CT of Primary Muscle Diseases

M. Jiddane, J. L. Gastaut, J. F. Pellissier, J. Pouget, G. Serratrice, and G. Salamon

Seventy-five patients with a variety of muscular dystrophies were studied using computed tomography (CT). At least 11 slices were taken in each patient, from the forearm to the lower leg. Sufficient information was obtained to provide some CT characteristics of several dystrophies, including Duchenne muscular dystrophy, facioscapulohumeral syndrome, limb-girdle muscle myopathies, and myopathic dystrophies. CT promises to be of increasing value in these areas in the future.

Computed tomography (CT) has not been widely used in the diagnosis and investigation of muscular dystrophies. The paucity of reports in the literature bears this out. The appearance of muscle tissues and muscle morphology are very characteristic on CT, however, and many of the features that are early indicators of dystrophic diseases (e.g., atrophied or pseudoatrophied lesions, degeneration, or fatty infiltrations) are easily recognized. While many muscular dystrophies are present at birth, some of them do not manifest clinically for many years. It was our intent to determine if these dystrophies had any useful information to provide on CT. We studied the CT appearance of muscle tissue in patients with a variety of muscular dystrophies. Because this study investigated relatively unresearched areas, it was our main purpose simply to look for correlations of CT appearance and disease.

Subjects and Methods

This work is based on a study of 75 patients with a variety of muscular dystrophies examined at the Neuro-Muscular Disease Clinic during 1981 and 1982. The same procedure was used in examining all patients. It involved one slice through the forearm, two slices through the upper arm, two or three slices of the scapular girdle and pelvis, three slices at thigh level, and three slices of the lower leg. The examination was performed on a CE 10,000 CT scanner. The scans were studied primarily for evidence of morphologic anomalies (atrophy and hypertrophy) and diffused or local anomalies (areas of necrosis, fatty infiltration, etc.) in the hope that we could correlate the appearance of certain features on CT with specific conditions.

Results

Our findings concern essentially four types of muscular dystrophies: Duchenne disease; Landouzy-Dejerine type facioscapulohumeral myopathies; limb-girdle muscular dystrophy; and Steinert myotonic dystrophy. Several other conditions were also examined (e.g., glycogenic muscle infiltration, cortisonic myopathy, polymyositis), but the data are too fragmentary to be discussed here.

Duchenne Muscular Dystrophy

Although many of the types of muscular dystrophies related to the X chromosome have been individualized (Becker syndrome, Emery-Dreysus syndrome), the most frequent form is that described by Duchenne in 1868. The disease is present at birth, and its signs generally manifest clinically when the child is about 3 years old. Walking difficulties appear gradually during childhood. Hypertrophy of the calves occurs early on, and then the disease spreads to the rest of the musculature. Because it is a genetic anomaly, the disease is almost always recognized in the young child, and is evidently hereditary. The evolution is fairly characteristic. Late walking, calf hypertrophy, and difficulty in climbing stairs or in rising from a sitting position are common. The damage finally reaches the arm muscles, and a scoliosis appears. At about 10 years of age the child's muscular degeneration is such that he or she can no longer walk and must be put into a wheelchair. Death is usually due to recurrent respiratory infections, and occurs when the patient is about 20. Victims of the disease also usually present some form of mental retardation. The disease's essential biologic criterion is an increase in creatine kinase [1, 2].

From an anatopathologic viewpoint, the numerous anomalies associated with Duchenne muscular dystrophy explain the importance of CT scan images. These include muscle fiber modifications, degenerative processes, muscle necrosis, and fat infiltration. At the end of the disease's evolution, the muscle has disappeared, leaving in its place fibrous fatty tissue. Ironically, when anatomic examinations are performed, they do not show any lesions in the central nervous system; it is the muscle biopsy that discovers the anomalies. Electromyography sometimes shows fibrillation, but more often demonstrates pseudomyotonic discharges and continual voluntarily disturbed traces that are small, of short duration, and interfered with by the least movement.

We examined eight patients, aged 4 – 14 years. The length of diagnosed Duchenne evolution was 2 – 8 years. The patients examined at the onset of the disease presented CT anomalies, especially in the calf. These anomalies were pseudohypertrophy and decreased muscle density. The latter was also found at the level of both thighs (fig. 1). In the cases involving later stages of evolution, all the muscles were damaged. They presented an aspect of lowered density, evoking a fat infiltration (figs. 1 and 2).

CT examination allowed a very precise view of lesion topography and permitted us to follow the disease's evolution. Especially valuable was its ability to distinguish pseudohypertrophy from actual hypertrophy. In an actual hypertrophy, muscle volume increases...
and its density is normal while in a pseudohypertrophy the increased muscle volume is due to a fat infiltration of muscle and the increase of subcutaneous fat tissue.

Facioscapulohumeral and Scapuloperoneal Syndromes

The facioscapulohumeral myopathies described under the name of Landouzy-Dejerine disease can also be included in the group of hereditary myopathies. Scapuloperoneal atrophies belong to a smaller and less diverse group that is distinguished by a slightly different neurogenic origin.

