SYRINGO-MYELIA.

CLINICAL LECTURE DELIVERED AT THE PHILADELPHIA HOSPITAL.

BY WHARTON SINKLER, M.D.,

Neurologist to the Philadelphia Hospital, and Physician to the Philadelphia Orthopædic Hospital and Infirmary for Nervous Diseases.

[REPRINTED FROM INTERNATIONAL CLINICS, VOL. III., THIRD SERIES.]

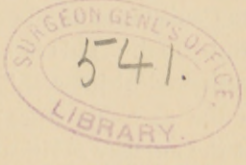
SYRINGO-MYELIA.

CLINICAL LECTURE DELIVERED AT THE PHILADELPHIA HOSPITAL.

BY WHARTON SINKLER, M.D.,

Neurologist to the Philadelphia Hospital, and Physician to the Philadelphia Orthopædic Hospital and Infirmary for Nervous Diseases.

[Reprinted from International Clinics, Vol. III., Third Series.]



SYRINGO-MYELIA.

CLINICAL LECTURE DELIVERED AT THE PHILADELPHIA HOSPITAL.[1]

BY WHARTON SINKLER, M.D.,

Neurologist to the Philadelphia Hospital, and Physician to the Philadelphia Orthopædic Hospital and Infirmary for Nervous Diseases.

GENTLEMEN,—I am going to present to you to-day two cases of syringo-myelia. This disease has been considered one of the rarities in medicine, and its name, syringo-myelia—from σύριγξ, "a tube," μυελός, "marrow"—derived from the Greek, means a cavity in the spinal cord. It has also been called hydromyelia, or hydrorrhachis internus. Now, the fact of there being cavities in the spinal cord has been known for a number of years. When D'Oliivier described them in 1826 he believed them to be of no clinical importance. Indeed, until quite recently cavities in the spinal cord were regarded as being of accidental occurrence, and not pathological. Even Erb, in his article on nervous diseases in Ziemssen's "Cyclopædia of Medicine," probably one of the most exhaustive treatises on the diseases of the nervous system, speaks of these cavities as being simply pathological curiosities; and in so recent a work as the "System of Medicine" edited by Pepper, the writers of the article on spinal diseases say, "among changes which occur probably at a late stage are the formation of cysts, large or small, either in the substance of the cord or consisting simply of dilatation of the central canal. This is known as syringo-myelia, and is exclusively of pathological interest."[2]

Within the past few years a number of cases have been recorded in which at the autopsy cavities were found in the cord, and on looking up the clinical histories it has been discovered that certain disorders of sensation and numerous trophic changes had existed during life. But few cases have been reported in which the diagnosis of syringo-myelia was made before death. I can find but five authentic cases reported in

---

[1] March 26, 1892.

[2] System of Medicine, vol. v. p. 1091.

which the diagnosis was verified post mortem. Two of these were reported by Schultze, one by Kahler,[1] and two by Déjérine.[2]

The disease is doubtless more common than is generally supposed. Schultze expressed the opinion that it is as common as multiple sclerosis.

The changes in the spinal cord are of two kinds. Either there is a simple dilatation of the central canal, which is probably a congenital condition and due to defective closure of the tube,—this is frequently known as hydromyelia, and is a condition not rare,—or there is a formation of new tissue in the cord in the shape of a gliomatous infiltration which is found in the gray substance. The glioma breaks down, and as a result cysts or cavities form in its substance. These cavities have a tendency to increase in size, and the encroachment on the sound tissue, together with hemorrhages which frequently occur, gives rise to new phenomena and changes in the old symptoms.

We should recall the fact that at about the sixth week of fœtal life the central canal of the cord is very large. This opening contracts by degrees and unites in the middle, the anterior portion forming the central canal, and the posterior part making the posterior fissure of the cord. Should the cavity, through some embryological defect, fail to contract, there remains an abnormally large opening in the cord, around which a glioma may develop. It is thought probable that most cases of syringo-myelia are the result of these congenital imperfections.

Cavities are most commonly found in the cervical or upper dorsal region of the cord. From the fact of these cavities being usually in the central part of the cord, the symptoms to which they give rise are principally those of a sensory character, and the trophic and muscular changes which take place are the result of pressure upon the anterior horns either by the dilated central canal or by the gliomatous growth itself. In some cases the glioma may start in the anterior horns.

On section the cord is found to contain in its substance a quantity of tissue of translucent appearance, which from its structure is evidently embryonal tissue in which the nerve-elements have failed to develop.

The degree of degeneration that takes place is sometimes enormous, the whole cord consisting simply of a large cavity with a thin band of tissue surrounding it.

The simplest form of syringo-myelia is a dilated central canal surrounded by gliomatous tissue, as seen in congenital cases. This is

---

[1] Charcot, Leçons du Mardi, 1888 and 1889, p. 488.

[2] Société de Biologie, February 8, 1890, and La Semaine Médicale, 1891, No. 6.

often abundant, and forms a distinct mass below the cavity. The posterior cornua are not uncommonly the seat of cavities which may extend throughout the length of the cord.

Cavities are often found post mortem in cases which presented no symptoms during life, and in young children these cavities apparently often exist without giving rise to symptoms.

Syringo-myelia occurs most frequently in persons who use their arms to a great extent, as butchers, tailors, etc. Pregnancy, injuries, and infectious diseases are said to give rise to it sometimes.

The symptoms of this disease are usually slow in developing, but, owing to the central location of the disease in the cord, there is pretty constant uniformity in the phenomena.

The disease may begin in one of two ways,—either with a predominance of the sensory symptoms or with muscular atrophy as the most conspicuous feature. In those cases beginning with sensory symptoms the disease is very much like the affection known as Morvan's disease, described by Morvan, a physician of Brittany.

In Morvan's disease a constant symptom is the extreme loss of sensibility in the upper extremities, so that various trophic changes, such as the formation of whitlows, occur without causing any pain whatever. It begins with pain and anæsthesia in the arms and hands, and muscular wasting occurs later. It was supposed by Morvan that the disease was peripheral in its origin, but the opinion now prevails that it is a variety of syringo-myelia.

In the cases in which muscular atrophy is present before the sensory symptoms, the wasting of the muscles usually begins in the shoulders and extends downward, and later in the course of the disease the various sensory disturbances show themselves. The sensory disturbances are impairment of tactile sense and pain sense, and, what is most characteristic, loss of heat sense. In many cases the tactile sense is preserved, and there is loss of power of perceiving heat, or it may be that only the ability to distinguish between heat and cold is lost.

