Acta Myologica • 2014; XXXIII: p. 34-42

MEMORIES BY A MYOLOGIST

Vladimir Karlovich Roth (1848-1916): the founder of neuromuscular diseases studies in Russia

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This article shortly examines the biography, scientific activity and scientific work on neuromuscular diseases of the famous Russian neurologist Vladimir Roth who was the founder of neuromuscular disorders study in Russia. In 1876 he was the first in Russia who performed an autopsy and a detailed histological study of a case of progressive muscular atrophy, in which he did not find changes in the nervous system. He called this disease "muscular tabes" i.e. myopathy. In 1884 Vladimir Roth expressed his opinion about the nosological place of the peripheral type of muscular tabes to be considered as a distal myopathy. Dr. Roth became well-known for his monograph of the neuromuscular diseases, published in Moscow in 1895 under the name "Muscular Tabes" in which he described the history of neuromuscular diseases in a very detailed way, analyzing 1014 cases published in the world literature from 1830 to 1893 and 125 personal observations in the period 1874-1894. He performed a thorough analysis of the pattern of muscle involvement using both electrodiagnostic and histological study of muscles and central/peripheral nervous system. We report a short review of this monograph and two cases of peripheral (distal) myopathy.

Key words: Muscular tabes, distal myopathy, peripheral neuromuscular involvement

Vladimir Roth is the founder of neuromuscular diseases studies in Russia. He was a disciple of A. Ya. Kozhevnikov – the founder of Russian neurology. Roth was the great clinician and neuropathologist. He was also identified as a brilliant master on electric diagnostic and electric therapy of the neuromuscular diseases.

Roth's scientific biography

Vladimir Roth (Fig. 1) was born on October 5, 1848 in the city of Orel in a family of a pharmacist, originating from Sweden. After gymnasium he was admitted to the Moscow University which he completed in 1871 with honors and stayed as a resident at the neurology clinic upon recommendation of Professor A. Ya. Kozhevnikov.

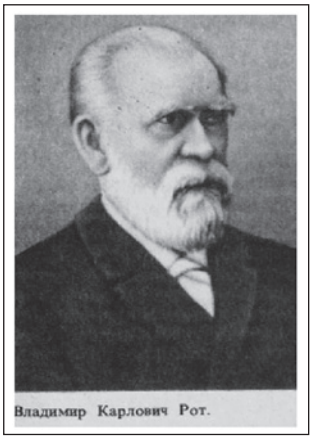

Figure 1. Vladimir Karlovich Roth (1848-1916).

In 1876, after completion of residency, Dr. Roth went to study and work abroad. During 4 years Dr. Roth worked at clinics and laboratories of Paris, Berlin and Vienna with Drs. Vulpian, Charcot, Magnan, Ranvier, Claude Bernard, Broca, Virchow, Leaden, Westphal, Meynert, Obersteiner and Benedikt.

After return to Moscow from 1881 to 1890 he headed a 40-bed Department of Neurology at the Old Catherine's Hospital and delivered lectures in diseases of nervous system and electric therapy. In 1895 he was appointed an Extraordinary Professor of Neurology of the Moscow University and headed out-patient clinic of nervous diseases (1890-1894). In 1899 director of the clinic of nervous diseases (after A. Ya. Kozhevnikov) and from 1902 to 1911 he was Staff Professor of Neurology in the Department of Medicine of the Moscow University.

Roth's scientific activity

Professional, scientific, teaching and social activities of Dr. Roth were associated with the Department of Medicine of the Moscow University, founded in 1755. This

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institution is currently named the First Sechenov State Medical University.

As an invited presenter Dr. Roth participated in many Russian and international scientific meetings: in Geneva (1877), Copenhagen (1884), Magdeburg (1884), Lubeck (1886), Berlin (1890), Bern (1895), Leipzig, Prague, Bordeaux.

