Hoffman’s syndrome – A rare facet of hypothyroid myopathy

Sir,
Hoffman’s syndrome is a specific, rare form of hypothyroid myopathy, which causes proximal weakness and pseudohypertrophy of muscles. It was first described in 1897 in an adult who developed muscle stiffness and difficulty in relaxation of muscles after thyroidectomy.[1] Similar presentation in children with cretinism is referred as Kocher–Debré–Sémélaigne syndrome.[2] We discuss a case of hypothyroid male with this unusual and rare presentation.

A 45-year-old male presented with complaints of weakness in lower limbs and easy fatigability for last 8 months. He had difficulty in getting up from squatting position and climbing stairs. He also complained of frequent muscular cramps and stiffness and pain in muscles. 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 was found to have periorbital puffiness and mild pedal edema. He also had a large tongue and generalized ichthyosis. He had a pulse rate of 56/min and his blood pressure was 140/90 mm Hg. Evaluation of cardiovascular and respiratory systems did not reveal any significant abnormality. Positive findings from neurological evaluation included bilateral proximal muscle weakness in lower limbs with grade 4 power. There was hypertrophy of bilateral calf muscles (gastrocnemius) without any associated tenderness [Figures 1 and 2]. No hypertrophy of thigh, arm, or any other muscle group was noted. There was classical delayed relaxation of bilateral ankle jerks. Laboratory investigation revealed a T3 level of 0.35 ng/ml (normal range: 0.60-1.81 ng/ml), T4 level of 1.54 μg/dl (normal range: 4.5-12 μg/dl), and the activity of thyroid stimulating hormone (TSH) to be 125 μIU/ml (normal range: 0.3-5.5 μIU/ml). Anti-thyroid peroxidase (anti-TPO) antibody was positive. Creatine phosphokinase (CPK) level was 1050 IU/l (normal level: <170 U/l). Serum lactate dehydrogenase (LDH) level was slightly elevated at 235 U/l (normal range: 90-185 U/l). Hemogram revealed mild anemia. Blood sugar and liver function tests were essentially normal. Blood urea and serum creatinine were normal. Urine examination was normal and myoglobinuria was absent. Lipid profile evaluation showed hypercholesterolemia (240 mg/dl) and hypertriglyceridemia (180 mg/dl). Electrocardiography (ECG) showed low-voltage complexes and sinus bradycardia. Two-dimensional echocardiography showed mild pericardial effusion with normal systolic and diastolic functions. Nerve conduction study was normal. Electromyography showed small-amplitude myopathic motor unit potential. Based on the above findings, a diagnosis of Hoffman’s syndrome was made. The patient was administered a starting dose of 100 μg/day of levothyroxine which was later escalated to 125 μg/day. On a routine follow-up later, his symptoms had improved though complete resolution of symptoms had not taken place. The calf hypertrophy, however, still persisted.

The neurological manifestations of hypothyroidism usually occur later, and it is unusual that they are found as initial symptoms.[3,4] Muscular symptoms are common in hypothyroid patients (varying from myalgia, weakness, stiffness, cramps, and easy fatigability in 30-80% of the patients).[3,5] The common symptoms
Hypothyroidism is a very common endocrine disease and clinicians should be aware of this atypical and rare presentation of hypothyroid disease spectrum. Hoffman’s syndrome represents those few forms of myopathy that completely reverse on prompt therapy and, hence, has a good outcome.

Swayamsidha Mangaraj, Ganeswar Sethy
Department of Internal Medicine, M. K. C. G. Medical College, Brahmapur, Odisha, India

Address for correspondence:
Dr. Swayamsidha Mangaraj,
Department of Internal Medicine, M. K. C. G. Medical College, Brahmapur - 760 004, Odisha, India.
E-mail: drsmangaraj@gmail.com

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