International Journal of Advances in Medicine
Sriharsha T et al. Int J Adv Med. 2022 Dec;9(12):1236-1238
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: https://dx.doi.org/10.18203/2349-3933.ijam20223029

Case Report

Hoffman's syndrome-a rare neurological presentation of hypothyroid myopathy

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Received: 08 October 2022
Accepted: 03 November 2022

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ABSTRACT

Hoffman's syndrome is a rare form of hypothyroid myopathy, which causes proximal muscle weakness and pseudohypertrophy of muscles. It is a complication of untreated or uncontrolled Hypothyroidism.The neurological manifestations of hypothyroidism usually occur late in the course of disease. It is quite rare to present with neurological manifestations in the earlier stage of disease. Very few cases of Hoffman's syndrome were reported from India. Here we report a case of 27-year-old male who presented to the medicine OPD with initial symptoms of proximal muscle weakness, fatigue and calf muscle hypertrophy. On further evaluation patient was found to have hypothyroid myopathy in the form of Hoffman's syndrome.

Keywords: Hoffman's syndrome, Hypothyroidism, Pseudo hypertrophy

INTRODUCTION

Hypothyroidism is the most common endocrine disorder. The frequency of myopathy in hypothyroidism ranges from 30 to 80%. The common symptoms of myopathy due to hypothyroidism are proximal muscle weakness, muscle cramps, delay in deep tendon reflexes, and rarely, development of muscle hypertrophy.[1] The muscular hypertrophy with muscle stiffness is reported in less than 10% of hypothyroid patients. Hoffmann's syndrome is a rare specific form of hypothyroid myopathy, which presents with features of proximal muscle weakness, fatigue, muscle stiffness, hypertrophy of muscles and delayed deep tendon reflexes.[2]

CASE REPORT

A 27-year-old male presented with complaints of weakness in lower limbs and easy fatiguability since last one month. He also complained of difficulty in climbing stairs and getting up from squatting position. He also complained of frequent muscular cramps since one month. There was no history of muscle pain, involuntary movements, fever. There was no history of bladder or bowel involvement. There was no history of hypertension, diabetes, or any other chronic illness. There was no history of any prolonged drug intake. On examination he had enlarged tongue. His pulse was 54/min, BP was 120/70 mm Hg. There was no thyroid enlargement. His calf muscles were hypertrophied bilaterally (Figure 1) without tenderness and there was non-pitting oedema over the ankles. No hypertrophy of arm, thigh and other muscle groups was present. Cardiovascular, respiratory and abdominal examination were normal. Neurological examination revealed normal higher mental functions and cranial nerve examination. Motor examination revealed tone was normal in all the 4 limbs, mild proximal lower limb muscle weakness with power of grade 4/5 and there was delayed relaxation of bilateral ankle jerks. Sensory examination was normal.

Complete blood counts, liver and kidney function tests were normal. MRI Brain was normal. Nerve conduction study was normal. Electromyography showed small

amplitude myopathic motor unit potentials. Thyroid profile revealed FT3-1.59 pg/ml, FT4-0.21 ng/dl, TSH>100 mIU/L. Anti-thyroid peroxidase antibody was positive. Creatine phosphokinase was elevated to 900 IU//L. Serum Lactate dehydrogenase was elevated. Urine examination was normal and myoglobinuria was absent. Lipid profile was normal. The Patient was administered on tablet levothyroxine 100 mcg and later increased to 125 mcg. On routine follow up his symptoms of muscle weakness and fatiguabilty got improved.

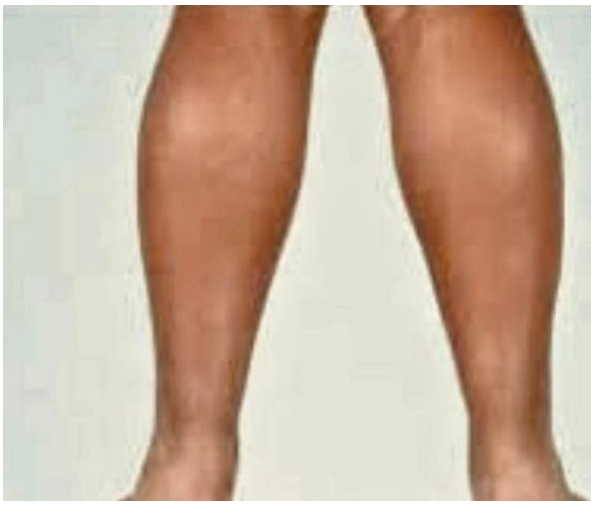

Figure 1: Bilateral calf muscle hypertrophy.

DISCUSSION

Hoffmann's Syndrome was first described by Hoffmann in 1897 in an adult who developed muscle stiffness and difficulty in relaxation of muscles after thyroidectomy. There are four subtypes of myopathy associated with hypothyroidism: (1) Kocher-Debre-Semelaigne syndrome, (2) Hoffman's syndrome, (3) atrophic form and (4) Myasthenic syndrome.