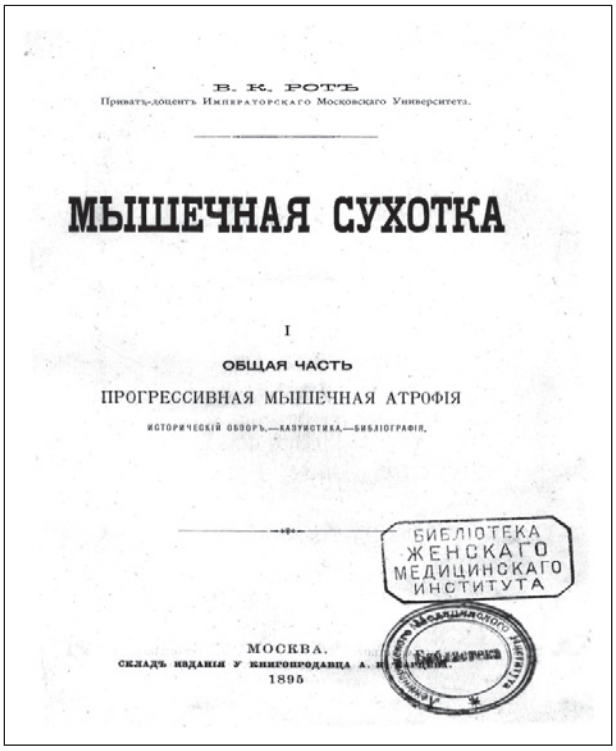
In 1897 Dr. Roth participated in the XII International Meeting of Medical Doctors in Moscow as a lead secretary and organizer. This was the first International medical meeting to be held in Russia. In 1890 Dr. Roth participated in establishing the Moscow Society of Neurologists and Psychiatrists and was elected a chairman of this society (after A. Kozhevnikov). He took part in the work of the "Society of Russian Doctors in the memory of N.I. Pirogov" established in 1881. In 1891 he became the chief editor of the "Korsakov's Journal of Neurology and Psychiatry". This was the first neurological journal to be written and published in Russia. In 1897 Roth edited the first Russian textbook on nervous diseases "Course of Nervous Diseases". He was the founder the Institute of Neurology at the Moscow Imperial University.

Roth's scientific work on neuromuscular diseases including myopathies from 1874 to 1895 and his famous book "Muscular tabes" (Moscow 1895) (Fig. 2).

Dr. Roth published 45 scientific works in Russian and foreign literature, devoted to different aspects of neuromuscular diseases.

In order to better understand the role of Dr. Roth in recognition of some neuromuscular diseases it is necessary to discuss some terminological definition which Dr. Roth used to distinguish these diseases. Dr. Roth described two groups of diseases with atrophy and weakness of muscles (1-3). The first group was called progressive muscular atrophy due to the affection of spinal cord and peripheral nerves which included primary and secondary spinal atrophy, polyneuropathies, neuropathies and amyotrophic lateral sclerosis. The second group was called muscular tabes resulting from disorders of muscle fibers themselves with fatty and connective tissue substitution. In other words the name muscular tabes included myopathies which Dr. Roth divided into basic (central), peripheral and transitional form of muscular tabes in his own material. However, it is necessary to note that when Roth described casuistics from the literature he used the name "peripheral type of muscular tabes" for designation of the neurogenic distal atrophy because according to the pattern of muscle involvement Dr. Roth considered that the cases described by Charcot et Marie, Tooth, Hoffmann, Joffroy, ets. connected with the lesion of muscle themselves but not peripheral nerves or spinal cord (see below and Tab. 1).

In 1873 Dr. Roth started his studies of progressive muscular atrophy, as suggested to him by Prof. Kozhevnikov. In 1874 Dr. Roth gave the information in the Moscow Medical Newspaper (4) about patient K., aged 22 (Fig. 3) with muscle affection similar to Duchenne's progressive muscular atrophy of childhood. After the patient's death of lung tuberculosis in 1876 Roth made a very detailed autopsy and post mortem histological study in which he did not find changes in the nervous system. These results were reported and published later (1, 2, 5). This study showed a pattern of myopathy (the normal and atrophic fibers, the fiber with granular degeneration, capillaries, connective tissue growth and formation of fat cells) while study of the spinal cord, medulla oblongata, sympathetic nerve, nerve roots and peripheral nerves showed no abnormality (description of histology of this case takes 25 pages (pp. 120-144) (6) (Figs. 6, 7). Autopsy and histology results were presented by the author at a meeting of the Society of Russian Medical Doctors and published among the proceedings of this society in 1880 (Moscow) (5). Slides of muscle and nervous tissue (spinal cord and nerves) were presented to Professors Babukhin, Kozhevnikov and Klein in Moscow, as well as to Professors Charcot and Vulpian in Paris, Benedikt in Vienna, who agreed with the author's conclusions.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МЫШЕЧНАЯ СУХОТКА

I

ОБЩАЯ ЧАСТЬ

ПРОГРЕССИВНАЯ МЫШЕЧНАЯ АТРОФІЯ

ИСТОРИЧЕСКІЙ ОБЗОРЪ.—КАЗУИСТИКА.—БИБЛІОГРАФІЯ.

МОСКВА.
СКЛАДЪ ИЗДАНІЯ У КНИГОПРОДАВЦА А. И.
1895

Figure 2. Roth V.K. Muscular tabes. I. General part. Progressive muscular atrophy: historical review, casuistry and references. Kartzev Publisher, Moscow, 1895.

Dr. Roth became well-known especially for his monograph of the neuromuscular diseases published in Mos-

Table 1. Review and grouping of casuistry from literature (1830-1893, author's note) (A total of 1014 cases).

