CT of primary muscle diseases.

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

AJNR Am J Neuroradiol 1983, 4 (3) 773-776
http://www.ajnr.org/content/4/3/773

CT of Primary Muscle Diseases

M. Jiddane,[1] 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-Déjérine 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-Dreyfus 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 anatomopathologic 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

[1]All authors: Laboratory of Neuroradiology, INSERM U6, 300 Boulevard de Sainte Marguerite, 13009 Marseille, France. Address reprint requests to G. Salamon.

AJNR 4:773–776, May/June 1983 0195-6108/83/0403-0773 $00.00

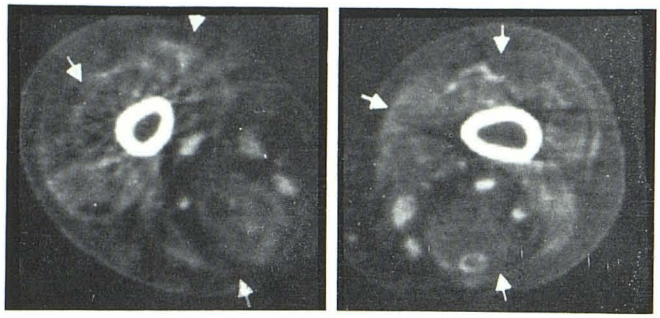


Fig. 1.—Duchenne muscular dystrophy. At thigh level all muscles have disappeared (*arrows*), which is characteristic of disease's late stage.

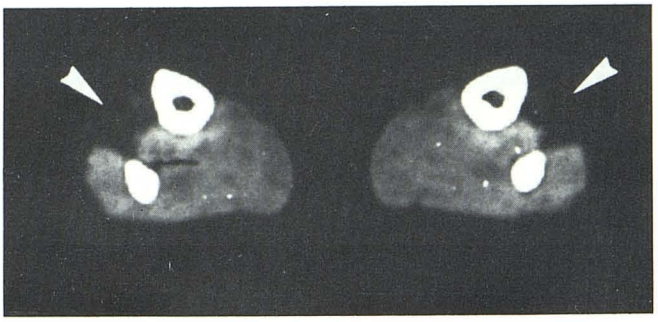


Fig. 3.—Landouzy-Déjérine syndrome (facioscapulohumeral syndrome) in 38-year-old woman with 10 year clinical evolution. At leg level there is disparity of third peroneal muscle on both sides (*arrowheads*). Pattern is frequently observed in this syndrome and especially in so-called scapuloperoneal syndrome.

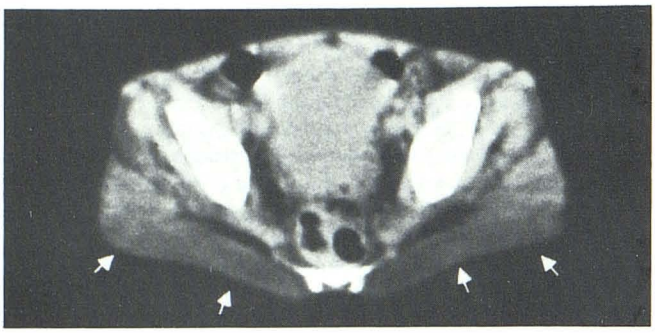


Fig. 2.—Duchenne muscular dystrophy in 9-year-old child. Density of gluteus maximus on sides is very low (*arrows*), while other muscles are normal. This is usual finding in Duchenne at this stage.

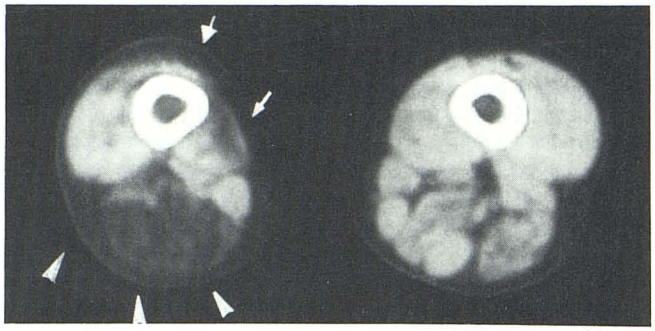


Fig. 4.—Landouzy-Déjérine syndome (facioscapulohumeral syndrome) in 36-year-old woman with 17 year clinical evolution. Slices at thigh level show asymmetry of lesions. Involvement of posterior thigh muscle parts on right side: biceps, semitendinous, semimembranous, gracilis, and great adductor (*arrowheads*). Minor champs on rectus femoris and vastus medialis (*arrows*).

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

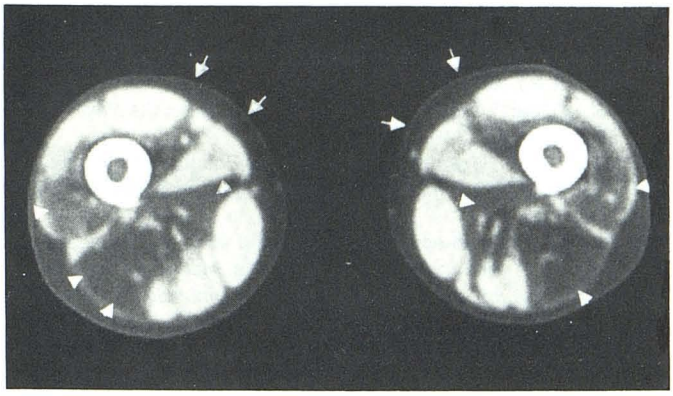
The facioscapulohumeral myopathies described under the name of Landouzy-Déjérine disease can also be included in the group of hereditary myopathies. Scapuloperoneal atrophies belong to a smaller and less diverse group that is distinguished by a slightly different neurogenic origin.

