

Short report

Hypothyroidism with true myotonia

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SUMMARY A patient with subclinical hypothyroidism who presented with true myotonia is described. There was no evidence that either he or members of his family had dystrophia myotonica or myotonia congenita. Treatment with thyroxine resolved his symptoms completely.

There are two recognised circumstances in which myotonia is seen in thyroid disease (Pearce and Aziz, 1969). Firstly, *clinical pseudomyotonia* involving slow contraction and relaxation of the muscles is seen in severe hypothyroidism and is one of the features of the Hoffmann syndrome. Secondly, hypothyroidism may reveal *true myotonia* in patients who suffer from dystrophia myotonica (Brumlik and Maier, 1972) or myotonia congenita (Jarcho and Tyler, 1958). Pseudomyotonia and myotonia are readily distinguishable by electromyography (Waldsterm *et al.*, 1958). Except in the setting of dystrophia myotonica or myotonia congenita, true myotonia has not been recognised as a feature of hypothyroidism. We now report a patient with subclinical hypothyroidism (Evered *et al.*, 1973) who had true myotonia, proven electromyographically, and who showed complete resolution after treatment with thyroxine. There was no evidence, either clinically or on electromyography, of myotonia in his family.

Case report

A 42 year old driver presented in March 1976 with disabling muscle stiffness. During the winter of 1974-75, while working out of doors on night shift, he developed episodes of muscle stiffness particularly at the beginning of movement. These resolved by early spring of that year but recurred in January 1976, and progressed to such an extent that by March he was unable to work. He complained that he could not initiate walking though after several steps his movements became easier. Hand movements, particularly grip, were difficult, and he noticed slowness in relaxation. He

had occasional cramping pains in the forearms and legs, and after a hiccough he was aware of the slow relaxation of his abdominal muscles. Direct questioning elicited no symptoms of hypothyroidism or other disease. There was no family history of muscle, endocrine, or autoimmune disease.

EXAMINATION

On examination he was a well-built man without evident stigmata of hypothyroidism. He had a smooth, non-tender goitre of moderate size without audible bruit. He was in sinus rhythm at 92 beats per minute, and his blood pressure was 140/80 mmHg. All his limb muscles were bulky and woody, and there was obvious clinical myotonia on attempted relaxation of the grip of both hands. However, there was no percussion myotonia or myoedema, and his muscle stretch reflex time was not prolonged. No fasciculation was observed. Muscle power and co-ordination were normal, and sensation was intact in all modalities.

LABORATORY INVESTIGATIONS

Serum creatine kinase was 330 U/l (0-50 U/l), serum thyroxine 13 nmol/l (60-150 nmol/l), thyopac-3 136 (92-117), free thyroxine index less than 10 (50-160), thyroid stimulating hormone 25.4 mU/l. Thyroid cytoplasmic and gastric parietal cell antibodies were present. Thyroid tanned red cell antibody titre was greater than 1/10 000 and thyroid microsomal antibody titre was 1/1600. Electromyography showed an increase in insertional and spontaneous activity in both the left deltoid and abductor pollicis brevis muscles. In the deltoid "bizarre high frequency discharges" (Simpson, 1969), formerly called "pseudomyotonic potentials," were recorded. Furthermore, true myotonic discharges were seen clearly (Figure). Muscle biopsy of the right deltoid was normal.

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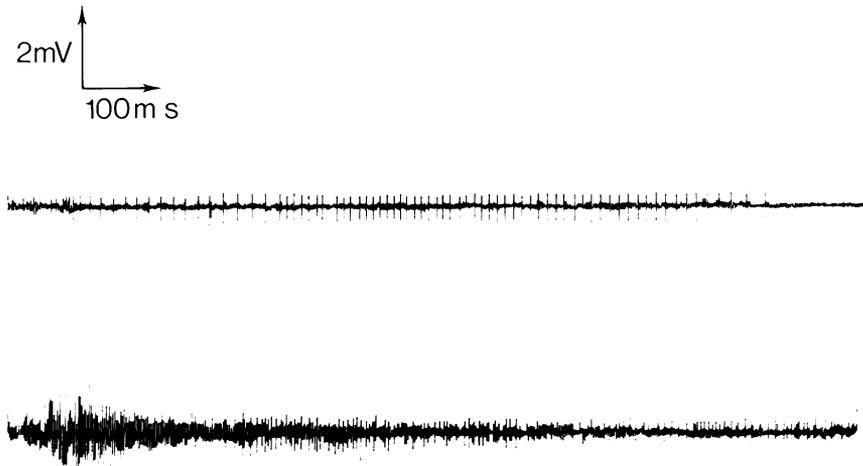


Figure Initial electromyogram to show bizarre high frequency discharge (upper tracing) and true myotonic potential (lower tracing).

FAMILY STUDIES

The patient had three children all of whom were clinically normal and had normal serum creatine kinase, thyroxine, and thyroid stimulating hormone levels. Serum thyroid antibodies were absent, and electromyography was normal.

PROGRESS

The patient was treated with L-thyroxine but after one month showed no improvement, and had quite profound stiffness and muscle pain. After a further four weeks there was a striking improvement. He no longer experienced muscle stiffness or cramps, and had lost 12 kg in weight. At the time his serum creatine kinase was 43 U/l, thyroxine 108 nmol/l, and thyroid stimulating hormone 5 mU/l. The electromyogram no longer contained myotonic potentials or "bizarre high frequency discharges."