Time of publication	Cases of different neuropaties attributed by the author to progressive muscle atrophy							Spinal amiotrophy								Basic form of muscular tabes									
	Amyotrophic lateral sclerosis	Glyomatosis and syringomielia	Complex spinal and subacute	Spinal pachimeningitis	Nevritis	Nonspecific nevrotic diseases	Origin unknown	Pure, protopathic and type Aran-Duchenne-Charcot	Humeroscapular type	Radial type	Childhood hereditary type	Adult hereditary spinal type	Policencephalomielitis chronica	Peripheral type of muscular tabes	Hypertrophic neuritis Dejerine	Descending form of children and adults	Ascending form of children (pseudohypertrophic)	Ascending form of adults	Typical	Indefinite transitional cases	Total cases of basic muscular tabes	Total spinal forms	Percentage relation (%) of the basic muscular tabes to all amyotrophies collected	Relation of the basic muscular tabes to pure amyotrophies only	Total cases which were analysed
Under 1853	2	1	6	-	2	2	1	-	1	-	-	-	-	2	-	1	12[1]	-	1	-	14	15	45%	14:1	31
From 1854-58	12	1	16	1	4	2	8	8	-	-	-	-	-	2	-	3	1	2	4	-	10	52	16%	5:4	64
,, 1859-63	14	2	5	2	7	2	1	3	1	-	-	-	-	2	-	15[2]	7	-	-	3	25	37	46%	8:1	64
,, 1864-68	6	1	5	-	2	2	1	3	-	-	-	-	-	-	-	4	35[5]	-	-	-	39	20	66%	13:1	59
,, 1869-73	10	5	2	1	4	2	-	4	-	-	-	-	-	17[1]	-	23[3]	24	7	3	6	63	28	58%	18:1	108
,, 1874-78	7	8	10	-	5	1	16	7	1	-	-	-	-	-	-	5	41	4	5	1	56	55	50%	8:1	111
,, 1879-83	5	4	8	-	6	2	-	2	-	1	-	-	1	-	7	84[6]	7	2	4	104	28	79%	52:1	133	
,, 1884-88	5	4	2	1	-	-	3	5	2	-	-	-	-	28	-	42	74	12	15	14	157	22	71%	39:1	207
,, 1889-93	4	4	-	-	-	-	-	7	2	8	18	4	4	22	2	58	76	20	7	1	162	51	70%	23:1	237
	65	30	54	5	30	13	30	39	7	9	18	4	4	74	2	158	354	52	37	29	630	308	62	16:1	1014

1) 10 cases of Eichorist and 5 cases of Friedreich
2) and 3) Many cases in Duchenne's book: 2 nd and 3d editions
4) 8 cases of Meryon
5) 10 cases in Duchenne's monography
6) 24 cases of Gowers

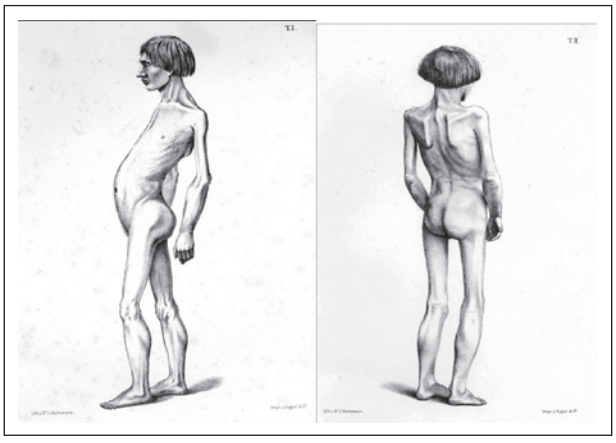


Figure 3. Observation I. (1874 year). Patient A.K., aged 22. Basic muscular tabes, typical form.

cow in 1895 under the name "Muscular Tabes. General part: Progressive muscular atrophy" (Fig. 2) at which he worked during 20 years. For this monograph Dr. Roth was awarded the degree in Medicine and the title of Extraordinary Professor of Neurology. This book includes 478 printed pages plus 19 isolated pages with photographs and drawings of Roth's own patients and histological preparations with their description (6).

This book included:

1) historical review with short clinical analysis of published cases and results of autopsies, histological and elecrtrodiagnostic studies in different neuromuscular diseases;
2) review and grouping of casuistic material;
3) review and grouping author's own observations and description of author's own observations (cases with muscular tabes i.e.myopathy, mainly as well as amyotrophic lateral sclerosis and primary and secondary spinal muscular atrophy) (pp.115-394);

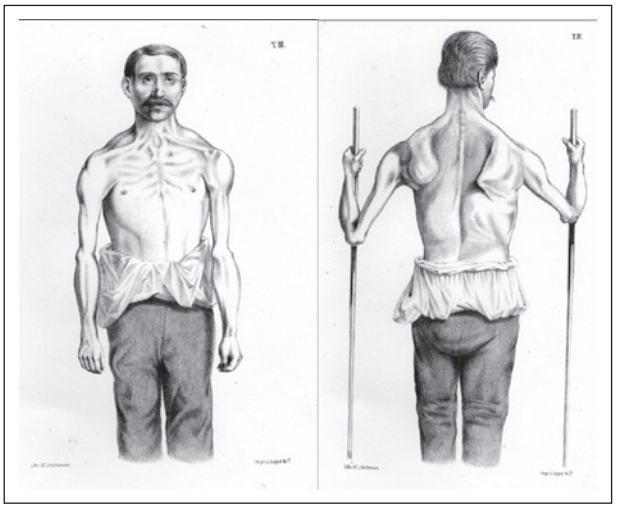


Figure 4. Observation XXIV (1879 year). Patient A.U., aged 31. Basic muscular tabes, descending form.

