CASE REPORT

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Hoffman's syndrome presenting as sole manifestation of hypothyroidism: A rare entity

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ABSTRACT

Myopathy is commonly seen in hypothyroidism. The most common symptoms include muscle cramps, weakness, and myalgia. Occasionally, hypertrophy of muscles may be seen which commonly involves muscles of tongue, thigh, and leg. The syndrome encompassing these clinical features is called as Hoffman's syndrome. It is commonly associated with symptoms suggestive of other systemic manifestations of hypothyroidism such as cold intolerance, constipation, and delayed deep tendon reflexes. Laboratory investigation abnormalities such as dyslipidemia, low voltage complex, and pericardial effusion are also commonly associated. Hoffman's syndrome as the only presentation of hypothyroidism in the absence of other symptoms and in the absence of other laboratory abnormalities is very rare. We present here an extremely rare case of Hoffman's syndrome as the only manifestation of hypothyroidism in the absence of other systemic and laboratory abnormalities.

Key words: Hoffman's syndrome, hypothyroidism, myopathy

INTRODUCTION

Hypothyroidism is an endocrine disorder which is commonly seen in clinical practice. It can present with various symptoms including cold intolerance, constipation, hoarseness of voice, and dryness of skin to name a few. On clinical examination and investigations, one can demonstrate dry skin, delayed deep tendon reflexes, dyslipidemia, low voltage complexes on electrocardiography (ECG), and pericardial effusion. Involvement of muscles in hypothyroidism is also common and manifest as muscle cramps, weakness, and myalgia.[1] Hypertrophy of muscles can also be seen which commonly involves the muscles of tongue, thigh, and leg. In adults, the syndrome which encompasses these signs and symptoms involving muscles is called as Hoffman's syndrome. It is a rare syndrome which was initially described by Hoffman in 1897.[2] Involvement of muscles occurs late in the course of the disease and is usually associated with other clinical features and laboratory investigations suggestive of hypothyroidism. Thus, it is very rare to find Hoffman's syndrome as the sole manifestation of hypothyroidism in the absence of other clinical features and laboratory abnormalities of hypothyroidism. We present here such a case of Hoffman's syndrome as the only manifestation of hypothyroidism.

CASE REPORT

A 40-year-old male, driver by occupation, presented with gradually increasing swelling of both the calf muscles for 1 year along with mild weakness in both the lower limbs. The patient also complained of cramps in the calf muscles along with stiffness which was aggravated on climbing stairs and prolonged walking. There was no history of suggestive of involvement of any other muscles in the body. The patient was in good health before this complains with no history of any diabetes mellitus, hypertension, or any other chronic illnesses. He had normal bowel and bladder habits and was not on any medications. He did not have any addictions. On examination, general appearance was normal. His pulse rate was 70/min, blood pressure-130/82 mmHg, and respiratory rate-12/min. There were no pedal edema, clubbing, or lymph nodes enlargement. Tongue examination did not reveal any abnormality. Systemic examination including cardiovascular, respiratory, and abdominal systems was found to be normal. On neurological examination, hypertrophy of bilateral calf muscles [Figures 1 and 2] was noted without any tenderness. Other groups of muscles were found to be normal. Weakness of proximal muscles of lower limbs was elicited with grade of 4. Deep tendon reflexes were normal. There was no sensory deficit. Other aspects of neurological examination were found to be normal. On investigations, his creatinine phosphokinase (CPK) was elevated - 875 IU/l. Levels of serum lactate dehydrogenase (LDH) were raised - 305 U/l. His thyroid-stimulating hormone level was raised significantly at 310 mIU/l. Free T3 level was 0.18 mcg/dl and free T4 level was 0.30 ng/dl, both of which were significantly low. On further evaluation, antithyroid peroxidase antibody was negative. His liver and kidney function tests, urine examination, and blood sugar levels were found to be normal. ECG and two-dimensional (2D) echocardiography were normal. Evaluation of lipid profile did not reveal any abnormality. Nerve conduction velocity study was found to be normal. Small amplitude myopathic motor unit potential was demonstrated on electromyography. Based on the clinical presentation, examination findings and laboratory investigations, diagnosis of Hoffman's syndrome was made and the patient was initiated on 100 mcg/day of levothyroxine. On follow-up of patient 1 month later, the weakness of lower limbs had improved to grade of 5. CPK

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Figure 1: Hypertrophy of bilateral calf muscles

Figure 2: Hypertrophy of calf muscle lateral view

levels had reduced to 175 U/l and LDH levels reduced to 105 U/l. The calf hypertrophy had only mild reductions on measurements. The patient was asked to follow-up regularly.

DISCUSSION

Thyroid hormone is an integral part of cellular metabolism regulating the various receptors and mitochondrial pathway. Deficiency of thyroid hormones in the body causes change in the muscle fiber types as well as their contractile properties. The path physiology involving the muscle hypertrophy is not completely elucidated. It is believed to be due to increase in both number and size of the muscle fibers as well as increase in the connective tissue.[3] Another mechanism postulated is that the hypertrophy is due to glycosaminoglycan accumulation which has been demonstrated on the histopathological examination of the muscle fibers which also appears swollen and pale with loss of normal striations.[4,5]

Various symptoms of hypothyroidism include weakness, fatigability, and muscular pain.[6] Proximal muscle weakness is commonly seen along with delayed deep tendon reflexes. Hypertrophy of the muscle is reported in <10% of hypothyroid patients. The commonly involved muscles include the tongue, thigh, leg, and arm. Elevation of muscle enzymes may be seen in hypothyroid myopathy; however, its levels bear no correlation with the weakness.[7] Severe form of hypothyroid myopathy can lead to a life-threatening complication of rhabdomyolysis which, in turn, can lead to acute kidney injury.[8] In majority of the reported cases of Hoffman's syndrome, other systemic features suggestive of hypothyroidism have been associated. Presentation of neurological manifestation as the first symptom of hypothyroidism is a rare entity as they occur late in the course of the disease. Mangaraj *et al.* had reported a case of Hoffman's syndrome as the presenting manifestation of hypothyroidism. However, in that case, hypertrophy of tongue muscle was present along with hypertrophy of calf muscles along with other systemic manifestations. Dyslipidemia, low voltage complex on ECG, and pericardial effusion on 2D echo, which are characteristics of hypothyroidism and were also present in the case.[9] Our case thus is extremely rare, as the only presentation of hypothyroidism in our case was Hoffman's syndrome in the absence of any other systemic features and the absence of other laboratory investigations which are typical of hypothyroidism.

CONCLUSION

Hoffman's syndrome is a rare presentation of a very common endocrine disorder, hypothyroidism. The positive aspect of this rare manifestation is that it is reversible with early diagnosis and treatment. Isolated hypertrophy of calf muscles without involvement of other muscles with the absence of other systemic features of hypothyroidism is extremely rare. Thus, the clinicians should be aware of this rare entity of a common disorder to initiate the therapy at the earliest.

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