The loss of sensation to heat is explained on the supposition that the posterior gray commissure is the path of painful impressions to the antero-lateral tracts.

In a number of cases of this disease which have been reported the first indication the patient had of there being anything the matter was that he would receive severe burns without being aware of them. One patient, reported by Dr. Vought,[1] burned his fingers frequently with

[1] New York Medical Journal, November 21, 1891.

the cigar which he was smoking. The other organs of special sense usually escape injury, but the eyes are sometimes affected. Sometimes the pupils are unequal, and there may be nystagmus; occasionally there is ptosis.

Trophic changes are very constant. The skin of the hands becomes thickened and myxœdematous in appearance. The hue is dusky or purplish, and frequently blebs and ulcers occur. The nails become thick, ridgy, and brittle. The temperature of the hands is usually low. Arthropathies are occasionally met with.

The diagnosis depends chiefly upon the association of the sensory and trophic changes, together with the muscular weakness and atrophy which accompany them, and the disassociation of thermal sensibility.

Cervical pachymeningitis presents symptoms which bear a striking resemblance to some cases of syringo-myelia.

In one of the cases which I shall show you to-day the appearance of the hands reminds one of a case of pachymeningitis. In the latter disease there is usually acute pain in the cervical region. The onset is rapid, and there is no disturbance of heat sense and pain sense.

Progressive muscular atrophy may be confounded with syringo-myelia, but in the former there are no sensory changes.

Peripheral neuritis is more sudden in its onset. There is more pain and hyperæsthesia; there is tenderness over the nerve-trunks, and, moreover, the whole body is usually affected within a short time.

Poliomyelitis is sudden in its onset; there is usually a history of recession of the paralysis from certain parts, and the muscular wasting is not associated with disordered sensation.

A tumor of the cord, especially if located in the central portion, may give rise to symptoms which it is impossible to distinguish from syringo-myelia.

Leprosy may be confounded with syringo-myelia, but in the former tactile sense is lost, as well as pain sense and heat sense; and, besides, in leprosy it is not long before the characteristic ulcerations and loss of substance take place.

MM. J. Déjérine and A. Thuilant have within a short time reported a case of syringo-myelia,[1] which is interesting in connection with the symptoms presented by the patient before you. Although thermic sensibility was abolished above 68°, it was preserved below that figure. The patient, D——, was thirty-four years of age. No nervous diseases in his ancestors. His father died at the age of fifty, from

[1] La Médecine Moderne, February 5, 1892.

a febrile affection. His mother died of a disease undetermined. In 1873 he apprenticed himself to a wine-merchant. His business was to cork the bottles, which he did by striking the corks with the palm of the left hand. From this resulted an unnatural callosity, the beginning of a phlegmon of the hand, which, eleven years ago, kept him in the hospital for one month. It is not stated whether the phlegmon was painful or not, but the history of the case contains the fact that in 1872 he received a burn of the chest, which he was unaware of until the next day. From this time—that is, about 1880—he noticed that his arm was becoming emaciated, and that there was some difficulty in flexing the hand at the wrist.

In addition, towards the end of 1881 he began to be conscious of a weakness of the lower limbs. This weakness gradually increased until 1882. He was then admitted to Bicêtre, suffering from complete paraplegia. For six years the patient had felt in the lower limbs pains, sharp and shooting in character. They occurred in paroxysms every two or three days, and lasted an hour or two.

The patient entered the infirmary February 10, 1890, for pulmonary tuberculosis in its first stages. There was then pronounced atrophy of the thenar, hypothenar, and interossei muscles of the left hand. In the left forearm the group of flexors was much atrophied, in the anterior region the round pronator alone being preserved. In the posterior region the extensors were greatly emaciated; the radii and the long supinator were, on the contrary, preserved.

Upon examination of the right hand a slight emaciation of the thenar and hypothenar eminences was noted, which attenuation dated back only a few weeks, according to the patient.

The left forearm was slightly flexed towards the arm. The hand (simian hand) was in hyperextension, the metacarpus forming with the wrist an angle of 100° or thereabouts, which position was due to the preservation of the radii and the long supinator. The first phalanges were in the axis of the metacarpal bones; the second and the third were very slightly bent towards the palm of the hand. The thumb was on the same plane as the other metacarpal bones; its digital phalanx was slightly flexed. In other words, the deformity of the left hand was actually the same as that of cervical pachymeningitis flexion.

The face was absolutely intact. The pupils were normal, and reacted to light and accommodation. No paralysis of the eye-muscles.

There was complete paralysis of the lower extremities, and there were exaggerated knee-jerks and ankle clonus.

Sensation.—The touch sense was absolutely normal in the trunk,

the face, and the upper and lower limbs, as well as on the buccal and lingual mucous membranes.

The pain sense, carefully tested, was normal everywhere.

Thermic Sensibility.—D—— had very acutely the cold sense from 17° to 39° over the entire surface of the body. At 59° he experienced a sense of coolness over the entire trunk, the lower limbs, the neck, the face, the arms and forearms, and the right hand; this sensation was less marked on the palmar surface of the left hand. These examinations were conducted with a flask provided with a thermometer and containing water at different degrees of temperature.

Insensibility to heat reached its maximum of intensity in the hands, particularly on the palmar surface, the forearms, and the lower half of the arm. In all these regions it was possible by means of a bottle of hot water (water at 194° or 203°) to produce an erythema, and even a bulla, without any resistance from the patient.

On the neck and trunk, at the level of conjunction of the upper limbs with the trunk, the application of water at 194° on the skin gave at the very first a sensation of moderate warmth to the patient. This sensation increased, and finally became painful if the contact was prolonged for several seconds. Heat below 149° was not perceived in these regions. In the lower limbs the heat sense was normal.

The patient died January 20, 1891, of tuberculosis of the lungs.

