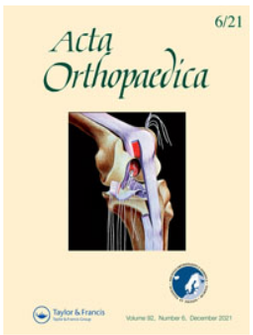

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UNILATERAL JUVENILE MUSCULAR ATROPHY OF UPPER LIMBS

By

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In 1959 *Hirayama* and co-workers (1) drew attention to a new clinical entity of juvenile muscular atrophy of unilateral upper limb based on their experience of 12 cases. In 1963 *Hirayama* and co-workers (2) reported a more detailed study including laboratory tests of 11 of the 12, and of 9 new patients. Early onset, localized muscular atrophy of one arm, the disease is progressive in the one arm for some years and then becomes stationary. This form of muscular atrophy is distinctly different from any other entities of muscular atrophy hitherto described. In 2 patients *Hirayama* and co-workers (1:2) found hyperactive deep reflexes of the leg on the affected side. This paper described a typical case, the symptoms and the diagnostic problems.

CASE REPORT

A 23-year-old male undergraduate (engineer) was admitted to the out-patient clinic at Orthopaedic Hospital in Aarhus on February 28, 1966. A few weeks before admission he had been found unfit for military service because of selective atrophy of the right upper arm.

The young student, who had never been in hospital before, now wanted to be examined.

His family history was noncontributory. During school-age he had been healthy. 8 years ago he started to work as an electrician apprentice and already the first winter he noticed that he fumbled when using a small screw-driver, and occasionally he unintentionally dropped the screw-driver—he was right-handed. Occasionally he had been aware of a fine tremor of the fingers of the right hand, especially when he was working with small objects. When it was cold the right hand was stiff and felt cold. He had no pain or paraesthesia. Three years previously, when he started his education as an engineer he was aware of a slight progressive weakness of the grip of his right hand and he felt some trouble in using a pencil for more than 3–4 hours

without a rest. He had no pain in the neck or trouble with the left hand or his lower limbs. The past 2 years he had not noted any progression or changes on the affected side.

Two years ago he had an attack of dizziness after he had been working for 5 hours at the drawing table. He stayed in bed for a week and was bothered by some dizziness for 3 months followed by jerks of the head. He has not have any such attack since, no headache, no tinnitus or disturbances of vision, he cannot recall any spell of fever or the possibility of polio-myelitis. He still uses the right hand more than the left.

Examination on February 28, 1966, revealed a well nourished and healthy looking young man with considerable atrophy of all intrinsic muscles of the right hand. The right thenar eminence was atrophic. There was decreased power of abduction of all the fingers. Some weakness of the flex. prof. to the IV and V finger together with weakness of the flexor pollicis longus and brevis. There was slight paresis of the extensor carpi ulnaris muscle. The other muscles of the right arm and shoulder girdle were normal.

There was a slight asymmetry of the face without changes of the mimic muscles or the sternocleido-mastoid muscles.

Very pronounced fascicular twitchings were observed in the atrophied muscles, especially in the I interosseous muscle. Appreciation of light touch (wisp of cotton wood) and superficial pain (pinprick) was normal. The V finger and the ulnar half of the IV finger felt dry, suggesting slight hypohidrosis, but the ninhydrin – sweat-test was negative.

The deep tendon and periosteal reflexes of the upper limb were normal. The groove of the ulnar nerve was normal apart from a skin scar after a boil. The abdominal skin reflexes were present. Patellar tendon reflexes were present and symmetric. The ankle jerk on the right side was more brisk than on the left. The plantar reflex on the right side was atypical and periodically positive. There was no paresis of the muscles of the lower limbs but 10 cm over the patella the right thigh was 1.5 cm smaller in circumference than the left.

ELECTROMYOGRAPHY AND NERVE CONDUCTION VELOCITY

Electromyographic examination was performed. The muscles examined were the first dorsal interosseous muscle and the abductor muscle to the fifth finger on both hands. On the left side the electromyographic pattern was normal. On the right side there was a neurogenic affection with moderate loss of motor units, denervation potentials and fasciculation. The mean duration of the potentials was within normal limits.

Conclusion: neurogenic affection on right side. Nerve conduction velocity in both ulnar nervus, measured from the region proximal to the sulcus nervi ulnaris to the wrist was normal (right nervus ulnaris 53 m/sec., left nervus ulnaris 58 m/sec.) (sign. H. J. Hansen).

Muscle biopsy: The biopsy was taken from the right abductor digiti quinti muscle. The muscle had preserved longitudinal and transvers fiber bands. In some areas there were fibres of normal diameter. In other areas there was uniform atrophy of the fibers. The nuclei were pressed together, some of them were pyknotic and in a row. The connective tissue was normal. The amount of fat was normal. There was no sign of infection. Microscopical diagnosis: Neurogenic atrophy (sign. Edith Reske-Nielsen). X-rays studies of the cervical spine did not reveal any abnormality.