4) references and alphabetic list of cited authors.

We give a very brief characteristic and only some from these parts.

1) Historical review included the following items:
   - I. Article of Aran on progressive muscular atrophy. Opinion of the physicians on muscle atrophy before XVIII century. Gerardi L.B. van Swieten. Pathology of muscle system and cases of progressive muscular atrophy before the Aran period. Hasse, Graves, Parry, Ch. Bell, Costa &. Gioja, H. Maoy, Froriep, Dubois, Partridge, Duchenne, Romberg, etc.
   - II. Period of Aran, Duchenne and Cruveilhier.
   - III. Further development of progressive muscular atrophy studies. First attempts of clinical differentiation between different forms. Prevalence of myopathic theory in Germany and England. Meryon, Oppenheimer, Wachsmuth, Roberts, Friedberg, etc.
   - IV. Neuropathic theory of the disease. Different anatomic facts. Sympathetic theory of the disease. Jaccound, Valerius, Dumenil, ets. Development of spinal (poliomyelitic) theory. Predecessors of Charcot. Work of Bergmann. Successes in studies of the anatomy nervous system. Autopsy of Luys, Clarke, Dumenil. Spinal cord affection in children's paralysis. Charcot theory of trophic role of large cells of the spinal cord anterior horns.
   - V. Confirmation of Charcot theory by new facts in pathologic anatomy. Autopsy of Hayem, Charcot-Joffroy. Development of teaching of paralysis bulbaris progressiva and it connection with pathogenesis of progressive muscular atrophy. Duchenne, Leyden, Kussmaul. Amyotrophic lateral sclerosis and deutoropathic amyotrophy. Studies of systemic myelitis. Triumph of Charcot's poliomyelitic theory (Vulpian, Charcot).
   - VI. Studies on Friedreich.
   - VII. Muscle pseudohypertrophy. Casuistic material. Monograph of Seidel and Duchenne. Autopsy of Cohnheim and Charcot. Friedreich's opinion in the attitude of pseudohypertrophy to progressive muscular atrophy.
   - VIII. Revival of myopathic theory in Germany. Leyden's opinion on the devision of progressive muscular atrophy into two different forms. Autopsy of Lichtheim. Möbius's opinions. Second monograph of Seidel. Autopsy of Erb–Schultze. Autopsy of Roth. Clinical grouping of the cases progressive muscular atrophy: Leyden, Damaschino, Gowers and Erb.

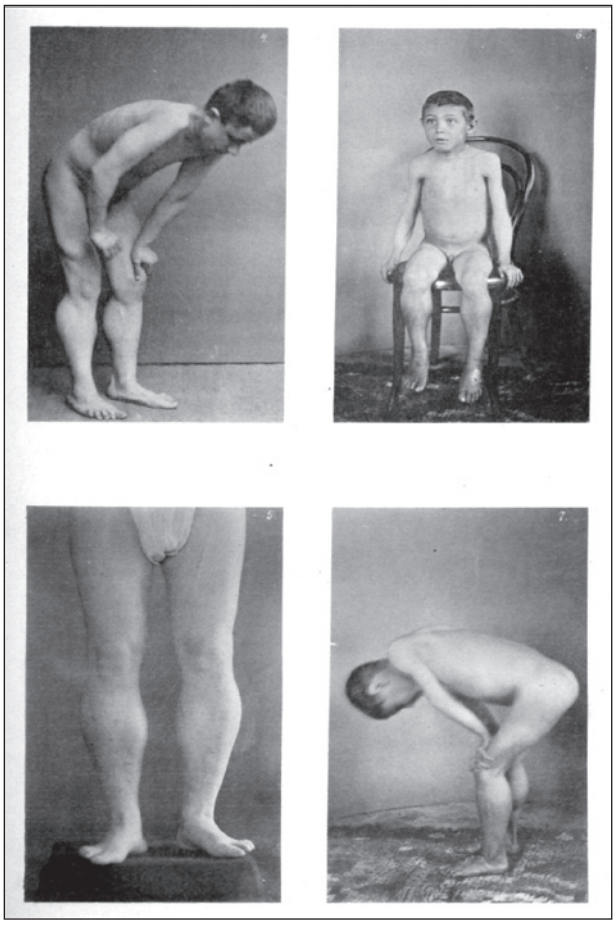


Figure 5. Two photos at the left side - Observation IV. (1891 year).Patient M.S., aged 8. Basic muscular tabes, ascending form (in childhood onset). Upper right - Observation V. (1888 year). Patient I.L., aged 16. Basic muscular tabes, ascending form (in childhood onset but with slow course); Lower right - The boy with pseudohypertrophy of muscles after influence is presented.