At the autopsy the following conditions were found. "The cerebrum, sensibly below its normal size, weighs ten hundred and ninety grammes. Nothing particular is to be noticed concerning either the cortex or the central nuclei. The lateral ventricles are not dilated. Cerebellum, pons, and medulla oblongata normal. Cervico-dorsal scoliosis at the left lateral convexity. Dura mater healthy. Extremely noticeable flattening of the marrow from the first cervical portion as far as the level of the lumbar enlargement. In the first half of its length the marrow has the appearance of a flattened ribbon; beginning at the upper half of the dorsal region it has the appearance of a fowling-piece with its two barrels in juxtaposition, and these two swelled halves are joined to each other in front by a small bridge of nervous substance, and at the back by the pia mater only. Under transverse section is discovered, through the entire length as far as the level of the lumbar enlargement, a large cavity, and it seems as though one were looking at fragments of macaroni, cooked and flattened. On inflating this cavity it is easy to determine its great size in proportion to the nervous substance which surrounds it. The cavity diminishes at the level of the commencement of the lumbar region, and here is noticed

the presence of gliomatous deposits. At the level of the lumbar enlargement a cylinder of glioma is again found in the centre of the marrow. The cutaneous nerves of the arms and the nerves going to the atrophied muscles showed neuritis."

Fig. 1.

William K., syringo-myelia. (From a photograph taken September, 1888.)

The history of the patient whom I shall show you first has been accurately prepared by my resident, Dr. G. H. Richardson, to whom I am indebted for his care in observing the case.

Case I.—William K., aged fifty-five years; a native of Germany; by occupation a butcher. Both parents are dead; one member of the family died of phthisis, and the rest of the family are well and healthy. He was admitted to the Philadelphia Hospital, July, 1886. He came to this country in 1868, was first employed as a hostler, and later as a butcher. He denies any specific or alcoholic history, and says that he was healthy until 1873, when he first noticed that the right arm was weak and heavy. The loss of power was particularly noticed in the shoulder group of muscles, which loss remained stationary for two years and a half, when the left arm began to be similarly affected. This was accompanied with increased disability in the right arm, and in time both became useless. The arms wasted, and there was a tendency to contraction of the extensors of the hands and fingers. He does not know whether he could recognize heat and cold at that time, but thinks he would have been able to notice the difference between hot and cold objects, and does not believe that he could have burnt his fingers without knowing it. His appetite is normal and his general health is unchanged. On admission he had complete loss of power in both arms and forearms, and this afterwards extended to the

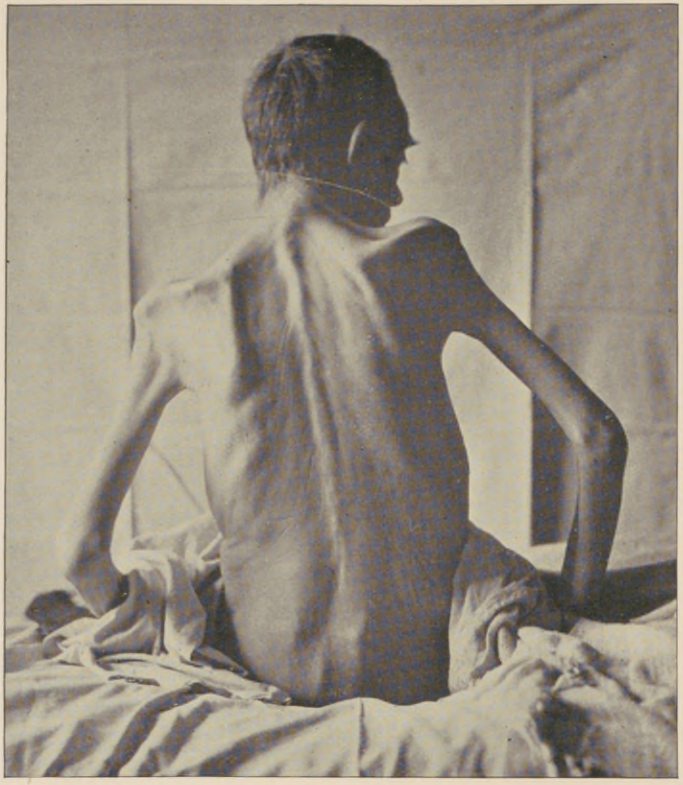


FIG. 2.—Wm. K., syringo-myelia. (From a photograph taken March, 1892.)

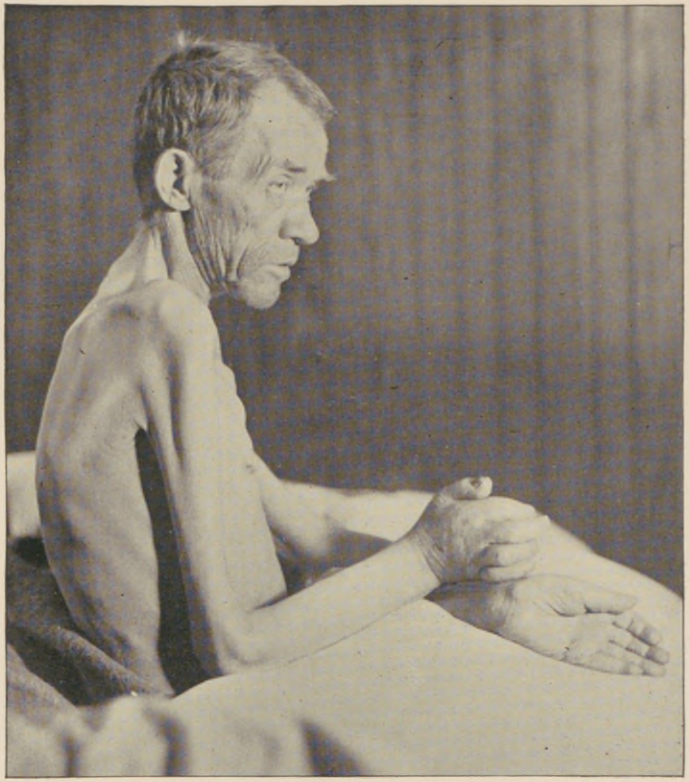


FIG. 3.—Wm. K., lateral view. (From a photograph taken March, 1892.)

flexors of the wrists and fingers. In 1891 he began to suffer from a violent diarrhœa, which has continued at intervals up to the present time. In November of 1888 he had trophic blebs on the first finger of the right hand, which disappeared without leaving any scar. About that time fibrillary tremors occurred in the muscles of the arm and forearm, with increased knee-jerk and the development of ankle clonus. In November, 1889, it was recorded that there was slight nystagmus, and tremulousness of the tongue on protrusion. The patient has gradually grown weaker and more emaciated, until he has reached the degree of extreme wasting which he now presents. For the past several weeks he has had severe diarrhœa, which accounts in some measure for the loss of flesh. A photograph taken in 1888, for which I am indebted to the kindness of my friend and colleague Dr. Dercum, shows the change in the man's condition. At that time, as you see, the body was well nourished, while the arms showed decided atrophy.

