SYRINGOMYELIA.

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FROM

THE MEDICAL NEWS,

July 22, 1893.

[Reprinted from THE MEDICAL NEWS, July 22, 1893.]

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IN a recent work[1] on diseases of the nervous system it is stated that "it is now quite a fashion in this country to report so-called cases of syringomyelitis." It it be a fashion it is one that seems to have but few votaries in the United States. For, since the publication of the first case by Dr. Starr,[2] certainly not many more than a dozen American cases have been recorded. The paucity of our literature on syringomyelia forms a striking contrast to the voluminous records of European authors on the subject. Schultze, Déjerine, Charcot, Hoffmann of Heidelberg, and many others have been enabled to base their studies of this disease on such abundant material as to justify the assertion that it is scarcely less common than disseminated sclerosis. Hoffmann alone recorded five cases in 1891,[3] and recently, in a very elaborate essay,[4] added a report of no fewer than eighteen.

Assiduously as syringomyelia has been studied for

[1] "A Treatise on Nervous and Mental Diseases," by Landon Carter Gray, M.D., 1893.

[2] American Journal of the Medical Sciences, May, 1888.

[3] Volkmann's Sammlung, 1891, No. 20.

[4] Deutsche Zeitschrift für Nervenheilkunde, Bd. iii.

several years past, the interest in it does not seem to flag. Its pathology and the relations of the structural changes in the cord to the clinical manifestations are still subjects of laborious research. The diagnostic difficulties arising from its resemblance to certain other diseases render its study especially important. The question of its identity with Morvan's disease has not yet been answered to the satisfaction of all; and its resemblance to anesthetic leprosy has called forth the sensational assertion that syringomyelia and anesthetic leprosy form "a single morbid entity," varied only by climate or the hygienic conditions of the patient.[1]

In view of the sustained interest in all that concerns this remarkable malady, the report of the following case may not be unacceptable.

The patient, K. F., is an unmarried woman, twenty-five years old, a native of Boston, Mass., of Irish parentage, and resident in California since her second year. She presented herself at the San Francisco Polyclinic on the 13th of January last, with the statement that her hands were wasting, and added that she was insensible to pain or heat in the upper extremities. The disease began about six or seven years ago with a prickling sensation ("like when the foot is asleep") in the right deltoid region; this sensation gradually descended to the elbow, and thence down the forearm to the wrist;

[1] Zambaco: Gaz. hebd., 1891. Quoted by Marestang, Revue de Méd., 1891, p. 783. In La Semaine Médicale, 1893, June 10, Zambaco maintains that not only Morvan's disease and syringomyelia, but also sclerodermia, sclerodactylia, aïnhum, morphea, and local asphyxia with symmetrical gangrene, are "nothing but varieties of one and the same prehistoric disease, old as the world, leprosy."

it has never been present on the left side. The patient found relief from the distressing feeling by directing her companions to strike her on the fleshy part of the right arm, and the more vigorous their blows the greater was the relief. She did not cease to be troubled in this way until about a year and a half later; in the meantime she had continued her occupation as a seamstress. While engaged in sewing and ironing she often sustained injuries to her hands which, although severe, entailed no suffering, and thus attracted her attention to the impairment of her sensation. She often burned herself and did not notice it until it had been pointed out to her; once she put her hand into very hot water and scalded it, but suffered no pain. Her fingers finally became very sore, so sore that she thought "they must rot off," and some of the nails fell off two or three times. About four years ago she abandoned her occupation and has since then injured herself less. While undergoing electric treatment the regulation of the strength of the constant current was occasionally left to the patient by her physician, and there being no painful sensation to warn her, a large running sore formed in the palm of the left hand, which healed very slowly, but did not distress her. The disturbance of sensation preceded the wasting of the hands. The right hand wasted first; there was twitching in the fingers and especially in the thumb, in which there was a sensation "like the fluttering of a bird trying to escape." The right hand had wasted to its present condition when (about three years ago) the atrophy of the left commenced. The patient is often annoyed by a jerking of the left lower limb; the jerking consists in violent flexion of the thigh and extension at the knee; it occurs most frequently and is most violent when the patient is recumbent

and, according to the testimony of the mother, during sleep. It sometimes occurs while she is sitting, and is then arrrested by standing. Fatigue after much walking intensifies the jerking. Occasionally there are cramps in the great toe of each foot, in which the toe is forcibly extended. Complaint is further made of pain at the back of the neck and in the palm of the hand.

