TWO CASES OF MYOPATHY LIMITED TO THE QUADRICEPS

BY

JOHN N. WALTON

From the Department of Medicine, King's College, University of Durham, and the Royal Victoria Infirmary, Newcastle upon Tyne

Bramwell, in 1922, demonstrated before the Royal Society of Medicine two cases suffering from a form of myopathy which appeared to be limited to the quadriceps. A third similar case was reported by Denny-Brown in 1939, but no subsequent descriptions of this condition have appeared in the literature. All of the three patients previously reported were over 40 years of age; this paper describes two further cases of this type, one in a young man.

Case Reports

Case 1.—J. O., a salesman aged 22 years, in March, 1951, four weeks after joining the Army, found great difficulty in doing physical training; his thighs seemed to stiffen and to slow him down. At the same time he noted considerable difficulty in rising from the floor or from the crouching position, and had to make use of his arms. After exercise he found it difficult to climb stairs, but after rest this was less noticeable. His previous health had been good; his parents and three sisters were healthy and there was no family history of muscular disease.

On examination on August 17, 1951, he was a healthy looking soldier: he walked with a normal gait, but on rising from the floor or from a crouching position he was compelled to use his arms. The cranial musculature, the upper limbs, and the trunk showed no abnormality; there was no myotonia. In the lower limbs there was striking hypertrophy of the lateral vasti (similar to that seen in Case 2, see Fig. 2), but the medial vasti were atrophic. There was marked weakness of extension of the knee joint and of adduction of the thighs. Although the left calf was somewhat larger than the right, and the circumference of the left leg at mid-calf was $\frac{1}{2}$ in. greater than the right, the muscles below the knee were otherwise normal in appearance and power. All the tendon reflexes were active and equal on the two sides.

On April 3, 1952, the patient was admitted to the Royal Victoria Infirmary under the care of Professor F. J. Nattrass. His clinical condition was unchanged; an intramuscular injection of 1.25 mg. of prostigmine produced no subjective or objective change in his condition. An electromyogram from the right vastus lateralis and from the left vastus medialis was recorded with concentric needle electrodes. The pattern was similar in the two sites; there was no spontaneous

activity but on volition the motor units were profuse, reduced in amplitude, and polyphasic; frequency analysis (Walton, 1952) revealed a shift to the right.

A muscle biopsy was taken from the right vastus medialis; it showed that many muscle fibres were severely atrophied while others were enlarged to more than 100 μ in diameter and showed central migration of sarcolemmal nuclei (Fig. 1). There was some fatty infiltration and replacement fibrosis; no necrotic change in muscle fibres, cellular infiltration, or regenerative activity was seen. Sections of a biopsy specimen from the vastus lateralis showed a considerable

FIG. 1.—Transverse section from the vastus medialis (haemalaun and eosin, $\times 100$). Many muscle fibres are atrophic, others are enlarged, rounded, and show central migration of sarcolemmal nuclei. There is some fatty infiltration and replacement fibrosis.

infiltration with fat. Many of the muscle fibres appeared healthy, many others were enlarged and showed central sarcolemmal nuclei; only a few were atrophic.

The patient was discharged from hospital and received 150 mg. α -tocopherol daily. On November 7, 1952, he was subjectively improved and no longer noted stiffness in the thighs although he still experienced difficulty in climbing stairs towards the end of the day. There was no objective change in his condition and the α -tocopherol was discontinued. He was seen again in April, 1953, and was again unchanged. In February, 1955, his symptoms remained slight; he walked six or seven miles daily and had clearly shown no deterioration. The hypertrophy of the lateral vasti was still very marked and was more evident on the right side; both adductor groups and medial vasti were atrophic and weak. The right knee jerk was active, the left depressed.

Case 2.—W. A., a railway shunter, was aged 57. In 1946, at the age of 48, he noted some weakness of the left leg; the knee tended to give way easily and he fell on two or three occasions. An orthopaedic surgeon could find no abnormality in the knee joint. During the succeeding months the knee tended to give way more frequently and similar, though less severe, weakness was noted on the right side. He began to have difficulty in climbing stairs and after bending or kneeling for some time he found considerable difficulty in standing up again. For about a year the weakness appeared to

become worse, but since that time there had been no further deterioration in his condition. He had been compelled to change his occupation, but was working regularly as an electrician's mate in September, 1951, although the knees continued to give way from time to time. Apart from symptoms of peptic ulcer the patient gave no history of any illnesses of note; his parents died in old age and had no symptoms of muscle disease, while five brothers and six sisters were well.

