Two further cases are reported here, both having malignant hypertension, one responding to nephrectomy and one not.

I am grateful to Dr. Horace Joules and Dr. R. A. J. Asher for permission to report cases admitted under their care. I am also grateful to Dr. J. H. O. Earle for his help in the pathological examination of these cases.

REFERENCES

Abeshouse, B. S. (1941). *Surgery*, **9**, 942 ; **10**, 147.
Barker, N. W. (1951). *Med. Clin. N. Amer.*, **35**, 1041.
Braasch, W. F., Walters, W., and Hammer, H. J. (1940). *J. Amer. med. Ass.*, **115**, 1837.
Butler, A. M. (1937). *J. clin. Invest.*, **16**, 889.
Crabtree, E. G., and Chaset, N. (1940). *J. Amer. med. Ass.*, **115**, 1842.
Everett, H. S. (1940). *Urol. Cutan. Rev.*, **44**, 557.
Goidsenhoven, F. van (1947). *Brux.-méd.*, **27**, 321.
Goldblatt, H., Lynch, J., Hanzal, R. F., and Summerville, W. W. (1934). *J. exp. Med.*, **59**, 347.
Langley, G. J., and Platt, R. (1947). *Quart. J. Med.*, **16**, 143.
Leadbetter, W. F., and Burkland, C. E. (1938). *J. Urol.*, **39**, 611.
Longcope, W. T. (1937). *Ann. intern. Med.*, **11**, 149.
Mulholland, S. W. (1940). *Bull. N.Y. Acad. Med.*, **16**, 244.
Nesbit, R. M., and Ratliff, R. K. (1940). *J. Urol.*, **43**, 427.
Ostry, H. (1948). *Canad. med. Ass. J.*, **58**, 498.
Pickering, G. W., and Heptinstall, R. H. (1953). *Quart. J. Med.*, **22**, 1.
Schroeder, H. A., and Steele, J. M. (1939). *Arch. intern. Med.*, **64**, 927.

A CASE OF MYOTONIA CONGENITA WITH HYPOTHYROIDISM

BY

A. F. WADE, M.B., B.S.

Late House-Physician, Medical Unit, University College Hospital, London

Symptoms and signs suggestive of myotonia have been described in association with hypothyroidism. Since these are relieved by treatment of the thyroid condition, they have been regarded as part of the hypothyroid state (Hoffmann, 1896 ; Debré and Sémelaigne, 1935).

This paper records a case in which myotonia congenita (Thomsen's disease) and hypothyroidism were present simultaneously and in which the myotonia was not completely relieved even when the patient was made hyperthyroid.

Myotonia congenita is a hereditary disease of muscle characterized by prolonged involuntary contraction following a voluntary contraction (active myotonia) or mechanical stimulation (mechanical myotonia). The condition was first described by Leyden (1874), and Thomsen later published his classical account (Thomsen, 1876).

Case Report

A man aged 20 was referred to University College Hospital in September, 1954, for assessment of his myxoedema, diagnosed nine months earlier. He stated that at the age of 5 he had suffered from stiffness in the legs. This did not greatly trouble him and disappeared after two years. At 12 years the stiffness reappeared. It was more severe and extensive than previously. His main difficulty was in starting a movement, especially if sudden, owing to stiffness of the muscles. The calves and forearms were particularly affected. He noticed that the muscle became prominent and hard and was occasionally painful. After performing a movement repeatedly the stiffness wore off. He was sometimes unable to move a limb for some seconds. The symptoms were worse in the mornings and when cold.

His athletic abilities, previously excellent, fell off markedly, and by the time he was 13 he had given up sport completely, though doing well at his studies. At 14 he was becoming slower physically and mentally. He gained weight rapidly and complained of lethargy and difficulty in concentrating, and his work deteriorated, so that instead of being near the top he was now near the bottom of his class.

At 18 he joined the Army and was constantly in trouble on account of slowness and clumsiness. On a cold morning he was almost incapacitated. While on sentry duty he once "went stiff all over" and fell to the ground when an officer unexpectedly appeared.

He next complained of frontal headaches, constipation, and occasional double vision. On investigation at that time he was puffy about the eyes, his skin was cold and dry, his pulse 55, his basal metabolic rate −25%, and serum cholesterol 470 mg. per 100 ml. A diagnosis of myxoedema was made and he was treated with thyroid. His weight decreased, the headaches became less severe, and the stiffness was less marked.

When first seen at University College Hospital there was, in the absence of full details, some doubt about the diagnosis, and thyroid therapy was discontinued. A month later, on admission, there was slight puffiness around the eyes, his skin was cold and dry, his temperature 97° F. (36.1° C.), his voice low-pitched, his pulse 44, and blood pressure 130/80 mm. Hg. The thyroid was not clinically enlarged. The muscles were well developed, the calves appearing abnormally bulky; tone was normal and there was no wasting. The arm-, knee-, and ankle-jerks were sluggish, and direct percussion of the calf muscles and forearm extensors produced a contraction lasting about five seconds with marked grooving at the site of percussion. Following a firm handshake there was considerable delay in relaxation of the grip.