IX. Work of Erb. Juvenile form. Dystrophia muscularis progressive. Question of progressive muscle atrophy at the VIII International Medical Congress. Moebius's opinion. Autopsy by Landouzy and Dejerine. Facio-scapulo-humeral type. New opinion of Charcot-Marie-Tooth.

X. Peripheral type of muscular tabes. "Special form" of Charcot-Marie: peroneal type Tooth; Hoffman's neural muscular atrophy. Autopsy of Dubreuilh and Hanel. Hypertrophic neuritis Dejerine. New spinal forms: radial type and hereditary form of Bernhardt. Autopsy of Strumpell. Children's spinal form of Werdnig and Hoffmann. Polioencephalomyelitic form.

XI. Casuistry of the basic muscular tabes. Theory of the vascular origin of this disease. Neuritic theory of Holland authors. Success of tropho-neuropathic theory. Pathogenic theory the disease of Gradenigo, Babinsky and Onanoff and Roth. Opinions of Schultze, Vulpian, Erb and Strumpell.

2) Review and grouping of cases from literature (Table 1): this table shows a review of 1014 cases of progressive muscle atrophy and basic muscular tabes collected by Dr. Roth in the World literature between 1830-1893.

3) Review and grouping of Roth's own observations (Table 2): this table shows

Table 2. Review and grouping of author's own observations (1874-1894 author's note)(A total of 125 observations).

	Glyomatosis of spinal cord	Lateral amyotrophic sclerosis	Don't confirm new cases of aran-Duchenne type	Pure cases of Aran-Duchenne type	Humero-scapular type spinal muscular atrophy	Humero-scapular type did not differentiated	Atrophia which beginning with posterior cervical mescles	Radial type	Atypical cases with affection of forth limbs	Accidental amyotrophies in neuritis, migrenis, poliomyelitis, procasa et others	Unknow origin	Peripheral muscular tabes	Basic muscular tabes, descending variant	Basic muscular tabes, ascending variant in children	Basic muscular tabes, ascending variant in adults	Typical basic muscular tabes	Atypical cases of basic muscular tabes	Total cases of basic muscular tabes	Total cases of progressive muscular atrophy
Author's observation	27	8	5	1	1	3	1	4	1	3	1	4	5	9	4	8	3	29	88
Ambulant patients observed at the Clinic of Neurology for 4 years (1890-1894)	12	2	1	-	-	1	1	2	-	3	-	1	2	1	-	2	-	5	28
Patients not admitted at the Clinic of Neurology in the same period	4	2	-	-		-	-	-	-	-	1	-	-	-	-	-	-	-	7
Patients treated in the Clinic of Neurology during 10 years (1884-1894)	13	12	-	1	-	2	1	1	3	1	-	1	4	2	-	3	-	9	44
All clinical patients excluding repeated cases	26	14	1	1	-	2	1	3	3	4	-	2	5	3	-	5	-	13	70
All author's cases and clinical cases excluding repeated cases	41	20	5	2	1	4	1	4	4	4	2	4	7	9	4	10	3	33	125

In the group of the patients with gliomatosis and lateral amyotrophic sclerosis the patients without muscular atrophies were not included.

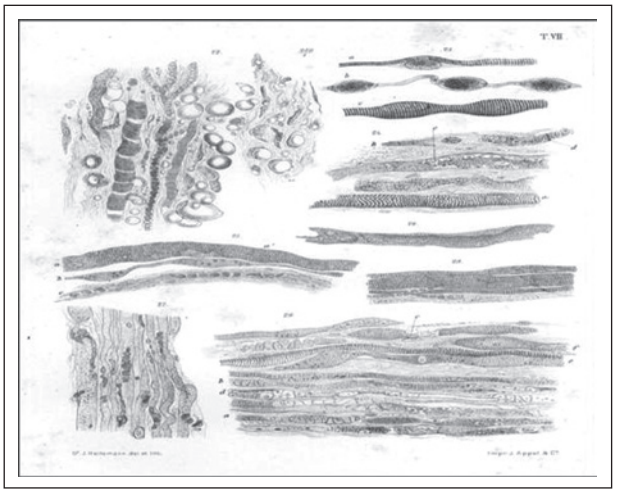


Figure 6. Upper right (Slide n. 22, m. sacro-lumbo-dorsalis). Patient A.K. Muscular tabes. Normal fiber in middle, atrophic fiber on the right, fiber with granular degeneration, capillaries, connective tissue growth and formation of fat cells on the left . Upper left (Slide n. 23, m. interosseius). -Observation XLII – syringomyelia. Down Left side (slides 24-26, 28-29, m. biceps brachii and slide 27, m. gastrocnemius). Observations XXXIV: amyotrophic lateral sclerosis. The histological study of XXXIV case was made in laboratories of Virchov and Klein. (The slides 23-29 were not described by authors of this article-author's note).