His present condition, as you see, is that of excessive emaciation and feebleness. Upon uncovering the body, what first attracts attention is the extreme wasting of the muscles about the shoulders and arms, almost every trace of muscular tissue having disappeared from the deltoid and supra- and infra-spinati muscles, and, in fact, all the muscles of the arm and forearm are wasted to a remarkable degree. He has no power of motion whatever in the upper extremities, with the exception of ability to extend the wrists and fingers. He extends the wrist of the right hand freely, and feebly extends all the fingers except the forefinger and thumb. In the left hand he extends the wrist fully, and extends all the fingers, but he is unable to close them. As a result of this long-continued action of the extensors, there is partial rigidity of the wrists in extension, so that they cannot be flexed without considerable force. They can be brought down only to the same plane of the forearm, and when the flexing power ceases the hands return to a position of extreme extension. I want to call particular attention to the extension of the wrists. It is the position which has been considered exclusively characteristic of cervical pachymeningitis, and it is precisely the same deformity that existed in M. Déjérine's case, quoted above.

Upon inspection, you will notice that the hands are rather darker in color than the arms; formerly they were deeply cyanotic, but since his recent illness they have lost some of the dusky hue. The fingers are thickened and clubbed at the ends, and the skin is glossy and purplish, and looks œdematous, but there is no pitting on pressure. There are numerous scars on the hands, the result of burns and other injuries.

Many of these burns were received without the patient being conscious of them, from his coming in contact with steam-radiators, etc. The finger-nails are curved, somewhat thickened, ridgy, and brittle, but especially the thumb-nail of the right hand, which is very much thickened; this, he says, was the result of a felon eight years ago, which was very painful.

I want to point out to you that there is a marked lateral curvature of the spine, with the convexity to the right, beginning in the cervical region and extending down to the twelfth dorsal vertebra like an S. There is some bulging of the right side of the chest, and the right sacro-iliac synchondrosis is more prominent than the left. There is no cyphosis; that is, there is no posterior angularity of the spine. You see occasionally fibrillary tremors in the muscles of the shoulders and arms; they are very noticeable in the infra-spinati. These fibrillary contractions are increased by a slight blow upon the muscles.

Reflexes.—All the tendon-jerks in the upper extremity are absent. When I tap on the biceps tendon there is no response, nor can I elicit any wrist-jerk. There is no epigastric, abdominal, or cremasteric reflex. The muscles all retain their irritability, and a tap on the belly of a muscle causes immediate contraction, and sometimes gives rise to slight fibrillary tremor for the time. This is strikingly marked in the pectoral muscles.

The knee-jerks are grossly exaggerated, and ankle clonus is present on both sides. You see that a tap on the patella gives a quick and excessive response on both sides, and ankle clonus is readily developed. Rectus clonus can be demonstrated on the right side. We have, then, in the legs exaggerated tendon-reflexes with ankle and rectus clonus, while we have in the upper extremities an entire absence of reflexes. The superficial reflexes are active in the legs; a little prick with the points of the compass gives rise to vigorous reflex contractions. The patient has no pain in the extremities, no formication, and no girdle sense. There has been a sensation of burning in the feet and legs when they are warmed by the clothing.

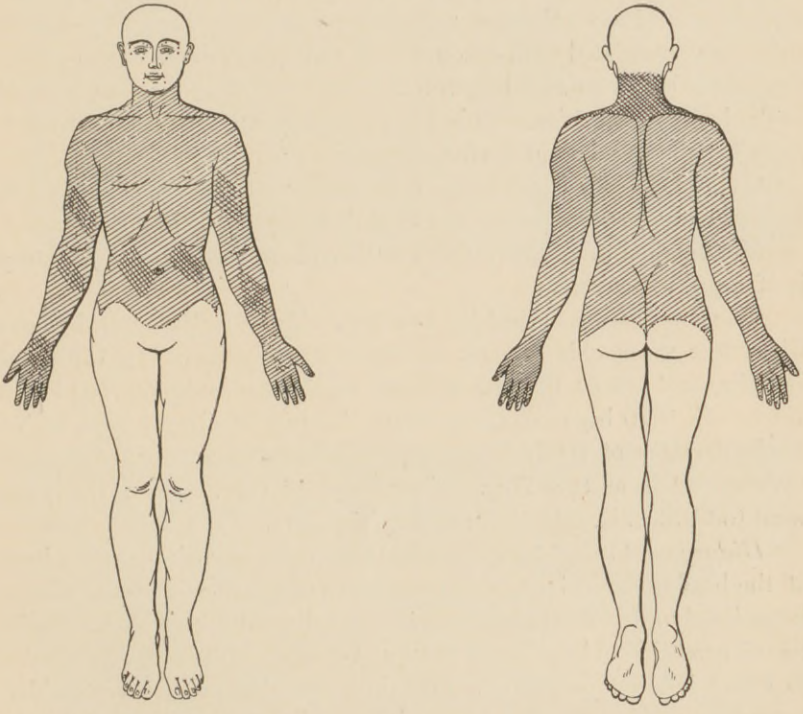
In the lower extremities we find considerable wasting, although this is chiefly from the loss of adipose tissue. There is but little muscular wasting in the legs.

He is able to walk without difficulty, but tires easily. His walk is somewhat peculiar, the right leg not being flexed at the knee in stepping forward. There is no incoördination in standing with or without the eyes closed.

The pathognomonic feature in this case is the addition of changes

in the temperature sense to the muscular atrophy. There is no change in the sensibility to touch or pain, but the sensibility to heat and cold is considerably altered. I have here two bottles, one of a temperature of about 40° F., the other quite hot. On applying these over the surface of the hands, it is found that both anteriorly and posteriorly there is loss of power to distinguish between hot and cold. In the face there is no loss of thermal sense, nor is there loss of this sense below the hips.

Fig. 4.



Case I.—William K. Light shading shows region in which there was confusion between heat and cold. In the dark patches neither heat nor cold was felt.