Investigations.—Plasma cholesterol was 275 mg. per 100 ml. (later 370 mg.). His basal metabolic rate was −30% ; radioactive iodine uptake at 24 hours was 5% of total dose. The electrocardiogram showed a rate of 39 with normal voltage curves. The blood sodium, potassium, chloride, calcium, magnesium, and inorganic phosphate were all normal. His intelligence quotient was 100.

Electromyography (Dr. W. D. Fletcher).—"The strength-duration curves of the right extensor carpi radialis were quite normal, although the type response was rather sluggish. Electromyography with needle electrodes, however, revealed trains of high-frequency oscillations on insertion of the needle and on mechanical stimulation of the muscle. These findings, taken together, are typical of myotonia congenita."

Improvement, objective and subjective, followed the administration of quinine. There was no urinary creatine. The creatine tolerance was high (71% retained) and administration of creatine had no observable effect on the myotonia. These findings are in keeping with other recorded observations in Thomsen's disease (Milhorat and Wolff, 1938 ; Poncher and Wade, 1938).

While under investigation the patient became more myxoedematous and his myotonia more marked. Thyroid therapy resulted in an improvement in both conditions. Correction of the hypothyroidism, however, was not accompanied by abolition of the myotonia, although active myotonia became less obvious.

He was discharged from hospital on December 3, 1954, taking 0.4 mg. of sodium L-thyroxine daily. Six weeks later he was readmitted for observation. He stated he was physically and mentally more agile, but complained of perspiring hands and palpitations. He had been less troubled by stiffness.

On examination he had lost a stone (6.4 kg.) in weight since taking thyroxine. His pulse was 68 a minute and regular. Active myotonia was not observed, the grip relaxing rapidly, but there was still marked mechanical myotonia, particularly of the calf and forearm extensors. His basal metabolic rate was now +53% and plasma cholesterol 247 mg. per 100 ml. No urinary creatine was detected. Electromyography showed no change, being still typical of myotonia ; the electrocardiogram showed no significant change. Slit-lamp examination revealed no lens opacities.

Histology of calf-muscle biopsy showed well-developed muscle fibres with no muscle atrophy. In spite of the weight

loss, arm and leg volumes showed no significant change since his first admission. He was discharged on 0.2 mg. of sodium L-thyroxine daily.

Family History.—The mother, a maternal aunt, and her daughter all gave a history of myotonia and were found on clinical and electromyographic examination (Dr. Fletcher) to exhibit myotonia. The mother's father had had an operation for a cataract at the age of 74. Another sister of the mother had suffered from " Derbyshire neck."

Discussion

The history and findings are so characteristic of myotonia congenita and of hypothyroidism as to leave no real doubt that this patient suffers from two separate conditions.

The early age of onset of myotonia and the prolonged muscular contractions, worse in the morning, when cold, and becoming less marked on repeated movement, are typical. Difficulty in relaxing a grip following a handshake is a usual complaint. His falling " muscle bound " on being suddenly surprised recalls the behaviour of myotonic goats who suffer from a condition which completely resembles myotonia congenita in man (Levin, 1934 ; Brown and Harvey, 1939). Further, the bulky calf muscles exhibiting mechanical myotonia, the electromyographic records, and the strong family history indicate that this is a case of myotonia congenita. The diagnosis of hypothyroidism cannot be in doubt, and the history suggests that the myotonia preceded the hypothyroidism by about nine years.

Very few cases of the two diseases occurring together seem to have been described. Gordon *et al.* (1952) reported four cases of myotonia congenita in a family, one of whom had a basal metabolic rate of −20% and a blood cholesterol of 371 mg. per 100 ml.

The reason for stressing the existence of two separate conditions is that a number of cases have been described in which myotonoid symptoms and signs were due to hypothyroidism.

Hoffmann (1896) described a syndrome in which myxoedema was associated with hypertrophy and slow action of the muscles. Electromyography in such cases is not typical of myotonia (Nevin, personal communication ; see Poncher and Woodward, 1936), and the condition responds completely to thyroid therapy (Malloret and Sigwald, 1939). Debré and Sémelaigne's cases exhibited the combination of myxoedema, muscle hypertrophy, and retarded growth. The condition occurs chiefly in young children (Thomasen, 1948) and responds completely to thyroid (Debré and Sémelaigne, 1935). Thomasen (1948) described a similar case to that recorded here, but the electromyographic records were not those of myotonia congenita, there was no family history, and all symptoms responded completely to thyroid therapy. Goldstone and Ford (1955) described the cases of two patients who developed severe myotonia as a complication of post-operative thyroid deficiency. The symptoms were completely relieved by the administration of thyroid extract.

The relief of myotonia with thyroid, noted in this case, is interesting, but cannot be a specific effect, for myotonia could still be demonstrated when the patient was hyperthyroid.