Nothing was elicited indicating a family tendency to nervous diseases; but there seems to be a tuberculous taint, as the patient's father and sister and a maternal uncle are said to have died of pulmonary tuberculosis.

The patient is at present small and anemic. In both hands there is advanced muscular atrophy. The interossei are wasted, the absence of the interosseus primus being particularly noticeable. The thenar and the hypothenar eminences are flattened, the right thenar less than the left. The distal and middle phalanges are flexed, and the proximal phalanx is extended, whereby the "claw-hand" is produced, more markedly, however, on the right than on the left side. Flexion and extension at the wrist are preserved; but the atrophy of the intrinsic muscles of the hands is so great as to render the left hand practically useless and to seriously impair the use of the right. The advantage of the right hand over the left is due to the better condition of the opponens and the adductor pollicis in the former, and of the muscles in the right forearm. Abduction, extension and flexion of the thumbs are fair. Both forearms are thin, but the left is thinner than the right, the latter measuring 13 cm., the former only 12 cm. in the middle. The muscles of the upper arms and shoulder appear a little flabby, but not more so than might be expected from the patient's general nutrition; their functions are in-

tact. The right arm measures 19.5 cm., the left 19 cm. around the middle. There is no atrophy of the trunk or of the lower limbs.

Fibrillary twitching is seen in the muscles on the inner side of the left forearm and in those of the hands; there is also a jerky tremor of the hands. As to the electric reactions, the muscles supplied by the musculo-spiral nerve react on both sides equally well and normally to direct and indirect galvanic and faradic excitation. The right median and ulnar nerves and the muscles of the forearm to which they are distributed respond fairly well but less promptly than the extensor muscles; in the same nerves and muscles of the left limb there is a considerable diminution of excitability by either current, without modal change, and the reduction of irritability is greater in the ulnar than in the median nerve and muscles. There is no typical reaction of degeneration in the small muscles of the hands; in accordance with what has been noted of the comparative preservation of motor power in the right thumb, contractions of the adductor and the opponens pollicis are obtainable by direct excitation or by excitation of their nerves at the wrist, with not very strong currents; in the other muscles of both hands the electric reactions are either greatly diminished or abolished.

The tactile, muscular, and stereognostic senses are preserved in all parts.

While the muscles of the upper extremities were being tested by faradism the patient gave no indication of pain even when strong currents were applied with a very small electrode. Other tests, however, disclosed that the analgesia was not absolute in these parts, deep punctures and strong pinches, particularly in the right hand and shoulder eliciting evidence of painful sensation. (Figs. 1 and 2.) The

analgesia is more profound throughout the left superior extremity than upon the right.[1] It is most pro-

FIG. 1.

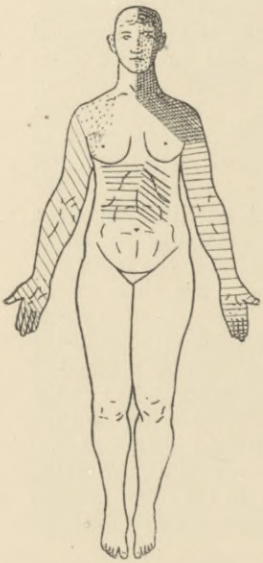
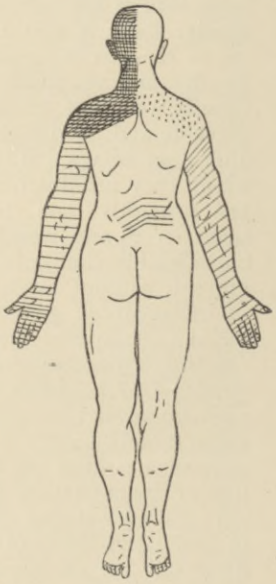


FIG. 2.



Analgesia.

The dots, slanting lines, horizontal lines, and crossed lines, denote different degrees of sensory impairment, the intensity increasing in the order in which they are mentioned.

found on the left side of the head (except a part of the face), on the left half of the neck and the left shoulder. It is slight, but still evident in compari-

[1] By a lamentable oversight the four illustrations produced in the reprint of the article were omitted from the article as it originally appears in THE MEDICAL NEWS.

son with the other side, on the median half of the left side of the face and on the left side of the

FIG. 3.