There are small areas on the anterior surface of the arms where the power to distinguish heat and cold is retained. In the greater part of the arms and trunk there is confusion between heat and cold; sometimes he says the bottle is hot when it is cold, and in some areas he can recognize cold but not heat. In other regions of the arms neither heat nor cold is recognized. The thermal anæsthesia is more marked on the right than on the left. (See Fig. 4.)

He can distinguish the compass-points correctly and promptly, and

distinguishes a sharp from a dull point. Painful sensations are readily perceived. The other senses seem unaffected. Smell and taste are normal, and the hearing is good. There is no disturbance of speech. His memory is good, and he is intelligent. There is no loss of bladder-control, and, notwithstanding the troublesome diarrhœa, the sphincter ani is intact.

We have, then, in this case these principal features: first, loss of power, beginning in 1873, nineteen years ago, with atrophy in the right arm, extending three years later to the left arm; later, the development of loss of thermal sense, without changes in tactile sense or pain sense, associated with extreme muscular atrophy and lateral curvature of the spine, reflexes lost in the upper extremities, but exaggerated in the lower extremities. This train of symptoms justifies us in pronouncing the case one of syringo-myelia.

Case II.—Thomas F., aged fifty-six years, white; birthplace, Ireland; occupation, teacher and book-keeper; single.

Mother died of phthisis pulmonalis; otherwise the family history is of negative interest.

Previous History.—Healthy as a boy. Came to the United States when five years old. Drank to excess when young. In 1863 had syphilis, and states that he suffered with secondaries in 1883 and 1884. In 1870 began to teach school, having previously kept books. Had an attack of erysipelas of the face in 1884, and suffered much with neuralgia at this time. He enlisted in the army in 1856, and went to California. He left the army in 1861.

History of Present Trouble.—In 1860 he was struck upon the back of the head with the butt of a gun; there was no headache nor vertigo immediately after this injury, but it was followed a year later by some loss of sensation in left side of head and face, which gradually extended to entire left side of body. Loss of power began in left shoulder, extending to left arm, left leg, and left side of trunk. Was treated in 1875 in a New Orleans hospital. At this time he found that he could not raise right arm to head. At no time has he been unable to walk, but there has been all the time a progressive loss of power in muscles.

He was admitted to this hospital in 1886. At that time there was complete loss of power in both arms and hands, although locomotion was not affected. The following note was made of his condition in 1886:

"Whole of left side is anæsthetic, it being possible in some parts to run a pin through the skin without the patient experiencing pain.

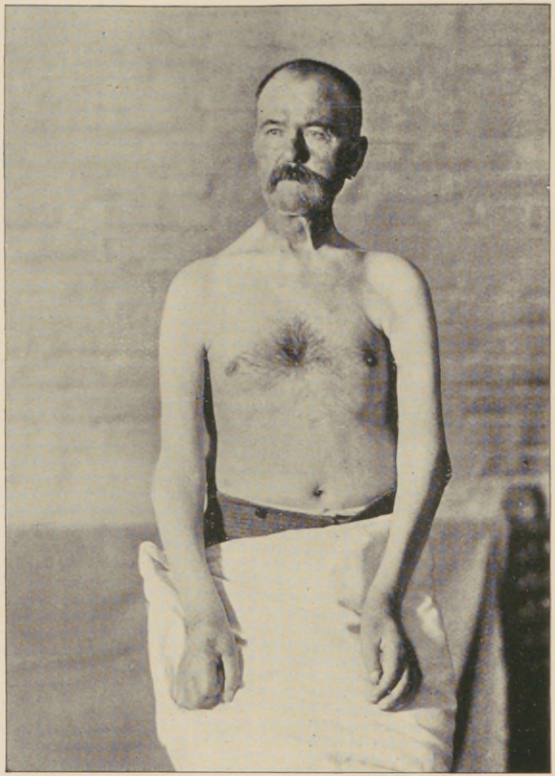


FIG. 5.—Thomas F., syringo-myelia.

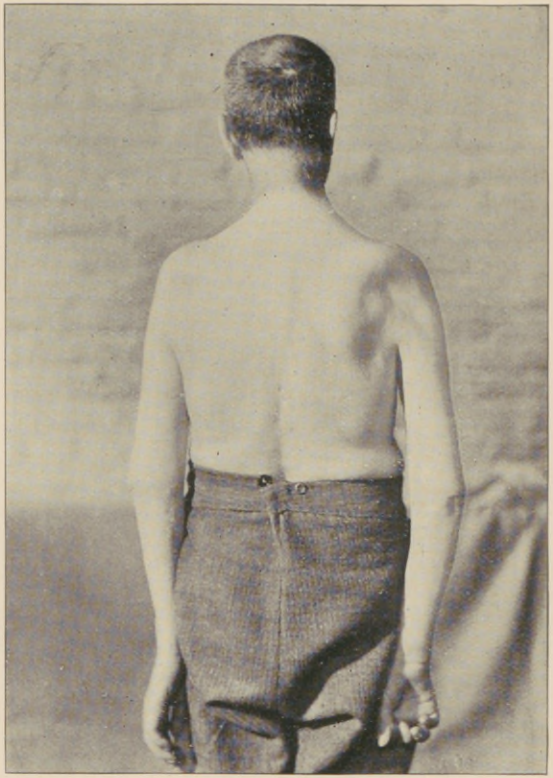


FIG. 6.—Thomas F., syringo-myelia.

On the right side sensation is blunted. Left pupil contracted and vision poor in left eye. Left arm can be raised but very slightly from the body, and forearm cannot be flexed more than nine degrees towards arm. Fingers are extended and can be scarcely moved. Loss of power complete in right arm and forearm. Right leg shows no loss of power. Left leg shows considerable loss of power and feels numb. Patellar reflexes diminished in both legs. Left arm emaciated, and hypothenar and thenar eminences of hand wasted. Infra- and supra-spinati much wasted."

March 23, 1888.—"Patellar reflexes exaggerated. Areas of anæsthesia over surface of left leg. Sensation in right leg retarded. Anæsthetic patches over both forearms. Sensation impaired in arms. Small areas of anæsthesia on left side of face."

Since admission to this hospital he has had a corneal ulcer of the right eye and several attacks of conjunctivitis.

