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Hoffmann's syndrome: A rare and treatable hypothyroid myopathy in adults

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Abstract: ***Background:*** *The symptoms of myopathy due to hypothyroidism are proximal muscle weakness, delay in deep tendon reflexes and rarely (10%) development of muscle hypertrophy. Hoffman Syndrome is a adult pseudohypertrophic myopathy associated with hypothyroidism, There have not been many cases reported in India.*

Case Report: *A 30 year male presented with weight gain and all four limbs proximal muscles weakness, history of change in voice since 6 months. On examination tongue was hypertrophied on Neurological examination Motor system revealed muscle bulk increased over both calf and biceps, power in all four limbs is 4/5 with predominant proximal muscle weakness, with delayed ankle jerk and systemic examination was normal. On investigations TSH>100µIU/mL free T3-2.04ng/ml free T4-0.11µg/dL, AntiTPO>1000 Iu/ml CPK- 19481U/L, other investigations are normal. Treatment with 100 mcg thyroxin per day showed significant improvement in the patient. The follow-up values of TSH and CPK showed return to normal values after 3 months*

Conclusion: *Hoffmann's syndrome is rare condition its initial presentation with myopathic manifestations can raise diagnostic problems as calf muscle hypertrophy can present in diseases like Duchenne and Becker's muscular dystrophy, amyloidosis, and focal myositis. Therefore special awareness about this syndrome will lead to the correct diagnosis. It is also important to emphasize that both the symptoms and serum levels of muscle enzymes return to normal with hormone replacement therapy.*

Key words: *Hoffmann's syndrome; Hypothyroidism; pseudohypertrophy of muscles.*

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I. Introduction

Hoffmann's syndrome is a rare specific form of hypothyroid myopathy, which causes proximal muscle weakness, pseudo hypertrophy of muscles and delay in deep tendon reflexes. The muscular pseudo hypertrophy with muscle stiffness is reported in less than 10% of hypothyroid patients.[1] The hypothyroidism presenting with initial neurological manifestations is very unusual and rare. There have not been many cases reported in India.

II. Case Report

A 30 year old male presented with weight gain and Weakness all four limbs. Weakness is more in proximal muscles than distal, in the form of difficulty in getting up from sitting position and combing hair. History of dryness of skin and change in voice and slowness in speech were there for 6 months. There was no history of sensory involvement and bowel/bladder disturbance.

On examination, Pulse was 76/minutes and BP- 110/60 mmHg, dryness of skin present. Neurological examination showed normal higher mental functions and cranial nerves. Motor system examination showed muscle bulk increased over calf and biceps,[figure 1 & 2] power in all four limbs was 4/5 with predominant proximal muscle weakness with delayed ankle jerk. Sensory system examination was normal. Other systemic examination was normal.

Laboratory investigations revealed TSH>100mIU/mL, free T3- 2.04pg/ml, free T4- 0.11ng/dL, Anti-TPO >1000IU/ml, AST-202U/L, cholesterol-340mg/dl, triglycerides-142mg/dl, HDL-40.5mg/dl, LDL-249.6mg/dl, VLDL-28.4mg/dl, CPK- 19481U/L, serum electrolytes, RFT, complete blood picture, ECG and 2D-ECHO were normal. ANA-15 screening negative. Muscle biopsy showed no inflammation or muscular dystrophy. Nerve conduction velocities were normal. Electromyogram (EMG) showed no evidence of myotonia. Treatment with 100 mcg thyroxin per day showed significant improvement in the patient. The follow-up values of TSH and CPK showed return to normal values after 4 months and dose was reduced to half.

Figure 1: Photograph of the patient in the case report showing Hypertrophy of biceps muscles of both the upper limbs

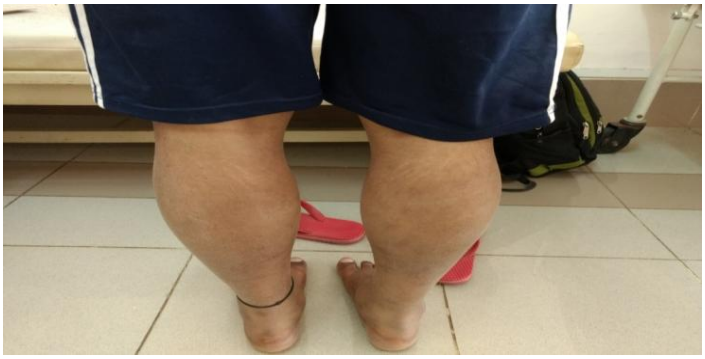

Figure 2: Photograph of the patient in the case report showing Hypertrophy of calf muscles of both the lower limbs

III. Discussion

Primary hypothyroidism accounts for 95% of the cases of thyroid insufficiency. The main etiology is Hashimoto's thyroiditis, an autoimmune chronic thyroiditis characterized by high levels of thyroid peroxidase antibodies (TPOAb) and thyroglobulin antibodies (TgAb). Both TgAb and TPOAb are found in almost 100% of patients with Hashimoto's thyroiditis.[2] The etiology of pseudohypertrophy in Hoffman's syndrome remains obscure and is a matter of discussion. It has been postulated that the mechanisms involved could include an increase in connective tissue, number of muscular fibers and hypertrophy due to accumulation of glycosaminoglycans. [3,4] The most common muscles groups involved are the tongue, arm and leg muscles. The muscle hypertrophy and muscle weakness will recede following treatment with thyroid hormones.

The creatine phosphokinase levels (CPK) will be elevated in thyroid myopathy and very high in some patients (10-100 times greater than the normal level). However, it has no correlation with weakness.[5] The CPK is the best biochemical marker of myopathies. The fall of the enzyme levels with the treatment occurs slowly, varying from weeks, months or even years. The nerve conduction studies (NCS) may show entrapment neuropathies and axonal sensorimotor polyneuropathy.[6, 7]

Our patient presented with predominant proximal muscle weakness and hypertrophy of calf muscles. His CPK levels were elevated 10 times the normal and TPOAb titers were also elevated. The patient was started on treatment and was discharged. On follow up after 4 weeks of treatment showed decreased CPK levels and improvement in muscle power with mild reduction in muscle bulk.[Table 1]

Table 1: Post Treatment Values Of TSH and CPK

	T3 (ng/ml)	**T4 (mg/dL)**	**TSH (mIU/mL)**	**CPK (IU/L)**
Normal	0.7-2.04	4.6-10.5	0.4-4.2	20-200
1 month	1.03	10.81	5.37	2225
4 months	0.71	9.6	0.1	692

TSH = Thyroid stimulating hormone, CPK = creatine phosphokinase

IV. Conclusions

Hoffmann's syndrome is very rare condition and its initial presentation with myopathic manifestations can raise diagnostic problems as calf muscle hypertrophy can present in other diseases like Duchenne and Becker's muscular dystrophy, amyloidosis, and focal myositis. Therefore special awareness about this syndrome will lead to the correct diagnosis. It is also important to emphasize that both the symptoms and serum levels of muscle enzymes return to normal with hormone replacement therapy.

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