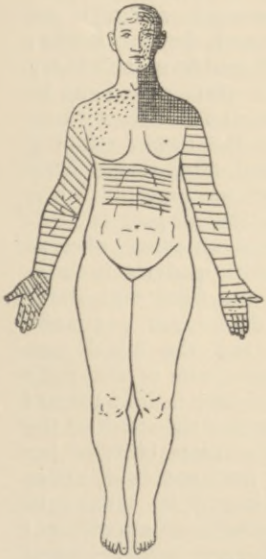
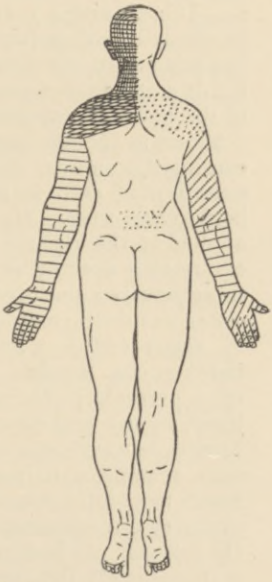


FIG. 4.



Thermo-anesthesia.

The dots, slanting lines, horizontal lines, and crossed lines, denote different degrees of sensory impairment, the intensity increasing in the order in which they are mentioned.

mouth and tongue. The upper and lower regions of the chest are slightly analgesic, while the breasts are normal or nearly so. Ammonia held to the nostrils produces equal irritation on both sides. The analgesia in the hands is not as great as the statements of the patient would lead one to believe.

The ulnar nerves are not sensitive to strong pressure at the elbows.

The insensibility to heat and cold coincides for the most part in distribution and degree with the anaglesia. (Figs. 3 and 4.) In the right hand it is less than in the left, and is far from absolute, although the impairment of the thermic sense is beyond doubt; but it is less than would be inferred from the history of the patient's experience. A piece of ice can be retained in the left hand almost indefinitely without causing discomfort, whereas in the right a stinging sensation of cold is announced after it has been held a short time. Water which was quite hot when applied to the right hand for the purpose of electric testing was not felt to be at all warm, whereas subsequently a test-tube filled with moderately warm water was distinguished from one filled with water of about 60° F.; and the distinction was made between the wooden handle and the metal part of a percussion hammer, when held in the right hand, by the temperature of the metal. The patient herself, however, when informed of the result of the tests, remains convinced of the insensibility of her hands to painful impressions, heat and cold, under ordinary circumstances. A piece of ice taken into the left side of the mouth is recognized as cold, but is immediately felt "several degrees colder" when shifted to the right side. An analogous difference obtains in regard to warm water. Dissociation of the temperature-sense itself was not observed.

Trophic changes are not pronounced apart from the muscular atrophy. A few insignificant scars indicate the sites of former injuries. The nails are not abnormal.

Nor are the vasomotor disturbances marked. On the chest and back the vasomotor irritability seems to be increased; this is especially noticeable

when the finger is passed along the row of spinous processes. The hands are, as a rule, subjectively and objectively quite cold.

The iris-reflex is normal in either eye. The left conjunctival reflex is absent; the right one is present. The pharyngeal reflex is preserved. A brisk jaw-jerk can be obtained. The triceps-tendon reaction can be elicited in either arm. On percussion the supinator longus on each side contracts actively. No periosteal or tendon-reflexes exist at the wrists. The epigastric reflexes are absent. The knee-jerks are both much exaggerated, and are equal on the two sides. The tendo Achillis reflexes are quite active. The front-tap contraction is obtainable on either leg. Ankle-clonus was elicited on the left side at one examination, but has not been produced since. The plantar reflexes are absent.

Hearing, taste, and smell are normal. The eyes were kindly examined for me by Dr. Pischl. V. with $+ 1.5 = 6/VI$ in both eyes. The field of vision determined for white and colors is normal. The ophthalmoscope reveals in the right eye a band of opaque nerve-fibers encircling the inner half of the papilla and equal in width to three-fourths the diameter of the disc. In the left eye a small patch of such fibers is seen at the upper, and another at the outer, edge of the disc. The papilla appears a little redder than is normal, which is probably due to the contrast with its white environment. There is a slight narrowing of the left palpebral fissure, its width being 8.5 mm., while that of the right is 9.0 mm.

The secretion of tears is more abundant in the left eye than in the right. As has already been mentioned, the left conjunctival reflex is abolished. The pupils react to light and in accommodation, and are equal. There is no nystagmus.