a review of 125 observations of progressive muscle atrophy and muscular tabes collected by Dr. Roth between 1874-1894. Dr. Roth subdivided muscular tabes into descending basic cases, ascending basic cases with onset in children and adults as well as the typical and atypical cases of basic muscular tabes and peripheral (distal) cases of muscular tabes.

4) Bibliography. Dr. Roth cites 1175 papers that he collected from the world literature from 1830 to 1893 and read in the original language, German, French and English. Of the most cited papers a very short summary of their content. Table 3, for example, presented, only two pages of references from 1830 to 1853 years.

The book "Muscular tabes" is very well illustrated with photographs and pictures of patients (performed by the painting-physician Dr. Heitzmann from Vienna) demonstrating various distinct phenotypes. Beside, detailed histological finding of muscle and central and peripheral nervous system are beautifully illustrated by color pictures. For example, we present from this monograph some photographs and drawings of patients with different types of muscular tabes (i.e. a myopathy) (Figs. 3-5, 8) and some histological slides (Figs. 6, 7).

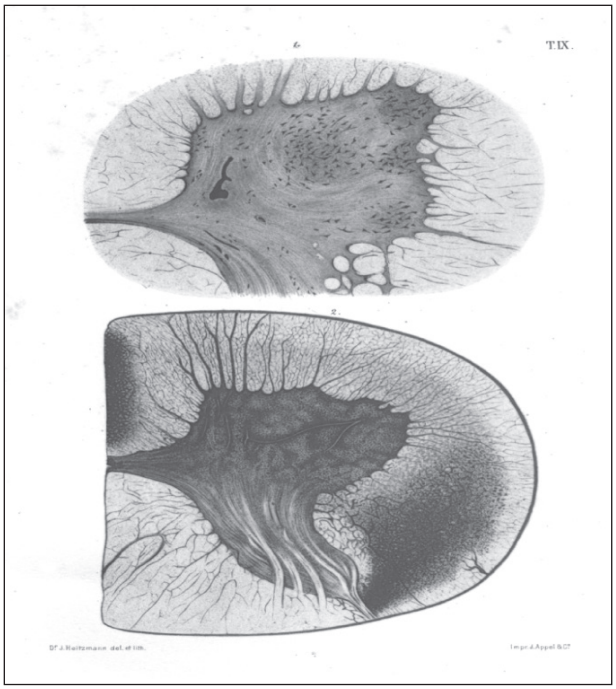
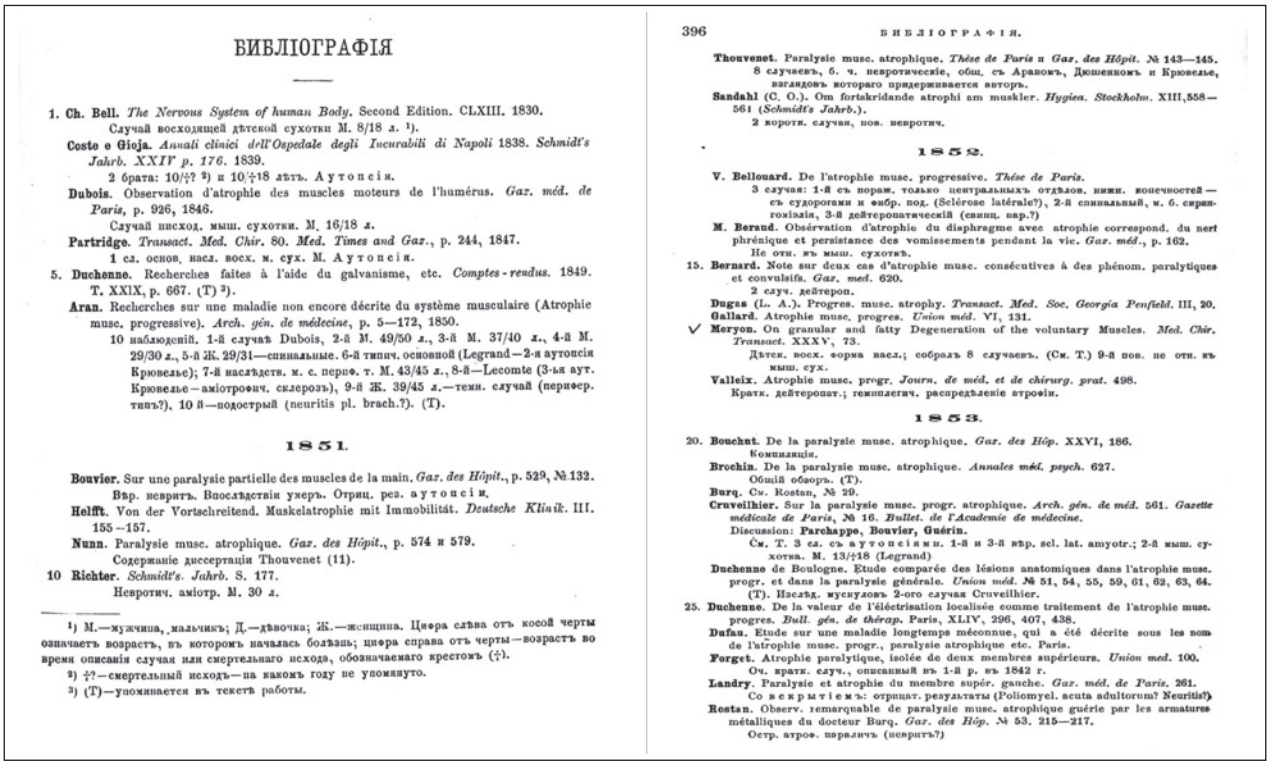


