

Hoffmann's Syndrome- A Rare Form of Hypothyroid Myopathy

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Sir,

Varying manifestations of hypothyroidism are seen in clinical practice. Patients with hypothyroidism can present with mild fatigue or life-threatening myxoedema coma. Hypothyroidism may present as Hoffmann's Syndrome (HS), which is a rare form of hypothyroid myopathy. HS occurs in adults and is characterized by the presence of hypothyroidism, pseudohypertrophy of muscles and varying degrees of muscle weakness [1,2]. Few cases of HS have been reported from India. We hereby report a case of HS from Southern India.

A 45-year-old male, manual labourer by occupation presented to the Department of Medicine for evaluation of pedal oedema of nine months duration. He also gave history of swelling of face of nine months duration. On examination he had macroglossia and periorbital oedema. His pulse was 84/min, BP-120/70 mm Hg. There was no thyroid enlargement. His calf muscles were hypertrophied bilaterally with non-pitting oedema over the ankles [Table/Fig-1]. On neurological examination he had mild proximal lower limb muscle weakness (power 4/5) and deep tendon reflexes were sluggish. Systemic examination was normal.

Thyroid profile report revealed, Thyroid Stimulating Hormone (TSH) was 292.1 μ IU/ml (0.2-4.2), T4- 0.922 μ g/dl (5-14) and T3- 0.195 ng/ml (0.8-2). Creatine Phosphokinase (CPK) was 9594 U/L (85-170). Electromyography (EMG) was suggestive of a myopathic disorder. Thyroid peroxidase antibody was not done in view of financial constraints and our patient refused muscle biopsy. He was treated with levothyroxine 50 μ g. He was lost to follow up.

There are four subtypes of myopathy associated with hypothyroidism: Kocher-Debre-Semelaignesyndrome, HS, atrophic form and myasthenic syndrome [1]. Muscular pseudohypertrophy is a key feature of HS. Muscle pseudohypertrophy in HS is probably due to deposition of glycosaminoglycans and increased muscle fibre size [1,2]. The muscles commonly involved are the tongue, arm and leg muscles [2,3]. Calf muscles (gastrocnemius) are usually involved but generalized hypertrophy can occur [2,4]. When such an entity occurs in children it is known as Kocher-Debre-Semelaigne syndrome. Differential diagnosis for calf muscle hypertrophy are muscular dystrophy (Duchenne, Becker), sarcoid granulomas, amyloid and focal myositis [2,3,5]. The indexed patient had elevated TSH with pseudohypertrophy of calf muscles so a diagnosis of HS was made. The cause of hypothyroidism in HS is usually primary (Hashimoto thyroiditis) [3,6]. Secondary hypothyroidism can rarely present as HS [6]. Muscle biopsy is usually not required to confirm the diagnosis [2,5]. Muscle MRI may be help in the diagnosis of HS [4]. HS carries good prognosis once treatment is started and, the muscle enlargement usually regresses with time [2,4,5]. In some cases muscular hypertrophy may persist [7]. [Table/Fig-2] showing the similar case reports on HS published in literature [4,6-12].

Hypothyroidism is a common endocrine disease. Sometimes uncommon manifestation of a common disease is encountered in routine practice. HS is a classic example for this, so clinicians



[Table/Fig-1]: Hypertrophy of calf muscles of both the lower limbs.

Author	Year	Feature
Nalini A et al., [4]	2014	Long duration of illness on presentation and generalized muscular hypertrophy
Kumar H et al., [6]	2016	Hoffman's syndrome due to Sheehan syndrome
Qureshi W et al., [7]	2005	Persistence of muscular hypertrophy despite treatment
Ahmed GS et al., [8]	2014	Hoffman's syndrome with acute kidney injury
Kiran HS et al., [9]	2011	Typical Hoffman's syndrome
Mangaraj S et al., [10]	2014	Typical Hoffman's syndrome
Sundarachari NV et al., [11]	2013	Typical Hoffman's syndrome
Lee KW et al., [12]	2015	Typical Hoffman's syndrome
present case	2017	Typical Hoffman's syndrome

[Table/Fig-2]: Case reports about Hoffmann's syndrome published in the last decade.

should be aware of this rare disorder. Serum TSH must be done in all patients presenting with any degree of weakness and muscular hypertrophy to rule out HS as it has a better prognosis when compared to other causes of muscle hypertrophy.

REFERENCES

- [1] 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;61:851-54.
- [2] Udayakumar N, Rameshkumar AC, Srinivasan AV. Hoffmann syndrome: Presentation in hypothyroidism. *J Postgrad Med*. 2005;51:332-33.
- [3] 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.
- [4] 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.
- [5] Senanayake HM, Dedigama AD, De Alwis RP, Thirumavalavan K. Hoffmann syndrome: a case report. *Int Arch Med*. 2014;7:2.
- [6] Kumar H, Singh VB, Agrawal J, Meena BL. Chronic Sheehan syndrome presenting as pseudo-hypertrophic myopathy (Hoffman syndrome): A very rare combination. *Neurol India*. 2016;64(3):564-66.
- [7] Qureshi W, Hassan G, Khan GQ, Kadri SM, Kak M, Ahmad M, et al. Hoffmann's syndrome: a case report. *Ger Med Sci*. 2005;3:1-4.
- [8] Ahmed GS, Zaid HM, Moloney M. Hashimoto's thyroiditis presenting as Hoffman's syndrome, rhabdomyolysis and acute kidney injury. *BMJ Case Rep*. 2014;2014. pii: bcr2013203269.
- [9] Kiran HS, Sudharshana Murthy KA, Aparna AN. A young lady with swelling and stiffness of calf muscles. *Indian J Endocrinol Metab*. 2011;15:130-31.
- [10] Mangaraj S, Sethy G. Hoffman's syndrome - A rare facet of hypothyroid myopathy. *J Neurosci Rural Pract*. 2014;5:447-48.
- [11] Sundarachari NV, Sridhar A, Lakshmi VP. Rare yet treatable: Hypothyroid myopathy (Hoffman's syndrome). *J Dr NTR Univ Health Sci*. 2013;2:203-04.
- [12] Lee KW, Kim SH, Kim KJ, Kim SH, Kim HY, Kim BJ, et al. A Rare Manifestation of Hypothyroid Myopathy: Hoffmann's Syndrome. *Endocrinol Metab (Seoul)*. 2015;30:626-30.

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