Status Præsens.—Upon superficial examination, we find several scars on different parts of his body, the largest of which is one just above the elbow of the left arm and is the result of a burn caused by leaning against a steam-pipe while taking a bath. I mention that fact because in the history of cases of syringo-myelia there are many instances of burns having been received without the patient being aware of them at the time. The attitude of the patient is peculiar. He has a very marked stoop and is unable to place himself in an erect position, and there is a noticeable curvature of the spinal column, the convexity being to the right. I may observe in passing that in nearly all cases of syringo-myelia there is a right lateral curvature. There is also a prominence in the clavicular region on the right side. The face is flushed, and both arms, from the shoulders to the finger-tips, are deeply congested and cold. The shoulder-muscles are wasted, especially the supra- and infra-spinati and deltoid muscles. You notice the fibrillary tremor which occurs in these muscles. The arms hang flaccidly by the sides, and from the elbows down there is great congestion of the surface, the hands being particularly cyanosed, and the skin is dry. The hands are puffy, the skin of the hands and fingers being very much thickened and swollen, and are of a purplish, glossy appearance. There is some thickening of the finger-nails; they are rigid, very brittle, and grow slowly. Upon tapping over the tendons of the biceps and triceps and over the wrist tendons there is no response whatever. In other words, there is absence of tendon-reflexes in the upper extremities, as in the other case. The knee-jerks are grossly exaggerated on both sides, and there is ankle clonus. The plantar reflex is present. He walks, as you see,

fairly well, but there is a slight incoördination and clumsiness in his gait, which is probably due to weakness. There is considerable sway when his eyes are closed, so much so that he would fall if he were not held. He has absolutely no power to move the arms in any direction. He has slight power to flex and extend the right wrist, but on the left side he is absolutely unable to extend or flex the wrist. The muscles of the arms and forearms are wasted, and the thenar and hypothenar eminences are markedly atrophied. The fingers of the right hand are contracted and have the appearance of the "claw-hand." The fingers of the left hand are slightly contracted, mostly the third, fourth, and fifth. I have carefully gone over the electrical reactions, with the following results:

Electrical Reaction in Left Arm.—Muscles of shoulder respond to galvanic current ten milliampères. Deltoid muscle, K C C strongest. Supinator radii longus, K C C strongest. Extensors of hands give no response to the strongest current. Flexors of hands and fingers, A C C = K C C. The scapular muscles feebly respond, A C C > K C C.

In the right arm the current is felt more distinctly, but a much stronger current is required to produce the same degree of contraction than in the left arm.

In the deltoid, triceps, supinator, extensors, and flexors of hands and wrist, K C C > A C C. In the hand muscles, A C C = K C C.

In the left leg muscles, all respond to five milliampères, and the sensibility to the current is much more acute.

To the faradic current in the left arm, all the muscles of the upper arm respond except the deltoid.

In the forearm the supinator radii longus responds to no current.

In the right arm the reactions to the faradic current are the same as in the left, except that the flexors and the extensors of the wrist respond, but the intrinsic hand muscles do not.

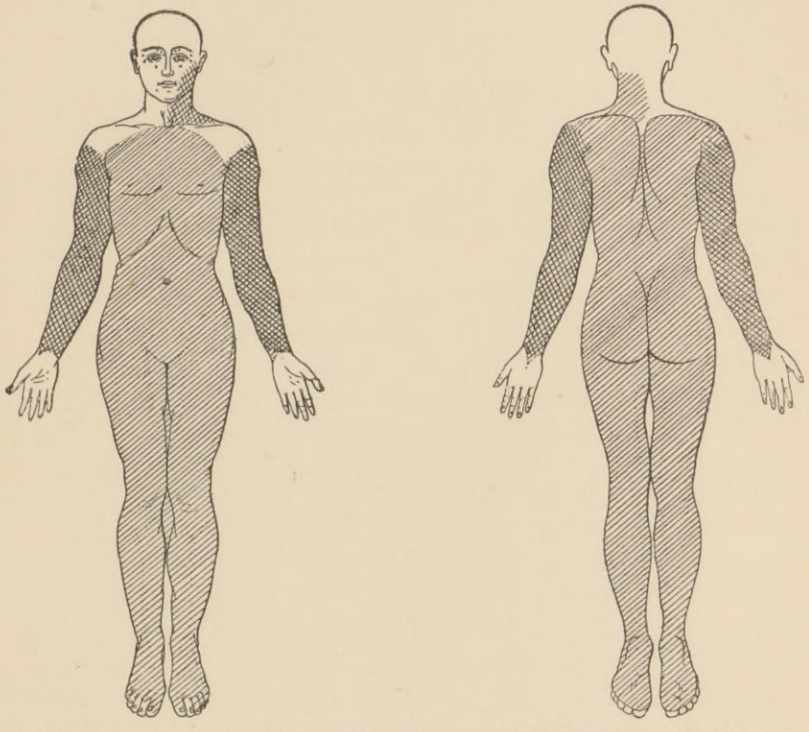
In both legs there is good response to the faradic current.

From the above it will be seen that in both arms the reaction of the galvanic current is reduced quantitatively, and in the muscles of the left arm there is distinct qualitative change. In the scapular and other muscles there is distinct reaction of degeneration.

There is impairment of the tactile sense in the hands, so that he cannot distinguish the compass-points nearer together than an inch and a half, and the sensation is delayed. There is considerable difficulty in differentiating between the sharp and blunt ends of the æsthesiometer. You see I have pricked his skin to a considerable depth with

the sharp end of the instrument without his perceiving pain, and he says that I am touching him with the blunt end. On the abdomen he cannot distinguish a sharp point from the finger-end, and has no idea of the distance between the points of the compass. The sensibility of the face is generally good to the compass-points and to light and deep touch. On the chest and shoulders he cannot distinguish between sharp and dull points, nor can he localize the touch accurately.

FIG. 7.



CASE II.—Thomas F. In area of light shading there is confusion between heat and cold. In the areas shaded dark the patient does not perceive either heat or cold.

Wherever the sharp point of the compass is placed on the skin, small wheals are formed in about a minute after the prick.

Temperature Sense.—I now test the ability of the patient to discriminate between heat and cold by using the hot and cold bottles. Along outer aspect of right forearm answers are confused as to heat and cold. Over upper third of forearm and over the supinator longus he calls hot "cold." Temperature sense in right hand is fairly preserved, also on the back of the left hand. Hot is called "cold" over the left arm

and forearm. On the left chest hot is called "cold." On the right chest patient states that the hot bottle feels a little warmer by contrast than on the left shoulder, and on the left neck and face he cannot distinguish heat and cold. On the right neck, face, and shoulder he appreciates the hot bottle, and, although perception is very much delayed, he calls it "warm." The right arm is slightly more sensitive than the left. He cannot distinguish heat from cold over surface of legs. (See Fig. 7.)

