

muscles, which we shall presently describe, is not uncommon in the juvenile form and is here never present, there is no question but that the two affections are identical, and that only in some cases, from reasons not as yet understood, the interstitial connective tissue becomes early increased, while in others nothing else can be demonstrated but simple atrophy, with increase in the number of muscle nuclei and here and there the formation of vacuoles in the fibres. The diagnosis is so much facilitated by the "myopathic facies"—that is, the expression produced by the sinking in of the cheek, the somewhat dependent lower lip, and the inability to close the eyes—that the experienced diagnostician is frequently able to recognize the disease at the first glance. Marie and Guinon have called attention to the possibility of confounding the disease with *lepra anæsthetica*, in the course of which also weakness of the facial muscles exists (cf. lit.). It is interesting to note in this connection that sometimes disturbances of function in the facial muscles may constitute a congenital defect which under certain circumstances may be followed by an actual atrophy; further, that in sisters or brothers of individuals who suffer from this myopathy which we have just described, a certain imperfection in the development of the facial muscles may be found, although the disease never breaks out in them. These are facts which Strümpell especially has pointed out, but the cause still remains wholly unexplained. About the treatment we need add nothing to what we have said with reference to the juvenile form.

The third form of the muscular diseases now under consideration—the so-called pseudo-hypertrophy—is connected with an increase in the interstitial adipose tissue which, in spite of the atrophy of the muscle fibres, lead to an apparent increase in the volume of the affected parts. The disease was known and described by Griesinger in 1864, and again by Duchenne in 1868. It begins generally in the muscles of the trunk and attacks, in contradistinction to the two forms just described, by preference the lower parts of the body, the muscles of the back, loins, and thighs. Though for a long time the patient can use his arms and hands just as well as usual, the walk, owing to the affection of the erector muscles of the spine, becomes altered in the characteristic manner which we have described on page 363. The condition of the patient may remain unchanged for years before the arms also take part in

the process. When this happens it occurs in the same manner as in the juvenile form. The diagnosis is very much facilitated by the appearance of the patient. The enlargement of the calf muscles, the thighs, and the glutei (which are sometimes colossal), give to him the appearance of a giant and suggest a supernatural strength (cf. Fig. 130); but the fact that these great masses feel spongy and soft, and that the electrical excitability is considerably decreased owing to the diminution in the number of the muscle fibres, readily explains why these sturdy-looking persons are feeble and without strength, and almost wholly deprived of the use of their limbs.

In its onset the disease resembles closely the other forms. Here also only children become affected, more especially those between the ages of four and nine. Again, the disease may occur in several members of the same family, so that we must undoubtedly assume a hereditary predisposition; and here also the fibrillary twitchings are not met with. Duration and treatment are the same as in the juvenile atrophy.

Congenital atrophy of the muscles may be found in cases of malformation of the arms and hands. Fig. 131 represents a boy aged thirteen in whom the forearms are absent; some of the fingers are grown together and some deformed. A similar case has been reported by Wilkin (*Lancet*, page 1265, December 14, 1887), where there was atrophy of the biceps and the *brachialis anticus*.

Absence of certain individual muscles is rarely observed. Erb has reported a case in which there was an almost entire absence of both trapezii (*Neurolog. Centralbl.*, i, 1889). Among earlier instances the pectorales (Ziemssen), the biceps (McAlister), the deltoid, and gastrocnemius (Gruber), were wanting. These cases possess no clinical interest.

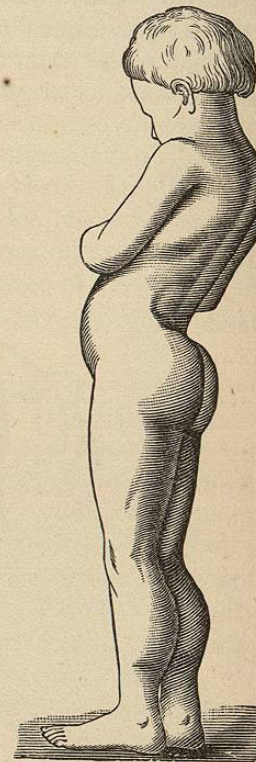


Fig. 130. — PSEUDO-HYPERTROPHY OF THE MUSCLES OF THE LEGS WITH ATROPHY OF THE MUSCLES OF THE BACK. (After DUCHENNE.)

The sensory disturbances which are peculiar to the muscles, but about the exact anatomical nature of which we know nothing, are called myalgias or muscular rheumatisms. *Ætiologically*, overexertion, strains (possibly rupture of certain muscle fibres, which may happen during gymnastic exercises or other violent bodily exertion), must be mentioned in this connection. Sometimes we are unable to find any such cause, and we have to attribute the trouble to the influence of cold.