The facioscapulohumeral myopathies are transmitted by dominant autosomal chromosomes and especially affect the facial mimic muscles. Difficulty in closing the eyelids and a smooth appearance of the facial muscles are two common signs. Scapular muscle troubles appear early (difficulty in raising the arms above the head) and progress to problems with the biceps and triceps. The deltoid is often untouched. In contrast to Duchenne disease, hypertrophy is not observed, and muscle damage is often asymmetrical. The muscles of the inferior limbs are often damaged, and in severe forms the paraspinal muscles are often attacked, causing lordosis. Although the disease’s prognosis is much better than that of Duchenne disease, the infantile forms are very severe. Cardiac anomalies are noted. The evolution is usually very slow [3]. The anatomicopathologic examination shows collections of sarcoplasm between groups of myofibril and type II fiber hypertrophy. Inflammatory reactions and areas of necrosis are also possible. When it was possible in our study to examine the central nervous system, it was always normal.

We examined 10 patients aged 17–40. The length of evolution varied greatly. The most common CT anomalies were located at the levels of the brachial biceps and triceps and the scapular girdle. Even in the early stages, inferior member muscle damage was almost always found. Locations of this muscle damage included the buttocks, the thigh, and the posterior and anterior parts of the leg; in scapuloperoneal syndromes, the damage predominated in the anterior part of the leg (fig. 3). For facioscapulohumeral syndrome, the three most interesting features demonstrated by CT were the asymmetry of the lesions (fig. 4), the frequent inferior member muscle damage, and the paraspinal muscle damage.

Limb-Girdle Muscle Myopathies

This group of diseases consists of various conditions classified as limb-girdle muscle myopathies by Walton and Natras [4]. Actually, they belong to an enormous group of primitive muscle diseases that do not affect the face, are transmitted by autosomal heredity (usually recessive or sporadic), and have a benign evolution. (Duchenne considered the cases he encountered to be a form of progressive fatty muscle atrophy.) They can attack both genders, and often appear when the individual is about 30 years old. Half the cases begin with damage to the pelvic belt; the other half with the
scapular muscles. Both the shoulder and pelvic girdles are attacked eventually. Generally, 10–20 years intervene between the onset of the disease and major walking difficulties. It is sometimes very difficult to make a diagnosis between hereditary myopathies or acquired myopathies (e.g., polymyositis or endocrine or toxic myositis) and muscle disease having a spinal origin (Kugelberg-Welander disease). Generally, the pelvic and shoulder girdle muscles are the most frequently damaged, but arm and thigh damage are also common. Electromyography shows myopathic changes (potentially small, short-term, and polyphasic), which are sometimes associated with anomalies suggesting denervation [5]. The anatomopathologic examination demonstrates the myofibril size variations, necrosis zones of muscle fibers, fatty excess, and fibrous infiltration processes. Biologically, there is an increase in serum creatine kinase.

We examined 25 patients with limb-girdle muscle myopathies (15 women and 10 men), aged 20–65. The length of evolution varied greatly. In most cases the muscle damage involved the perihumeral, the buttock, and especially the thigh and calf muscles.

CT revealed a number of interesting facts, including the symmetrical distribution of the muscle damage as opposed to facioscapulo-humoral myopathies (figs. 5–7), the importance of fat infiltration in damaged muscles (figs. 6 and 7), the importance of the increase in subcutaneous fat tissue (figs. 5A and 6) and the frequent attack on spinal muscles (fig. 5B). The attack on the muscles of the thigh and of the leg's posterior muscles was especially notable (fig. 7).

**Myotonic Dystrophy (Steinert Disease)**

A different clinical entity from Thomsen congenital myotonia, Steinert disease is hereditary and transmitted by dominant autosomal chromosome [6]. It attacks both genders and has a slow, progressive evolution. The disease especially affects the distal, face, and neck muscles. In the severe forms there may be pharynx and larynx damage. In every case, the myotonic phenomena are characterized by an abnormally long decontraction after a voluntary contraction or percussion of the muscle; the slowness of its return to a relaxed state is remarkable. Widespread other muscle damage can be noted: cardiac, ocular, endocrine, and digestive. Electromyography is characterized by myotonic discharge. The anatomopathologic examination reveals a proliferation of nuclei, muscular atrophy, fibrosis, and sometimes fat infiltration into the muscle. The
atrophy involves type I fibers in particular, and there are also a
great many neuromuscular and spinal insertions.

Seven patients with myotonic dystrophy were examined. In each
case the evolution was at least 4 years long, and CT always revealed
an important lesion in the distal muscle. In many cases the damage
also concerned the quadriceps muscles. CT characteristics of my-
otonic dystrophy included a decrease in muscle volume, irregular
hypodensity of the muscles, and spinal muscle damage.

Discussion

There are few reports of CT examination of muscles. In 1976 the
value of CT was assessed for the study of neurogenic muscular
atrophies involving last cranial nerve palsies (Wolf, unpublished
presentation). In 1979, Bulcke et al. [7] reported on CT examination
of 24 normal subjects. They proposed a density scale for every
muscle, but did not report on any pathologic cases. In 1981, Bulcke
et al. [8] presented the results of a CT examination of three cases
of Becker disease. In 1977, O’Doherty et al. [9] performed CT on
10 patients. Five had Duchenne muscular dystrophy, one had
fasciocomphalohumeral dystrophy, two had Kugelberg-Welander syn-
drome, one had subacute polymyositis, and one had sarcoid my-
opathy.

We have tried to show the value of CT examination for certain
kinds of muscular dystrophy. CT can detect damaged muscles, and
it orients biopsy and electromyography better than does a clinical
examination. In some cases it can provide a nearly complete report
on the lesions and describe their evolution. For each of the diseases
concerned, it offers new elements that are not revealed by tradi-
tional clinical data, electromyography, or biopsy.

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