Discussion

The syndrome of muscular hypertrophy associated with delayed muscle relaxation was first described in athyreotic cretins by Kocher (1892), and later by Hoffmann (1897) in an adult who had undergone thyroidectomy. Hoffman differentiated the impaired relaxation of grip seen in his patient, now referred to as pseudomyotonia, from true myotonia by its failure to improve after several contractions. Subsequently percussion myoedema was described in Hoffman type patients (Wilson and Walton, 1959) and later authors recorded elevation of serum creatine kinase in such patients

(Graig and Ross, 1963; Pearce *et al.*, 1964). Pearce and Aziz (1969) described fibre hypertrophy, focal necrosis, and deposition of mucopolysaccharide on muscle biopsy. Waldsterm *et al.* (1958) demonstrated "bizarre high frequency discharges" on electromyography in such patients, but subsequently Wilson and Walton (1959) reported the absence of both the characteristic after-discharge of myotonia and bizarre insertional discharges during contraction. The constellation of features described above, when encountered in athyreotic cretins, has been named the Debre-Semelaigne-Kocher syndrome (Debre and Semelaigne, 1935). When encountered in hypothyroid adults it is usually referred to as the Hoffman syndrome, and it resolves with replacement therapy. We doubt whether this distinction now serves any useful purpose since the two syndromes are identical.

We emphasise that we are only considering myotonia in association with hypothyroidism. There are other neuromuscular syndromes occurring with hypothyroidism, and these have been reviewed elsewhere (Wilson and Walton, 1959; Astrom *et al.*, 1961; Sahay *et al.*, 1965; Norris and Panner, 1966; Pearce and Aziz, 1969; Golding, 1970; Takamori *et al.*, 1972). The main purpose of the present report is to discuss the occurrence of true myotonia together with pseudomyotonia in a patient with Hoffmann's syndrome. To date, the finding of true myotonia in the presence of hypothyroidism has been restricted to patients with dystrophia myotonica or myotonia congenita. In these patients, unlike those with classical Hoffmann's syndrome, there is improvement in

the ability to relax after several attempts, a myotonic discharge on electromyography, and usually a positive family history. Treatment with L-thyroxine may help to improve symptoms but true myotonic potentials persist and herald the reappearance of the underlying neuromuscular disorder.

Unlike those cases of dystrophia myotonica referred to above, our patient's symptoms resolved fully with thyroxine, and his electromyogram returned to normal. The other members of his family are normal, and we have found no evidence that he has dystrophia myotonia or myotonia congenita. We, therefore, believe that he demonstrates that true myotonia can occur solely as a result of hypothyroidism.

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References

- Astrom, K. E., Kugelberg, E., and Muller, R. (1961). Hypothyroid myopathy. *Archives of Neurology (Chicago)*, **5**, 472-482.
- Brumlik, J., and Maier, R. J. (1972). Myxedema and myotonic dystrophy. *Archives of Internal Medicine*, **129**, 120-122.
- Debre, R., and Semelaigne, G. (1935). Syndrome of diffuse muscular hypertrophy in infants causing athletic appearance. *American Journal of Diseases of Children*, **50**, 1351-1361.
- Evered, D. C., Ormston, B. J., Smith, P. A., Hall, R., and Bird, T. (1973). Grades of hypothyroidism. *British Medical Journal*, **1**, 657-662.
- Golding, D. N. (1970). Hypothyroidism presenting with musculoskeletal symptoms. *Annals of Rheumatic Diseases*, **29**, 10-14.
- Graig, F. A., and Ross, G. (1963). Serum creatine phosphokinase in thyroid disease. *Metabolism, Clinical and Experimental*, **12**, 57-59.
- Hoffmann, J. (1897). (Cited Wilson and Walton, 1959). Weiterer Beitrag zur Lehre von der Tetanie. *Deutsche Zeitschrift für Nevenheilkunde (Berlin)*, **9**, 278-290.
- Jarcho, B. W., and Tyler, F. H. (1958). Myxedema pseudomyotonia and myotonia congenita. *Archives of Internal Medicine*, **102**, 357-366.
- Kocher, T. (1892). (Cited Norris and Panner, 1966.) Zur Verhütung der Cretinismus und Cretinoider Zustände nache neuen Forschungen. *Deutsche Zeitschrift für Chirurgie (Berlin)*, **26**, 556-626.
- Norris, F. H., and Panner, B. J. (1966). Hypothyroid myopathy. *Archives of Neurology (Chicago)*, **14**, 574-589.
- Pearce, J. M. S., Pennington, R. J., and Walton, J. N. (1964). Serum enzyme studies in disease. *Journal of Neurology, Neurosurgery, and Psychiatry*, **27**, 96-99.
- Pearce, J. M. S., and Aziz, H. (1969). The neuromyopathy of hypothyroidism. Some new observations. *Journal of the Neurological Sciences*, **9**, 243-253.
- Sahay, B. M., Blendis, L. M., and Greene, R. (1965). Relation between myasthenia gravis and thyroid disease. *British Medical Journal*, **1**, 762-765.
- Simpson, J. A. (1969). Terminology of Electromyography. Report of a sub-committee of the International Federation of Societies for Electroencephalography and Clinical Neurophysiology. *Electroencephalography and Clinical Neurophysiology*, **26**, 224-226.
- Takamori, M., Gutmann, L., Crosby, T. W., and Martin, J. D. (1972). Myasthenic syndromes in hypothyroidism. *Archives of Neurology (Chicago)*, **26**, 326-335.
- Waldsterm, S. S., Bronsky, D., Schrifter, H. B., and Oester, Y. T. (1958). The electromyogram in myxedema. *Archives of Internal Medicine*, **101**, 97-102.
- Wilson, J., and Walton, J. N. (1959). Some muscular manifestations of hypothyroidism. *Journal of Neurology, Neurosurgery, and Psychiatry*, **22**, 320-324.