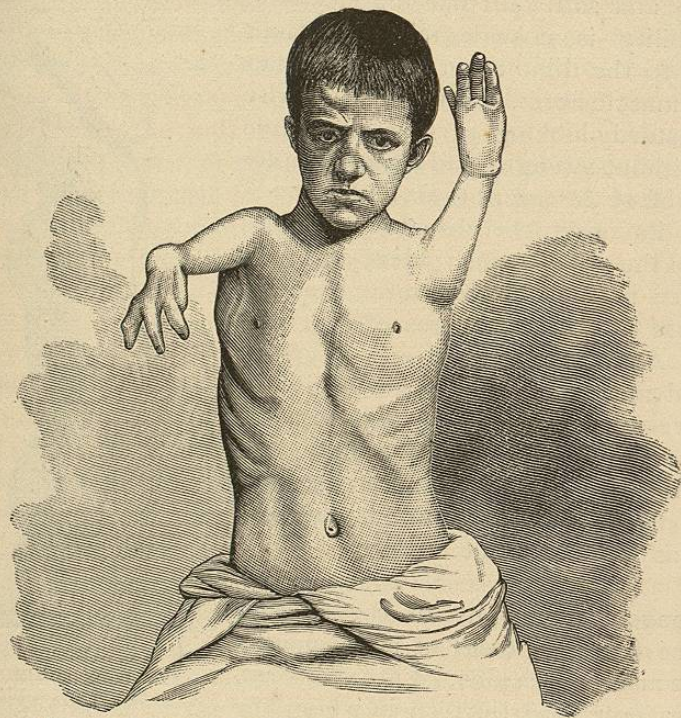


Fig. 131.—ABSENCE OF THE FOREARMS. The fingers are partly grown together. Atrophy of the muscles of the upper arms (personal observation).

There are persons who for years or tens of years suffer from myalgic pains which come and go and may disappear for certain periods of time completely, and it is just possible that chronic intoxications—e. g., alcoholism, perhaps also circulatory disturbances—have a predisposing influence. Among such myalgias, which may be very painful, even sufficiently so as to interfere with the occupation of the patient for a longer or shorter period of time, we have, for instance, the torticollis rheumatica, in which the muscles of the neck, the

myalgia lumbalis (lumbago), in which the muscles of the loins, the myalgia intercostalis, in which the intercostal muscles are attacked. The shoulder muscles may also be affected, and the myalgia in this region may become very obstinate without any implication of the brachial or cervical plexuses being demonstrable. In the diagnosis we must think of the possibility of an implication of the nerves and endeavor to exclude neuralgia. We must further remember that central diseases may give rise to muscular pains. The inexperienced may mistake the lancinating pains of tabes for chronic muscular rheumatism, and thus obscure the correct diagnosis for years. It will hardly be difficult to avoid confounding muscular rheumatism with articular rheumatism if we take into consideration the general condition of the patient, the appearance of the joints, the temperature, pulse, etc., which in the former affection remain normal.

In the treatment we should first of all endeavor to detect any underlying cause, and, if such exists, remove it. In recent cases, besides subcutaneous injections of morphine, salicylic acid may be tried internally; yet we should not spend much time with it if we perceive no effect, but should rather prefer local applications—irritants to the skin, poultices, mustard plasters, liniments, also massage and electricity—especially if the affection remains localized. If this is not the case, but if the pains travel round the body and the course assumes a more chronic type, treatment by sweating, steam baths, also mud baths or baths of *Pinus silvestris*, the non-medicated hot springs (Gastein, Johannisbad, Teplitz) or the sulphur springs, among others Pistyán, in Hungary, will be recommended. As a last resort, we may advise the patient to go to a well-conducted hydrotherapeutic establishment (Gräfenberg, Kaltenleutgeben, Nassau, etc.).

LITERATURE.

- Landouzy et Dejerine. De la myopathie atrophique progressive. *Revue de méd.*, Février-Mars, 1885.
 Marie et Guinon. Formes cliniques de la myopathie progressive primitive. *Ibid.*, Octobre, 1885.
 Westphal. Ueber einige Fälle von progressiver Muskelatrophie mit Betheiligung der Gesichtsmuskeln. *Charité-Annalen*, 1885.
 Charcot. Revision nosographique des atrophies musculaires progressives. *Progr. méd.*, Mars 7, 1885.
 Mossdorf. *Neurol. Centralbl.*, 1885, iv, 1. (Implication of the Facial Muscles in the Juvenile Muscular Atrophy.)