Thyroid therapy has been tried in uncomplicated cases of Thomsen's disease with either no effect or a worsening of the myotonia, and one case of Thomsen's disease with hyperthyroidism was improved by thyroidectomy (Thomasen, 1948).

Summary

A case in which myotonia and hypothyroidism coexisted is reported and reasons are put forward for believing that the patient suffers from two distinct diseases, myotonia congenita and myxoedema.

Treatment with thyroid brought improvement in both conditions. With correction of the hypothyroidism, however, myotonia could still be demonstrated.

I am grateful to Dr. W. D. Fletcher for performing the electromyographic studies; to Dr. D. R. Laurence and Dr. T. Geffen for their advice; and to Professor M. L. Rosenheim for his help and encouragement and for allowing me to publish the case.

References

Brown, G. L., and Harvey, A. M. (1939). *Brain*, **62**, 341.
Debré, R., and Sémelaigne, G. (1935). *Amer. J. Dis. Child.*, **50**, 1351.
Goldstone, H., and Ford, F. R. (1955). *Bull. Johns Hopk. Hosp.*, **97**, 53.
Gordon, W. H., Kaufman, J. M., and Kashton, H. A. (1952). *J. Mich. med. Soc.*, **51**, 188.
Hoffmann, J. (1896). *Dtsch. Z. Nervenheilk.*, **9**, 278.
Levin, M. (1934). *Arch. Neurol. Psychiat. (Chicago)*, **32**, 1286.
Leyden, E. (1874). *Klinik der Rückenmarkskrankheiten*, vol. 2. Berlin.
Malloret, P., and Sigwald, J. (1939). *Rev. neurol. (Paris)*, **71**, 513.
Milhorat, A. T., and Wolff, H. G. (1938). *Arch. Neurol. Psychiat. (Chicago)*, **40**, 1135.
Nevin, S. Personal communication. See Poncher, H. G., and Woodward, H., *Amer. J. Dis. Child.*, 1936, **52**, 1084.
Poncher, H. G., and Wade, H. W. (1938). *Amer. J. Dis. Child.*, **55**, 945.
Thomasen, E. (1948). *Myotonia*. Aarhus.
Thomsen, J. (1876). *Arch. Psychiat. Nervenkr.*, **6**, 702.

Medical Memoranda

Sensitization Reactions to Tulle Gras Dressing, B.P.C.

After the conquest of Peru in the sixteenth century balsam of Peru was introduced into European medicine. It is obtained from the trunk of the tree *Myroxylon pereirae*, and contains cinnamic and benzoic acids.

Fuller (1730) says it has many uses ; for instance, in treating coughs, spitting of blood, and dysentery, or with bees burnt, mouse dung, and honey for baldness. Graham (1832) states that the balsam is a stimulant and tonic, cleansing and stimulating foul and indolent ulcers.

More recently balsam of Peru has been used as a mild antiseptic. In the *Extra Pharmacopoeia* (1952) it is said to have a slight irritant local action and to stimulate epithelial regeneration, being employed, diluted with castor oil, for bedsores and chronic ulcers, and as an ointment for skin diseases such as eczema and pruritus. It is also mentioned as a treatment for scabies. Bloom and Lorincz (1954) used balsam of Peru, 10% in petrolatum, as a remedy in three cases of dermatitis repens ; they found, using *in vitro* tests, that the balsam had bacteriostatic effects in similar concentration on several pathogenic bacteria commonly found in skin lesions.

The commonest use of balsam of Peru at present is in curatio carbasi paraffini *B.P.C.*, otherwise known as tulle gras dressing. This consists of bleached cotton cloth impregnated with yellow soft paraffin containing balsam of Peru, 1 part in 80. Several identical proprietary brands are made as well as tulle gras with the addition of sulphanilamide, etc.

It is known that balsam of Peru may cause sensitization reactions. Andrews (1954) states that balsam of Peru may cause dermatitis when incorporated in perfumes (it has been used in brilliantine) ; Schwartz, Tulipan, and Peck (1947) mention the balsam as a cutaneous irritant. Alexander (1955) says: " The constituents of this drug [balsam of Peru], once so widely used, are largely cinnamein with esters of cinnamic and benzoic acids. It was an ingredient of a large number of proprietary preparations. According to Engelhardt, sensitivity to it was first described in 1880. It is a strong contactant, and if applied freely may sensitize some 10% of patients. Patch tests are readily secured. Which of its constituents is the determinant allergen is unknown, but both cinnamon and benzoic acid have the capacity to sensitize the skin."

Tulle gras dressing is widely used, and sensitization of the skin to the balsam of Peru may be overlooked. Sensitization is probably more likely to occur, and more easily overlooked, when the material is applied to already diseased skin.

Recently two examples have been seen. Both patients were women with hypostatic eczema of the legs ; following the application of tulle gras there was increased soreness and exudation. Patch tests with tulle gras were strongly positive,