Thyroid hormones significantly influence cellular metabolism. The deficiency of these hormones results in significant impairment of normal cellular functions. The muscle involvement in hypothyroidism is caused by changes in muscle fibers from fast-twitching type II to slow-twitching type-I fibers which results in slowness of muscle contraction associated with hypothyroidism.[3,4] The cause of hypertrophy may be due to an increase in connective tissue and increase in size and number of muscular fibers. Pseudo hypertrophy of muscles is characteristic feature of Hoffman's syndrome which is due to deposition of glycosaminoglycans. The commonly involved muscles are tongue, leg muscles and arm. Delayed relaxation of deep tendon reflexes is due to impaired calcium sequestration by sarcoplasmic reticulum, which prolongs twitch duration. Mangaraj et al had published a case report similar to our case who presented with muscle weakness and stiffness, our case had presented with proximal muscle weakness of bilateral lower limbs and calf muscle hypertrophy and recovered significantly after the treatment had started.[1]

The laboratory investigations usually shows increased levels of muscle enzymes. CPK is best biochemical marker for myopathies and is increased significantly in hypothyroid myopathy however it is not correlated with muscle weakness.[5,6] The fall of the enzyme levels following the treatment were seen but may take weeks, months or years. Our case showed decreased CPK levels in two months. Muscle weakness and hypertrophy will improve with treatment of thyroid hormone replacement.

Differential diagnosis for calf muscle hypertrophy is muscular dystrophies such as Duchenne and Becker's dystrophy, sarcoid granulomas, amyloid and focal myositis. Our patient had elevated TSH with pseudohypertrophy of calf muscles so a diagnosis of Hoffman's syndrome was made.[7-9] The cause of hypothyroidism in Hoffman's syndrome is usually primary like Hashimoto thyroiditis. Secondary hypothyroidism rarely presents as Hoffman's syndrome. Muscle biopsy is usually not required to confirm the diagnosis.

Hoffman's syndrome usually carries good prognosis once the treatment is started and the muscle enlargement usually regresses with time.[10-12] Sometimes persistent hypertrophy of muscles may be seen even after the treatment.

CONCLUSION

Hypothyroidism is a very common endocrine disorder in which hypothyroid myopathy is commonly encountered in many of the cases. Hoffman's syndrome is a rare entity of hypothyroid myopathy. It occurs due to uncontrolled or untreated hypothyroidism and is reversible with proper treatment and management. Clinicians should be well aware of this rare manifestation of hypothyroidism to prevent the associated morbidity by avoiding misdiagnosis and subsequent delay in management. Serum TSH should be done for all the patients presenting with any degree of weakness and muscular hypertrophy to rule out Hoffman's syndrome as it has better prognosis when compared to other causes of weakness.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Mangaraj S, Sethy G. Hoffman's syndrome-A rare facet of hypothyroid myopathy. J Neurosci Rural Pract. 2014;5(4):447-8.
2. Achappa B, Madi D. Hoffmann's Syndrome- A Rare Form of Hypothyroid Myopathy. J Clin Diagn Res. 2017;11(5):OL01-2.
3. Dudekula A, Fernandes M, Mahale A, Sharma H. Hoffmann Syndrome: An Unusual Cause of Proximal Muscle Hypertrophy. Res J Radiol. 2017;4:1-4.

4. Ranaweerage R, Perera S, Gunapala A. Hoffmann syndrome; a rare form of hypothyroid myopathy. Sri Lanka J Med. 2021;30(1):104-8.
5. Sidibe EH, Diop AN, Thiam A, Diagne PM, Sarr A, Toure M et al. Hoffmann's syndrome in hypothyroid myopathy. Report of a case in an African. Joint Bone Spine. 2001;68:84-5.
6. Melmed S, Polonsky KS, Reed Larsen P, Kronenberg HM. Williams Textbook of Endocrinology. 12th ed. Philadelphia: W.B. Saunders Company. The thyroid gland. 2011;410.
7. Udayakumar N, Rameshkumar AC, Srinivasan AV. Hoffmann syndrome: Presentation in hypothyroidism. J Postgrad Med. 2005;51:332-3.
8. Vasconcellos LF, Peixoto MC, De Oliveira TN, Penque G, Leite AC. Hoffman's syndrome: Pseudohypertrophic myopathy as initial manifestation of hypothyroidism. Case report. Arq Neuropsiquiatr. 2003;61:851-4.
9. Deepak S, Harikrishnan, Jayakumar B. Hypothyroidism presenting as Hoffman's syndrome. J Indian Med Assoc. 2004;102:41-2.
10. Horak HA, Pourmand R. Endocrine myopathies. Neurol Clin. 2000;18:203-13.
11. Scott KR, Simmons Z, Boyer PJ. Hypothyroid myopathy with a strikingly elevated serum creatine kinase level. Muscle Nerve. 2002;26:141-4.
12. Barahona MJ, Mauri A, Sucunza N, Paredes R, Wägner AM. Hypothyroidism as a cause of rhabdomyolysis. Endocr J. 2002;49:621-3.
13. Udayakumar N, Rameshkumar AC, Srinivasan AV. Hoffmann syndrome: Presentation in hypothyroidism. J Postgrad Med. 2005;51:332-33.
14. Praveen KA, Aslam S, Dutta TK. Hoffmann's syndrome: A rare neurological presentation of hypothyroidism. Int J Nutr Pharmacol Neurol Dis. 2011;1:201-03.
15. Nalini A, Govindaraju C, Kalra P, Kadukar P. Hoffmann's syndrome with unusually long duration: report on clinical, laboratory and muscle imaging findings in two cases. Ann Indian Acad Neurol. 2014;17:217-21.
16. Senanayake HM, Dedigama AD, De Alwis RP, Thirumavalavan K. Hoffmann syndrome: a case report. Int Arch Med. 2014;7:2.

Cite this article as: Sriharsha T, Kumar AA, Raghav RJ, Reddy NM, Vikrannth V, Raghavan V et al. Hoffman's syndrome-a rare neurological presentation of hypothyroid myopathy. Int J Adv Med 2022;9:1236-8.