- Krecke. Münch. med. Wochenschr., 1886, xxxiii, 14-16. (Implication of the Facial Muscles in Muscular Atrophy.)
- Ladame. Contribution à l'étude de la myopathie atrophique progressive. Revue de méd., Octobre, 1886.
- Landouzy et Dejerine. Nouvelles recherches sur la myopathie atrophique progressive, etc. Revue de méd., Decembre, 1888.
- Lichtheim. Ueber hereditäre progressive Muskelatrophie. Schweizer Corr.-Bl., 1888, xviii, 19, p. 603.
- Bernhardt. Ueber eine hereditäre Form der progressiven spinalen, mit Bulbärparalyse complicirten Muskelatrophie. Virchow's Archiv, 1888, Bd. 115, 2.
- Lesage. Note sur une forme de myopathie hypertrophique secondaire à la fièvre typhoïde. Revue de méd., 1888, viii, 11, p. 903.
- Sachs. Progressive Muscular Dystrophies. Journal of Nerv. and Ment. Diseases, November, 1888, xiii, 11.
- Stern. Ein Fall von progressiver Muskelatrophie (juvenile Form, Erb), mit halbseitiger Betheiligung des Gesichtes. Mittheil. aus d. med. Klinik in Königsberg. Leipzig, Vogel, 1888.
- Lichtheim. Ueber hereditäre progressive Muskelatrophie. Centralbl. f. Nervenheilk., 1888, xi, 20.
- Souza, Antonio Veiga de. Zwei Fälle von juveniler Form der Muskelatrophie. Inaug.-Dissert., Kiel, 1888.
- Troisier et Guinon. Deux nouveaux cas de myopathie progressive primitive chez le père et la fille. Revue de méd., 1889, ix, 1.
- Rémond. Une observation d'atrophie musculaire myélopathique à type scapulo-huméral. Progr. méd., 1889, 2.
- Winkler en van der Weyde. Primaire myopathie (type facio-scapulo-huméral) gecombineerd med ophthalmoplegia progr. superior. Nederl. Weekbl., 1889, i, 3.
- Scheuthauer. Histol. Untersuchung eines Falles von Pseudohypertrophie der Muskeln. Arch. f. Psych. u. Nervenkr., 1889, xx, 2.
- Herringham. Muscular Atrophy of the Peroneal Type affecting many Members of a Family. Brain, 1889, xi, p. 230.
- Pal. Ueber einen Fall von Muskelhypertrophie mit nervösen Symptomen. Wiener klin. Wochenschr., 1889, ii, 10.
- Auerbach. Zur Frage der wirklichen oder scheinbaren Muskelhypertrophie. Centralbl. für die med. Wissensch., 1889, 45.
- Limbeck. Fall von completem Cucullarisdefect. Prager med. Wochenschr., 1889, xiv, 36.
- Hitzig. Arch. f. Psych. u. Nervenkr., 1889, xxi, 2, p. 650.
- Stintzing. Deutsches Arch. f. klin. Med., 1889, 45, 3, 4. (Congenital and Acquired Defect of the Pectoral Muscles.)
- Gombault. Sur l'état des nerfs périphériques dans un cas de myopathie progressive. Arch. de méd. expérim. et d'anat. path., 1889, 5.
- Duda. Fall von Pseudohypertrophie der Muskeln. Inaug.-Dissert., Berlin, 1889.
- Muselier. Maladies générales chroniques et amyotrophiques. Gaz. méd., 1889, 20.
- Klaas van Roon. Over chronische en progressive atrophie van spieren. Akad. proofsch., Utrecht, 1889.

- Lenoir et Besançon. Myop. progr. primit. (type Landouzy). Revue de méd., 1890, 4.
- Annequin. Arch. de méd. et de pharm. mil., 1890, xv, 4. (Atrophy of the Rhomboid Muscles.)
- Bruns et Kredel. Fortschr. d. Med., 1890, 1. (Congenital Defect of the Pectoral Muscles.)
- Bielschowsky. Neurol. Centralbl., 1890, 1.
- Spillmann et Haushalter. Revue de méd., 1890, 6.
- Rovighi e Levi. Contribuzione allo studio della distrofia muscolare progressiva. Reggio Emilia, 1891.
- Guttman, P. Deutsche med. Wochenschr., 1891, 34.
- Krauss, William C. Muscular Atrophies. A Clinico-Pathological Study. The Buffalo Medical and Surgical Journal, April, 1891.
- Israel, A. Ueber Dystrophia musculorum progressiva. Inaug.-Dissert., Freiburg, i. B., 1891.
- Erb, W. Dystrophia muscularis progressiva. Volkmann's Samml. klin. Vortr., Neue Folge, November, 1892, 2.
- Muenzer. Zur Lehre von der Dystrophia musc. progressiva. Zeitschr. f. klin. Med., 1893, xxii, 6.
- Senator. Ueber acute Polymyositis und Neuromyositis. Deutsche med. Wochenschr., 1893, 39.
- Higier (Warsaw). Ueber primäre und secundäre Amyotrophien organischer und dynamischer Natur. Ibid., 1893, 38, 39.
- Strümpell. Deutsche Zeitschr. f. Nervenhk., 1893, p. 471.