There are no general sensory disturbances. He does not complain of pains shooting through his body, of numbness, or of prickling, but is very sensitive to cold, and is always shivering and complaining of being cold. There is no girdle sense. He says the sweat has disappeared from the upper part of his body, but there is no change in the tear-glands.

The condition of the rectum and bladder is normal, speech and voice are good, and his general nutrition is good. There are no diseased joints.

Both of these cases, as you see, very closely correspond in their history and in their present condition. We have in both of them wasting of the muscles of the upper extremities, with inability to distinguish heat and cold, and with some changes in the sensory condition. In the latter patient there is impaired tactile sense, while in the previous case there is no change in the tactile sense or pain sense. These conditions you can readily understand by the variations which may occur in the pathological conditions of the cord. The larger the cavity the greater amount of disturbance there would be, and the location of it would, of course, affect to a considerable extent the sensory disturbances. As far as we know of the physiology of the cord the tracks for the conduction of temperature sense and pain sense are in the region of the central canal in the gray matter (in the posterior commissure), and, as a cavity in the cord usually occurs in the centre, you can see how it is that the disturbance of temperature sense is among the most frequent conditions in these cases.

Case III.—I want to show you this patient in connection with the patient whom we had before us first, as he also exhibits extreme muscular wasting, but with a different condition of things. The case is one of amyotrophic lateral sclerosis.

The man is fifty-four years of age; was admitted to the hospital seventeen years ago. As you see, he has great wasting in the upper extremities, almost as much as in Case I. You see that he has power of motion in both upper extremities, but the movements are made

feebly and with the greatest difficulty. He has also the fibrillary tremors noticed in the other patients. You know fibrillary tremors are common in many forms of muscular wasting. There are no sensory changes whatever. He has no loss in the power of appreciating heat and cold, and tactile sense is unimpaired. The muscle-jerks are very notably increased. A blow upon the wrist causes a sudden violent contraction of all the muscles of the arm. A tap on the biceps tendon at once causes flexion of the arm, and there is also increased triceps-jerk. The condition of both arms is the same. The appearance of the hands is strikingly different from that of the hands in the cases of syringo-myelia. The atrophy of the hand-muscles is extreme, but there are no trophic changes whatever. There is no thickening of the tissues and no change in the color; the fingers are very thin and bony, and the shortening of the flexor tendons which has taken place causes contraction of the fingers, the so-called "claw-hand." You see this condition in both hands. In the right hand there is some power of extension of the middle finger. There are exaggerated knee-jerks. There is atrophy of the muscles of the leg as well as of the muscles of the arms, and the patient is able to walk only with the aid of crutches. He has a curious prominence in the muscles of the neck, from the weakness of the cervical muscles. The cervical vertebræ are abnormally prominent from the fourth down to the sixth, but on aiding him to put his head into an erect position the projection disappears.

In syringo-myelia the prognosis is absolutely unfavorable, and the disease may last for a great number of years,—twenty to twenty-five. In fact, in one of these cases we have a history of the disease having continued since 1873, and in the other it probably has lasted as long. There is nothing that we know of in the way of treatment that has ever proved beneficial in these cases, and even in cases with a marked syphilitic history the use of anti-syphilitics has been of no value.

Subsequent History of Case I., with Autopsy.—The diarrhœa from which the patient had been suffering when he was brought before the class was relieved only for a time, and again became troublesome and exhausting. The patient gradually sank, and died July, 1892. There was no notable feature during the last few weeks of life, nor was there any impairment of intellect.

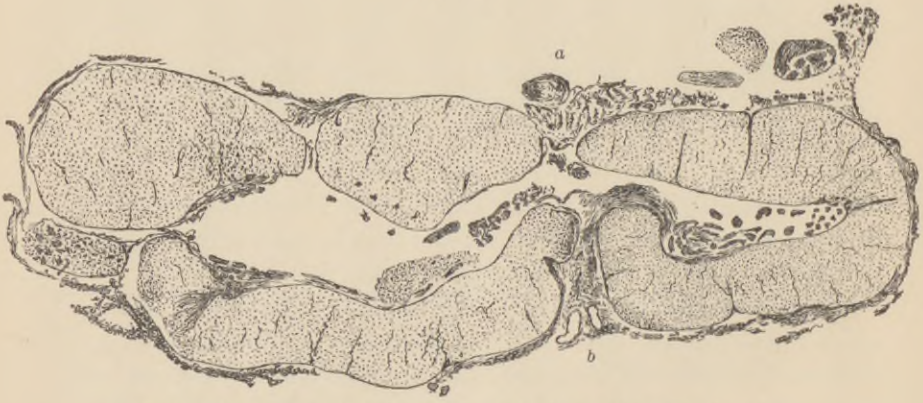
Owing to the fact that the autopsy had to be made under the most difficult circumstances, only the spinal cord was removed. It was through the efforts of my colleague, Dr. J. H. Lloyd, that the post-mortem was obtained. The medulla spinalis presented a large cavity, beginning in the cervical region and extending down to the beginning

of the lumbar portion, growing smaller from above downward. The cord was hardened in Müller's fluid and sent to Dr. C. W. Burr for examination, who has kindly made the following report :

" The dura is slightly adherent to the pia, and the latter to the cord in the cervical region ; otherwise the spinal membranes are normal. The cord is flattened antero-posteriorly from the upper cervical to the lower dorsal region, being most markedly so above, and assuming the normal form in the lumbar region. In the cervical region the cord measures one-half inch from side to side, and only one-eighth inch antero-posteriorly. The interior contains a centrally-placed cavity large enough to admit a quill. This cavity is lined throughout, except in the extreme upper portion, by a membrane stained light yellow by Müller's fluid. Its walls in the cervical region are very thin. It grows gradually smaller in cross-section, as sections are made lower and lower, until in the lower dorsal region only a small transverse slit remains. In the lumbar region nothing abnormal can be seen microscopically.

" *Microscopic Examination.* (Fig. 8.) *Upper Cervical Region.*— No gray matter is present except a small angle of the anterior horn

FIG. 8.