Figure 7. Up. Observation I. Basic muscular tabes. Abundance normal cells are shown in the anterior horn at the border between of C5-C6 nerve roots; **Down.** Observation XXXIV. Amyotrophic lateral sclerosis. Section of spinal cord at the C6 vertebra level. Sclerosis of the pyramidal tract and white substance around anterior horn. Absence of nervous cells in anterior horn and his hyaline infiltration. Atrophy of anterior and preservation of posterior nerve roots.

Distal myopathies in Roth's publications

There are four hereditary observations of peripheral muscular tabes (the same as a distal myopathy) in Dr. Roth's material: 2 observations with typical form (Observation XXXI, 1885, hereditary with severe weakness and atrophy of hand and feet, forearm and lower leg muscles (pp. 291-295); observation XXXIII, 1893**,** hereditary with severe weakness and atrophy of feet and lower leg muscles, only (pp. 298-301) and 2 atypical hereditary forms that Dr. Roth called as a "transitional forms of muscular tabes" (Observation XXVIII, 1886 (pp. 275-283); see description of this observation below) and observation XXIX, 1886, the sister of previous patient (pp. 283-286) (6). In all Dr. Roth's observations the disease began with affection of the lower leg muscles, extensors of feet and fingers. As an example, we present a very short description of two hereditary observations (a. with typical peripheral muscular tabes and b. with atypical, transitional form of muscular tabes).



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[1]) М.—мужчина, мальчикъ; Д.—дѣвочка; Ж.—женщина. Цифра слѣва отъ косой черты означаетъ возрастъ, въ которомъ началась болѣзнь; цифра справа отъ черты—возрастъ во время описанія случая или смертельнаго исхода, обозначаемаго крестомъ (†).
[2]) †?—смертельный исходъ—на какомъ году не упомянуто.
[3]) (Т)—упоминается въ текстѣ работы.

396 БИБЛІОГРАФІЯ.

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Table 3. Fragments of the references (1830-1853) including a total of 1175 articles with short summaries from Roth's monograph "Muscular tabes." Moscow 1895

a. Observation XXXIII, peripheral muscular tabes

Patient, of German origin, merchant, 39 years old, presented with symptoms of anxiety. The examination revealed a motor disorder in the lower limbs existing for a long time, but which the patients did not complain. The patient had 4 brothers and 4 sisters. The father and 1 of 4 sisters had a similar gait. The brother was normal. Clinical examination. Able to walk unaided without a stick. Main impairment was lack of dorsal flexion of the feet. Dr. Roth noted that the muscle wasting and weakness predominantly affected the anterior compartment of lower legs (posterior compartment muscles were minimal affected). Dr. Roth gave a very accurate and detailed analysis of the mechanisms of course steppage gait in this patient and noted that the leg muscle weakness was symmetrical. Small muscles of the feet were not affected. There was the contracture of the Achilles tendon. No fibrillations. The knee and Achilles tendon reflexes were absent. No sensory changes. All other muscles have not motor disturbances. Electric diagnosys: Roth himself made electric diagnostics of 22 muscles including feet and lower leg muscles and hands as well as mm. vastus lateralis and medialis and also peroneal nerves bilaterally of this patient. Results: no reaction of degeneration, no electric excitability in paralyzed muscles and its decrease in a weakened muscle. There were no electric signs of neuropathy. Dr. Roth concluded that this was a hereditary case of peripheral muscular tabes (i.e. primary distal myopathy, author's note).