SIGNS AND SYMPTOMS

The symptoms and signs in the 20 cases from the literature (1:2) and in our own case may be briefly summarized as follows:

Muscular atrophy: The atrophy is unilateral and confined to the hand and part of the forearm. The brachioradial muscle had preserved its volume, so that the forearm seemed to be more slender in the mid-portion. Fascicular twitchings of the atrophied muscles were observed. The atrophy progressed for some years and then stopped. Tremorlike movements of the fingers were noted, especially when working in the cold with small tools. Motor weakness was always present and often one of the first symptoms, especially when closing and spreading the fingers. Some patients found it difficult to work with the affected hand, especially in cold wheather.

Reflexes: The deep tendon reflexes of the upper limb were usually symmetric. Abdominal skin reflexes were normal. In our patient there was a brisk ankle reflex on the affected side as in 2 cases of Hirayama and co-workers. They did not find positive Babinski reflex on the affected side. In our case there was an atypical plantar reflex on the affected side.

A sensory disturbance was noted in a few patients, hypesthesia in the affected hand, but not in our case. In our case there was slight hypohidrosis of the V and half of the IV finger on the affected side. Hypohidrosis was noted once (case 8) by *Hirayama* and co-workers (2).

DIAGNOSTIC PROBLEMS

The diagnostic problems in our case were several. We first thought the disease to be early acute anterior poliomyelitis: There had not been any spell of fever, no polio-epidemic or polio in the patient's relatives

before onset of the disease. The stationary course, together with the localized atrophy with unique distribution and the characteristic age of onset enabled differentiation from the spinal progressive muscular atrophy.

The slow progress, the cessation of the disease together with the age at onset argue against amyotrophic lateral sclerosis. The possibility of syrengomyelia of the cervical cord was ruled out by the lack of sensory impairment. Cervical spondylosis and cervical rib were not seen. Hermatomyelia, multiple sclerosis and various kinds of inflammatory myelitis were differentiated by mode of onset and course.

Carpal tunnel syndrom was easy to exclude as were other peripheral nerve injuries. Atrophy in these disorders shows a different distribution and is accompanied by sensory disturbances.

The so-called parietal atrophy described more than a century age (3) may also be considered. This supranuclear lesion may lead to atrophy of the contralateral half of the body, or part of it. However, there is no phenomena of denervation or fasciculation. Welander's disease can be ruled out by the age of onset. Myopathies, such as progressive muscular dystrophy and myositis are easily excluded. The clinical course, the muscle biopsy and the electromyographic findings did not sustain the diagnosis.

The nature of the lesion and the cause of the disease are still unknown. Juvenile onset, stationary course and male sex preponderance are characteristic. No autopsy data are available. The final diagnosis should await complete pathologic studies.

SUMMARY

One case of juvenile muscular atrophy of unilateral upper extremity is described for the first time in Scandinavian literature. The case is discussed together with 20 cases found earlier in Japan. The disease is not inherited and predominantly males are affected. Juvenile onset, localized unilateral muscular atrophy, especially of the arm, and non-progressive course in the stage are cardinal features of this disorder. Electromyography demonstrates neurogenic affection, and normal nerve conduction velocity in motor nerves. Muscle biopsy shows the picture of neurogenic atrophy. The disorder is distinctly different from any hitherto known entities of muscular atrophy. As yet no pathologic studies have been reported.

RESUME

Il est décrit pour la première fois dans la littérature scandinave un cas d'atrophie musculaire juvénile unilatérale de l'extrémité supérieure. Il est discuté de ce cas rapproché de 20 autres trouvés antérieurement au Japon. La maladie n'est pas héréditaire et elle atteint principalement le sexe mâle. Attaque juvénile, atrophie musculaire localisée notamment dans le bras d'un côté et n'ayant pas une évolution progressive dans le dernier stade sont les éléments cardinaux de ce trouble. L'électromyographie montre une affection neurogène et une vélocité de conduction normale des merfs moteurs. Une biopsie du muscle donne le tableau d'une atrophie neurogène. Ce trouble est nettement différent de toute autre atrophie musculaire décrite jusqu'ici. Il n'a pas encore été effectué d'études pathologiques.

ZUSAMMENFASSUNG

Ein Fall von juveniler Muskelatrophie der oberen Gliedmasse einer Seite wird zum ersten Male in der skandinavischen Litteratur beschrieben. Der Fall wird zusammen mit 20 Fällen, die früher in Japan gefunden wurden besprochen. Die Krankheit wird nicht vererbt und männliche Personen werden vorzugsweise ergriffen. Beginn in der Jugend, örtliche Muskelatrophie, besonders im Arm einer Seite und nicht fortschreitender Verlauf im Spätstadium sind die Hauptzüge dieser Krankheit. Elektromyographie zeight eine neurogene Affektion und normale Nervenleitungsgeschwindigkeit in motorischen Nerven auf. Muskelbiopsie gibt das Bild einer neurogenen Atrophie. Die Krankheit unterscheidet sich deutlich von allen bisher beschriebenen Einheiten muskulärer Atrophie. Bis jetzt sind noch keine pathologischen Studien vorgenommen worden.

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