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CASE REPORT

HOFFMANN'S SYNDROME: A CASE REPORT

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ABSTRACT: The neurological manifestations of hypothyroidism are very unusual to see as initial symptoms and they usually occur late in the course of disease. Hoffmann syndrome is a rare form of hypothyroid myopathy in adults characterized by presence of muscle weakness, stiffness and pseudohypertrophy. We present a case of 27 years old male presenting with features of proximal muscle weakness of both upper and lower limbs with hypertrophy of calf muscles. His TSH and CPK were markedly raised and EMG showed myopathic disorder. We report this case because of its rarity.

KEYWORDS: Hypothyroidism, Hoffmann's syndrome, Pseudohypertrophy.

INTRODUCTION: The most commonly observed symptoms of myopathy due to hypothyroidism are proximal weakness, cramps, painful muscles, myxoedema on percussion, delay in deep tendon reflexes and development of muscle hypertrophy.[1-3] Severity of myopathy generally correlates with the duration and the degree of thyroid hormone deficiency.[4,5] Serum CK (creatine kinase) elevation is usually observed even in the absence of overt muscle weakness.[4,6] .Although muscular symptoms may occur in many patients with hypothyroidism, muscular hypertrophy with muscle stiffness is reported in less than 10% of the patients.[1] The adult and childhood forms characterized with increased muscle volume, slow movements and stiffness are known as Hoffmann's syndrome and Debre Kocher Semelaigne respectively.[1,5]

CASE REPORT: A 27 years old male presented with history of weakness and fatigue of all four limbs and difficulty in getting up from squatting position and doing work above the shoulder level for last six months. There was no history of sensory involvement, bowel/bladder disturbance.

The clinical examination revealed mild periorbital puffiness, large tongue, and dry coarse skin. His pulse rate was 56/minute and blood pressure was 120/78 mmHg. Neurological examination showed normal higher mental functions and cranial nerves. Motor system examination revealed muscle power of 4/5 in all four limbs with predominant proximal muscle weakness, normal muscle tone with generalized hyporeflexia and delayed relaxation of ankle jerk. There was hypertrophy of calf muscles of both lower limbs (fig 1). His sensory, cerebellar, bowel and bladder functions were normal.

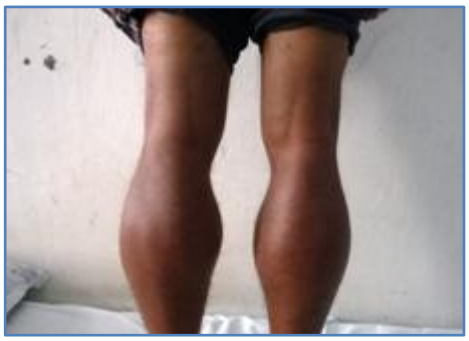

Fig. 1: Showing bilateral hypertrophy of calf muscles

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The laboratory examination revealed Hemoglobin of 12.9 gm/dL, serum creatinine of 0.8 mg/dL, Na 139 mmol/L, K 4.0 mmol/L, blood sugar fasting 86mg/dL. His Liver function tests revealed raised AST of 126 IU/L (5 – 40 IU/L), Lipid profiles showed raised cholesterol of 292 mg/dL (150-200 mg/dL), LDL 192 mg/dL and Triglycerides 290 mg/dL (60 – 160 mg/dL). His Creatine phospho kinase (CPK) was markedly raised to 14810 IU/L (24 – 170 IU/L) and also was LDH level of 442 U/L (140 – 280 U/L). Thyroid function test was abnormal with low free T4 0.43 ng/dL (0.89 – 1.76 ng/dL) and raised TSH of 153.62 mIU/mL (0.55 – 4.78 mIU/mL). His Anti TPO antibody was > 1200 IU/L. Electromyography (EMG) showed myopathic motor unit potentials (MUAPs) with small amplitude, duration and early recruitment in the proximal muscles suggestive of myopathic disorder patient didn't give consent for muscle biopsy.

He was treated with levothyroxine 100 microgram and he is presently doing well on 28 days follow up with improvement in muscle power and reduction in calf muscle bulk.

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.[6] It is characterized by muscle weakness and muscle (pseudohypertrophy).[7] Patients present with muscle cramps, muscle stiffness, weakness, hyporeflexia and delayed deep tendon reflexes.[8] Muscle enlargement (pseudohypertrophy) is very rare and its etiology remains controversial.[9] Calf muscles (gastrocnemius) are commonly involved.[10] The thigh, arm, tongue and forearm muscles are involved to a lesser extent.[10] Postulated mechanisms for muscle pseudohypertrophy include increase deposition of Glycosaminoglycans, increase muscle fiber size and number.[9,10] Hypothyroidism is associated with change in muscle fiber type from fast twitch type II to slow twitch type I and alteration of oxidative muscle enzyme activity with decreased calcium ATPase activity of fast twitch type II fibers leading to delayed relaxation.[7,10]

CPK levels are elevated in 70 – 90% patients of hypothyroidism, but do not correlate with severity of weakness. Other enzymes like AST, LDH and aldolase are also elevated. Our patient also had elevated CPK, AST and LDH. The electrophysiological study in hypothyroid myopathy may show findings compatible with neurogenic, myogenic, or a mix of those patterns. The EMG findings compatible with myogenic pattern are diminished duration, and amplitude of motor unit action potentials (MUAP).[8]

Hoffmann syndrome carries favorable prognosis once hormone replacement is instituted and most of the symptoms regress slowly with time, including the muscle enlargement. The decline of the muscle enzyme levels occurs slowly, varying from weeks, months or even years with the treatment.[8, 11] Electrophysiological findings may persist in some patients despite improvement of symptomatology and muscle enzymes.[8]

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