency is caused by primary hypothyroidism in 95% of instances. Hashimoto's thyroiditis, an autoimmune disease. It is a type of chronic tyroiditis showing marked high levels of thyroid peroxidase antibodies (TPOAb) and thyroglobulin antibodies (TgAb), is the most common cause. TgAb and TPOAb antibodies are present in nearly all Hashimoto's thyroiditis patients 2. If only one test is ordered, the TPOAb is the best choice because it has a stronger affinity and is found in larger concentrations. The cause of pseudohypertrophy in Hoffman's syndrome is unknown and is a matter of discussion. An increase in connective tissue, as well as an increase in the size and quantity of muscle fibres, have been proposed as possible mechanisms 1. There is also a shift in muscle fibres from fast to slow twitching type II fibres aberrations in oxidative enzymatic activity, and hypertrophy due to glycosaminoglycan buildup. The tongue, arm, and leg muscles are the most commonly involved muscle groups. Following treatment with thyroid hormones, muscle hypertrophy and weakness will subside.
Thyroid myopathy causes elevated creatine phosphokinase levels (CPK), which can be very high in some patients (10-100 times higher than normal), but there is no link between CPK and weakness 7. CPK is the most accurate biochemical

indicator of myopathies. The treatment causes a gradual decrease in enzyme levels, which might take weeks, months, or even years. In hypothyroid myopathy, an electrophysiological examination may provide data that are consistent with neurogenic, myogenic, or a combination of these characteristics. Reduced duration and amplitude of motor unit potentials are EMG results that are consistent with a myogenic pattern. Entrapment neuropathies can be detected using nerve conduction studies (NCS) and axonal sensorimotor polyneuropathyOur patient had predominantly proximal muscular weakness and calf muscle hypertrophy. His CPK levels were four times higher than usual, and his TPOAb titers were likewise high. The EMG revealed right and left lateral head of gastrocnemius showed myotonic pattern, and the nerve conduction examination are with in normal limits . The patient was discharged from the hospital after being started on L-thyroxine (200 mcg once a day). After giving levothyroxine for 4 weeks ;patient had decreased creatinine kinase levels, improvement in muscle power and bulk of muscle reduced

CONCLUSION

To summarize, Hoffman syndrome is a rare condition initially presented as calf muscle hypertrophy it will be difficult to diagnose as similar features seen in other conditions such as Beckers muscular dystrophy , dychenne muscular dystrophy, focal myosotis & amyloidosis. As a result, increased awareness of this illness will lead to an accurate diagnosis. It's also worth noting that after hormone replacement medication, both s.muscle enzymes levels & symptoms return to normal.

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