CASE I.—UPPER CERVICAL REGION.—*a*, anterior fissure; *b*, posterior fissure.

of the left side. A few shrunken motor cells, without processes, mere ghosts of cells, are found in this area. The commissures have disappeared. There is no sign of the normal central canal. The cavity is without distinct lining membrane. Its walls are made up of white matter, which is for the most part normal, with here and there microscopic spots of degenerated tissue. The cavity is prevented from com-

municating with the anterior and posterior longitudinal fissures simply by the folds of the pia mater.

"*Lower Cervical Region.* (Fig. 9.)—The gray matter is still absent. The cavity at this level, and below as far as it extends, is lined

FIG. 9.

CASE I.—LOWER CERVICAL REGION.—*a*, anterior fissure; *b*, posterior fissure.

by a tissue made up of many small round cells, with a dense interlacing of fine fibres. There are very few blood-vessels, and most of those present are at the periphery. This tissue is not distinctly marked off from the surrounding white matter, but sends prolongations into it.

"*Mid-Dorsal Cord.*—With a low power the anterior and posterior horns are distinctly visible, but the intermediate gray substance is pushed to either side by the cavity. A thin strip of the anterior commissure persists. With a high power a few normal gray cells are visible, but the greater number are shrunken and without processes. The white matter is normal, except a band of degenerated tissue along either side of the great longitudinal fissure and a very slight area in the corneal pyramidal tracts. The posterior nerve-roots are normal; in the anterior there are areas of degeneration.

"The lower dorsal and lumbar regions of the cord are normal, except small areas of degeneration in the lateral pyramidal tracts."

The following case was under the care of Dr. S. Weir Mitchell, at the Philadelphia Infirmary for Nervous Diseases, a short time ago. Through Dr. Mitchell's courtesy I am permitted to give the report of the case.

CASE IV.—The patient was a man twenty-seven years of age, who had enjoyed excellent health up to the age of twenty-one years. He

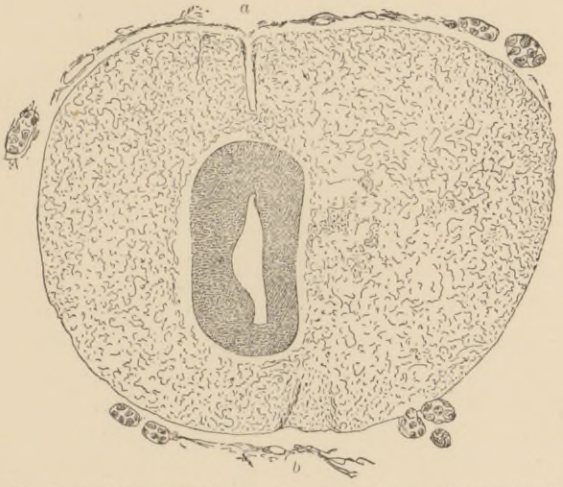
had never had syphilis, and was temperate in his habits. About six years before coming under observation he had an attack of intense pain in the back of the neck, which lasted about six weeks, and the patient was obliged to take large amounts of morphia for the relief of the pain. Immediately after this attack he had typhoid fever, and during convalescence he had a second attack of pain in the neck, lasting two weeks. The character of the pain was cutting and severe; it was continuous, but became worse at night, and extended to both shoulders and down the arms as far as the elbows. He has had several attacks like this since. About four years later he had a febrile attack of some kind which confined him to bed for five weeks. A few months later the right arm began to grow weak, and in about a year loss of power appeared in the right leg. There was no headache, no loss of control of the sphincters, and the general health was very good.

On admission to the Infirmary, his condition was as follows. The patient was well nourished and had the appearance of fairly good health. The right arm was very feeble, in fact almost entirely powerless; he could abduct the arm by the deltoid; could feebly flex the forearm on the arm; could flex and extend the wrist; flexion and extension of the fingers were feeble, and the thumb could only be slightly abducted. Pronation and supination were fair. The arm was one-fourth of an inch smaller in circumference than the left. There was no marked interdigital wasting, but the right thenar and hypothenar eminences were atrophied. All the muscles of the arm were flabby. The right pectoral muscle was smaller than the left. Dynamometer: right, five; left, fifty-nine.

The examination as to sensation was not made with any great accuracy, but in a general way it was found that there was loss of touch sense and pain sense in the right hand, and that there was perversion of temperature sense in both hands. There were no trophic changes, the nails were not ridged, and there were no blebs or ulcers on the hands. The knee-jerks were exaggerated, and ankle clonus was present. Achillis jerk and plantar reflex were present on both sides. Cremasteric and abdominal reflexes were absent. There was exaggerated muscle irritability on both sides. There was no deformity of the spinal column, but on pressure over the fourth cervical spine there was tenderness. The symptoms all pointing to a growth in the cervical region of the cord, it was decided to perform the operation of laminectomy. The cord was accordingly exposed in the cervical region, but it was not practicable to remove the growth. The patient died on the eighth day after the operation.

Autopsy.—The spinal canal in the cervical region is completely filled by the swollen and distorted cord. The dura and pia are adherent to it. On section, the interior of the cord is found to be occupied by a tumor, centrally placed, having pushed the anterior horns before it, and covered posteriorly by a thin band of white matter. It is reddish in color and infiltrated in spots with blood. It extends as a solid mass from the highest cervical region down to the fifth cervical nerve, at which point a central cavity appears, which, surrounded by the tumor, extends to the fourth dorsal nerve. On microscopic examination the tumor proves to be a glioma. There is marked degeneration of the anterior and crossed pyramidal tracts. There is marked acute myelitis at the seat of operation. The accompanying figure (Fig. 10) shows well the gliomatous mass surrounding the central canal in the

FIG. 10.



CASE IV.—LOWER CERVICAL REGION.—*a*, anterior fissure; *b*, posterior fissure.

lower cervical region. The proper structure of the cord is destroyed by the acute myelitis.

I have appended an account of this case because of its interest in connection with the others. In Case I. we had a typical illustration of a syringo-myelia resulting from a congenitally dilated central canal with subsequent development of the gliomatous growth around the cavity. In Dr. Mitchell's case the glioma was the primary lesion, and the cavity in the centre was the result of degeneration and absorption of the glioma.

I wish to acknowledge my indebtedness to Dr. Alfred Stengel for the microscopic drawings.