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 anatomopathologic 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.

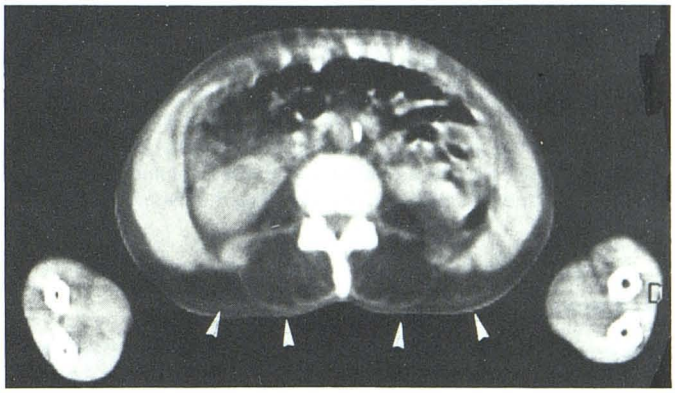
We examined 10 patients aged 17–40. The length of evolution varied greatly. The most common CT anomalies were located at the levels of the brachial biceps and triceps and the scapular girdle. Even in the early stages, inferior member muscle damage was almost always found. Locations of this muscle damage 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 Natrass [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



A



B

Fig. 5.—Limb-girdle syndrome in 61-year-old man after several years of clinical signs. **A,** Slices at thigh level show only minor infiltration of fat into subcutaneous space (*arrows*). This is observed mainly in men. Muscles are infiltrated symmetrically. At same level adductors and vastus lateralis are extensively involved by abnormal infiltration (*arrowheads*), which here proved to be glycogenic. **B,** Disparity on both sides of paraspinal muscles (*arrowheads*).

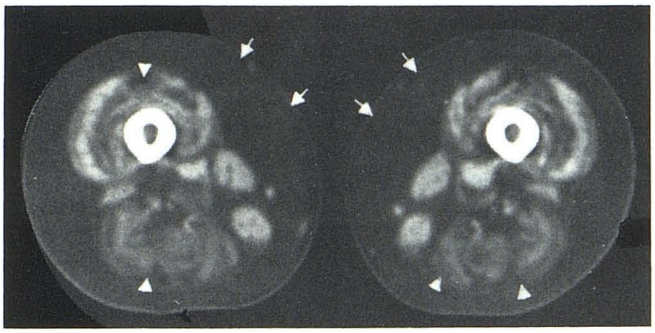


Fig. 6.—Limb-girdle syndrome in 22-year-old woman. Infiltration of subcutaneous fat is enormous (*arrows*), which is common in women with limb-girdle syndrome. All muscles of thigh are affected on both sides (*arrowheads*).

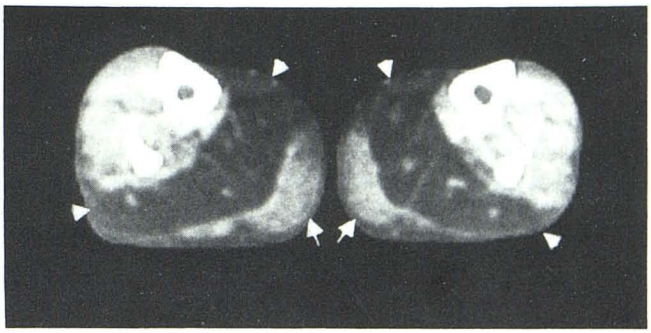


Fig. 7.—Limb-girdle syndrome. Patient had clinical hypertrophy of muscles of leg. Soleus on both sides is totally infiltrated (*arrowheads*). Only gastrocnemius (*arrows*) appears partly normal. Thus, biopsy might prove normal on the latter muscle.

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 facioscapulohumoral myopathies (figs. 5–7), the importance of fat infiltration in damaged muscles (figs. 6 and 7), the importance of the increase (especially in women) of 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 myotonic 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 facioscapulohumeral dystrophy, two had Kugelberg-Welander syndrome, one had subacute polymyositis, and one had sarcoid myopathy.

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 traditional clinical data, electromyography, or biopsy.

REFERENCES

1. Rowland LP, Layzer RB. X linked muscular dystrophies. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 40, part I: *Diseases of muscle.* Amsterdam: Elsevier/North Holland, **1979**:349–414
2. Serratrice G, Gastaut JL, Pellissier JF, Pouget J, Desnuelle C, Cros D. *Maladies musculaires.* Paris: Masson, **1982**:77–109, 132–143
3. Carroll JE. Facio-scapulo-humeral and scapulo peroneal syndrome. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 40, part 1: *Diseases of muscle.* Amsterdam: Elsevier/North Holland, **1979**:415–431
4. Walton JN, Nattrass PJ. On the classification, natural history and treatment of the myopathies. *Brain* **1954**;77:170–231
5. Bradley WG. The limb girdle syndrome. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 40, part 1: *Diseases of muscle.* Amsterdam: Elsevier/North Holland, **1979**:433–469
6. Roses AD, Harper PS, Bossen EH. Myotonic muscular dystrophy. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*, vol. 40, part 1: *Diseases of muscle.* Amsterdam: Elsevier/North Holland, **1979**:485–532
7. Bulcke JA, Termotte JL, Palmers Y, Crolla D. Computed tomogaphy of the human skeletal muscular system. *Neuroradiology* **1979**;17:127–136
8. Bulcke JA, Crolla D, Termote JL, Baert A, Palmers Y, Van Den Bergh R. Computed tomography of muscle. *Muscle Nerve* **1981**;4:67–72
9. O'Doherty DS, Schellinger D, Rapotopoulos V. Computed tomographic patterns of pseudohypertrophic muscular dystrophy: preliminary results. *J Comput Assist Tomogr* **1977**;1:482–486