On September 20, 1951, the patient was seen to be a healthy looking, vigorous man who walked briskly with a normal gait. He was slow in climbing stairs and made use of his hands on rising from the floor. No abnormality was noted in the cranial musculature, upper limbs, or trunk. The lower limbs showed marked hypertrophy of the lateral vasti and undoubted atrophy of the medial vasti (Fig. 2). There was marked weakness of knee extension and of adduction of the thigh, but abduction was powerful. The muscles below the knee were normal in appearance and power, the knee jerks were moderately depressed, but all other tendon reflexes were active and equal.

An intramuscular injection of 1.25 mg. prostigmine produced no subjective or objective change in the patient's condition. An electromyogram from the right vastus lateralis was recorded with a concentric needle electrode. There was no spontaneous activity; many areas of the muscle were electrically silent on volition, suggesting fatty infiltration, but in other areas many motor units were polyphasic; a sustained, complex, low-amplitude interference pattern was obtained, and on frequency analysis there was a shift to the right. Muscle biopsy was refused.

The patient was seen again on November 18, 1952, June 22, 1953, and September 22, 1955, and showed no subjective or objective change. There was no evidence that the myopathic process had spread to involve other muscles.

Discussion

Despite the difference in age, the similarity of the clinical manifestations in these cases suggests that both patients were suffering from a myopathic process, limited to the quadriceps, and producing atrophy of the medial vasti with a remarkable enlargement of the lateral vasti. It is probable that the latter change resulted at least partly from compensatory hypertrophy, even though the electromyographic findings in Case 2 indicated that much of the swelling was due to accumulation of tissue other than muscle. Furthermore, in Case 1, although the vastus lateralis was powerful, a biopsy specimen from this muscle showed early myopathic changes and fatty accumulation. It is of interest that Bramwell (1922) described compensatory hypertrophy of the tensor fasciae latae in one of his cases, while Denny-Brown's (1939) patient showed selective atrophy of the vastus medialis. The mechanism of muscular enlargement in patients

FIG. 2.—Photograph of the lower limbs of Case 2. There is striking hypertrophy of the lateral vasti while the medial vasti are atrophic.

with muscle disease is poorly understood. The fact that the calf muscles of a patient with progressive muscular dystrophy are often characteristically enlarged and that histological examination reveals a massive infiltration with fat has given rise to widespread use of the term pseudohypertrophy. However, this muscle group characteristically remains powerful until a late stage of the disease and it appears that some muscle fibres in such muscles may show true hypertrophy. Certainly in Case 1 of the present report the biopsy specimen from the vastus lateralis showed certain findings which could be interpreted as being due to a combination of compensatory hypertrophy and pseudohypertrophy.

In Denny-Brown's case, histological examination of the affected muscle revealed changes which were characteristic, not of progressive muscular dystrophy, but of "late-life" (Nevin, 1936) or "menopausal" (Shy and McEachern, 1951) muscular dystrophy. Adams, Denny-Brown, and Pearson (1953) and others believe this disorder to be a form of polymyositis, nosologically distinct from true muscular dystrophy. It thus appears that occasionally the polymyositis syndrome may be limited in its clinical effects to the quadriceps. Laurent (1953) has observed a case which may be of this type. A woman of 52 years gave a history of 11 years' progressive weakness and wasting of both quadriceps; she showed a dramatic improvement in power following an intramuscular injection of prostigmine and on continuous oral therapy this improvement was maintained until her death many years later. In this connexion it is of interest that I have observed a variable response to prostigmine in some cases of polymyositis.

So far as the cases in the present report are concerned, however, it is clear from the histological

findings that Case 1 was not suffering from polymyositis. In this patient the muscular changes were those which are believed to be characteristic of progressive muscular dystrophy. In the absence of histological evidence it is impossible to be certain as to the nature of the pathological process in Case 2 but the similarity of his symptoms and signs to those of Case 1 was striking. It seems probable that both patients were suffering from a true muscular dystrophy which for some reason was localized to the quadriceps. Probably this disorder may reasonably be regarded as a *forme fruste* of the limb-girdle type of muscular dystrophy, corresponding to the abortive cases of the facioscapulo-humeral type, which are much more common (Walton and Nattrass, 1954).

Summary

Two cases are reported of a form of myopathy limited to the quadriceps, in which there was atrophy and weakness of the adductors and medial vasti but enlargement of the lateral vasti.

It is pointed out that certain cases of this type may be found to be suffering from that form of the polymyositis syndrome referred to as "late-life" or "menopausal" muscular dystrophy. Reasons are given, however, for suggesting that the two cases of the present report were examples of a *forme fruste* of limb-girdle muscular dystrophy.

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