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OBJECTIVES

Hoffman's Syndrome is a rare form of hypothyroid myopathy characterized with muscle hypertrophy, stiffness and weakness. In most of the cases, the level of muscular enzymes is elevated, with no relation to the severity of the myopathic symptoms. The hormonal replasman is started, the myopathy could be reversible with good prognosis.

METHODS

43 years-old woman admitted to our hospital with a complaint of fatigue, progressive muscular weakness with muscular cramps and myalgia started three months before. Thyroid gland examination was normal. On neurological examination she had proximal and lower limb muscle weakness (3/4) and hyporeflexia. Her calf muscles was hypertrophic. Laboratorial investigation reveals (Table 1): increased serum levels of muscular enzymes, dislipidemia, severe hypothyroidism. Electromyography (EMG) muscles revealed low amplitude and short duration motor unit action potentials (MUAPs) with early recruitment suggestive of a myopathic disorder. The patient was diagnosed of having severe hypothyroidism with Hoffman syndrome. Oral L -thyroxine treatment was started (100mcg/ day) and dose was elevated 150 mcg/ day after two week later. After one months therapy her hypothyroid symptoms reduced, pseudohypertrophy of the calf muscles regressed, muscle enzymes were reduced.

Table 1. Laboratorial exams.

Period	TSH	ST4	TG	CHOL	CPK	LDH	AST
Before Treat	>150	0,2	622	250	4267	621	140
After Treat (one week)	89	0,69	421	198	1588	499	54
After Treat (one Months)	3,64	1,4	191	158	278	341	23

TSH: (0.5. 5.6mUI/mL); Free thyroxine(0.88-1.72 ng/dl)

TG: (50-200mg/dl Cholesterol(0-200 mg/dl).

CPK:(29-200 U/L,LDH (125-243U/L);

AST:(5-34 U/L)

RESULTS

Hoffman syndrome is a rare presentation of hypothyroid myopathy. The characteristic features of Hoffman's syndrome include localized or generalized hypertrophy of muscles in addition to muscle weakness, stiffness, cramps and pain as compared to the classic symptoms of hypothyroidism. The laboratorial investigation usually shows increased levels of muscular enzymes. The creatinophosphokinase (CPK) is the best biochemical marker of myopaties. Mild to moderate elevation of serum CPK level is seen in 70-90% patients with hypothyroidism indicative of muscle involvement but does not correlate with the severity of weakness. The clinical presentation and biochemical features might make it a bit difficult for the physician to differentiate it from polymyositis or muscle dystrophies. Regarding the association and etiology of Hoffman's syndrome autoimmune thyroiditis is commonly seen among these patients. Patients have anti-thyroid peroxidase and anti-thyroglobulin antibodies positive in >90% of cases as seen in our case as well.

CONCLUSIONS

Hoffman Syndrome has good prognosis if diagnosed earlier and treated appropriately. This case report shows that Hoffman syndrome, though a rare presentation of hypothyroidism has a good prognosis with timely diagnosis and appropriate management. In the differential diagnosis of myopathy with pseudohypertrophy, Hoffmann's syndrome should be considered. It is an infrequent cause of myopathy, with good prognosis.

References

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