In two other hereditary observations (brother and sister) in which the disease began with the atrophy and weakness of the distal part of legs and arms and later of the proximal muscles of the limbs, pelvic and shoulder girdle muscles involved in a less degree. Dr. Roth called this disease as a transitional form of muscular tabes.

b. This was an observation XXVIII (Fig. 8), which we present with very short Roth's remarks:

"A distinctive feature of this case is that… muscles of the pelvic girdle are affected less than thigh muscles, whereas muscles of the lower leg are completely gone, while muscles of the feet are minimally affected". "…On the upper extremities muscles of the scapular and shoulder girdle and upper arm muscles are much less severely affected than muscles of the forearm and hand. The lesions are symmetrical and muscles of the body are preserved". … "All tendon reflexes are absent"… "No fibrillation" … "Here we also see terminal atrophy, hypertrophy and lipomatosis of some muscles, characteristic of this disease, however, distribution of atrophy is quite distinctive". …

"No reaction of degeneration, no electric excitability in paralyzed muscles and its reduction in weakened muscles". There were not electric signs of neuropathy (Roth himself made electric diagnostics of 22 muscles in arms and legs and peripheral nerves (radialis, ulnaris, cruralis, peroneus, tibialis) on both sides of this patient-author's note)…"In our opinion this case represents transitional form of muscular tabes between the basic (central) type and peripheral type".

Figure 8. Observation XXVIII. (1886 year). Patient L.G., aged 32. Transitional form of muscular tabes (the same as atypical form of distal myopathy- author's remark).

In 1884 (1, 2) Dr. Roth at the International medical congress in Copenhagen presented a report "Issue of amyotrophic lateral sclerosis and its relation to progressive muscular atrophy" where he presented a classification of progressive muscular atrophies (without affection of the anterior horns of the spinal cord and nerves) which he called "muscular tabes" (the same as a myopathy) and described two main forms: 1. The most frequent – basic (central) form of muscular tabes – affecting muscles of the body and proximal segments of the extremities, pelvic and shoulder girdle. Often the disease has a tendency to generalize; 2. the second – peripheral form of muscular tabes – with muscular atrophy and weakness of the lower legs and in a lesser degree in feet, hands and forearms. In case of the peripheral form generalization of atrophy is less frequent.

In the review of progressive muscular atrophies in 1887 (1, 3) Dr. Roth makes the following conclusions based on his own data and literature:

1. Some cases of progressive muscular atrophies depend on primary affection of the spinal cord gray matter cells - amyotrophia spinalis progressive (protopathia Charcot). The cases described can be divided into two types according to the distribution of atrophy: a) Hand type – should not be called the Aran-Duchenne type and cannot be used as a prototype of "progressive muscular atrophy", because it is primarily (and may be exclusively) found in deuteropathic spinal atrophies; b) Shoulder-scapular type of Vulpian, the existence of which has not been proven yet.
2. The most frequent cases include progressive protopathic essential form of muscle atrophy (muscular tabes - the same as a myopathy) subdivided into two main forms according to the distribution of the disease: basic (central) or peripheral. The basic (central) form can be ascending or descending in children and adults. The peripheral form of muscular tabes deserves to be distinguished by the basic (central) form due to the presence of its peculiar clinical manifestations (7).

In the period 1884-1895, Dr. Roth published 4 hereditary cases of distal myopathy which he called "peripheral form of muscular tabes" (2 typical cases) and "transitional form of muscular tabes" (2 atypical cases) and presented (in 1884) this special type of muscular tabes i. e. distal myopathy as a nosological entity.

However, in Welander's survey of the literature (1884-1947) on distal myopathy, the name of Dr. Roth was not mentioned (8), although Davidenkov – a great expert on neuromuscular diseases in Russia – in his textbooks on hereditary diseases on nervous system (9, 10) wrote: "A special type of myopathy with a peripheral onset, recently called the "Naville type", deserves a separate description…Probably the G. family described by Roth (cases XXVIII, XXIX from his monograph), where brother and sister, born by healthy, probably related, parents, were affected, belong to the same group of cases" (9) and " Distal form, distinguished from all other myopathic forms by the onset in distal parts of upper and lower extremities with the following spread in the proximal direction to limb girdle and body muscles, is probably an independent form. Similar cases were described by Roth, Naville, Rimbaud-Giraud, Kolmaus und Sweerts, Kristen, Frommel and Van Bogaert" (10).

Conclusion

The development of studies on neuromuscular diseases, in particular myopathies, began in Russia in the second half of the XIX century and is tightly connected with the name of Vladimir Roth, professor of Neurology at the Moscow University. However his contribution to the study of the clinical pictures, etiology, pathogenesis and classification of hereditary myopathies and spinal muscular atrophies has remained underestimated in the world literature on neuromuscular diseases.

By a historical point of view, the name of Vladimir Roth should be remembered among the Authors who first described new clinical forms of myopathies, in particular those with a distal distribution of atrophy and weakness.

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