The spinal column is not tender to pressure. There is no scoliosis. On passing the finger down the row of spinous processes a peculiarity is felt in the shape of some of them in the lower dorsal and upper lumbar regions. The ninth dorsal spinous process is normal; the tenth is clubbed; the eleventh and twelfth dorsal and the first lumbar are clubbed and bifid; the second lumbar is like the tenth dorsal and the rest are normal.

The treatment since the patient has been under my care has consisted chiefly in the exhibition of iron. Her general health has improved, and she is now free from the pain in the back of the neck, but still complains occasionally that the hands ache.

The combination of symptoms such as have been described in the foregoing notes, consisting in progressive muscular atrophy of the Duchenne-Aran type and dissociation of sensory impressions, stamps this as a classical case of syringomyelia. If it be at all necessary to differentiate this malady from Morvan's disease, the absence of painless whitlows will be conclusive of the diagnosis.

How close the clinical resemblance between syringomyelia and anesthetic leprosy may be, despite the difference in pathology, is evident from the descriptions and illustrations furnished by Marestang. Dissociation of sensation has been observed by this author in leprosy; but only in one case among over one hundred was there retention of tactile sense with analgesia and thermoanesthesia, while in two cases there was retention of pain and of the thermic sense with tactile anesthesia. On the other hand, in syringomyelia loss of sense of contact has not infrequently been met with (which is not surprising,

in view of its pathology). Such dissociations, however, are exceptional in leprosy, and the preservation of tactile sensation in syringomyelia is the rule. The muscular wasting in the extremities is similar in the two diseases, and even the atrophy of facial muscles, often seen in leprosy, has been observed in four cases of syringomyelia. Nevertheless a proper appreciation of what is frequent and what is rare in each, or the recognition of some characteristic of leprosy, such as anomalies of pigmentation, loss of hair, nodular thickening of nerves, the bacillus, etc., will generally lead to a diagnosis. But that errors may occur has been recently shown by Pitres[1] in his report of the case of a man who, presenting trophic disturbances and the sensory dissociation of syringomyelia, was for a long time considered to be suffering from that disease until the detection of Hansen's bacillus in a cutaneous nerve proved him to be affected with leprosy. This man had lived for three years in Martinique, and it is in countries which are in frequent communication with endemic foci that special caution is required. The Pacific coast is exposed to the occasional intrusion of lepers from Hawaii or China, and in the public eleemosynary institutions of San Francisco there are now thirteen lepers. But in the present instance, besides the absence of anything in the patient's antecedents likely to excite a suspicion of leprosy, and the lack of any symptom characteristic of that disease, the

[1] Gaz. des Hôpitaux, 1892, No. 137; abstract in Neurolog. Centralblatt, 1893, p. 102. See also Kalindero: Wiener med. Presse 1892, No. 39.

insensitiveness of the nerves to pressure, the fact that the disturbance of sensation is less profound at the peripheral parts of the upper extremities than in certain other regions, the fibrillary twitching, the augmented knee-jerks and the narrowing of the left palpebral fissure, decide in favor of syringomyelia.

Hysteria, that arch-mimic of organic nervous disorders, has also been known to simulate syringomyelia. Cases of this kind have been reported by Charcot[1] and by Rossolimo.[2] The latter tells of a girl, aged twenty, with interosseal paresis, analgesia, and thermoanesthesia, but only slight diminution of tactile sensation, in her hands. This combination of symptoms had recurred six times, in the course of a year and a half, at regular intervals, and disappeared promptly after an hypnotic séance. To discuss the possibility of the hysterical nature of the case now under consideration would appear like raising objections to a diagnosis simply for the pleasure of refuting them. Nor does it seem necessary to enumerate the features distinguishing cervical hypertrophic pachymeningitis from syringomyelia, the anesthesia in the district of cranial nerves alone sufficing to exclude the former.

The possibility of diagnosticating syringomyelia during life is not yet universally conceded. In the discussion of a case presented before the Neurological Society of New York, in 1890, the president, Dr. L. C. Gray, said: "The only way to make a diagnosis of syringomyelia was to make an autopsy."[3]

[1] Leçons du Mardi, 1888–1889, p. 518.

[2] Neurolog. Centralblatt, 1892, p. 493.

[3] Boston Med. and Surg. Journal, 1890, vol. cxxiii, p. 570.

And the same view is expressed in his recent *Treatise on Nervous and Mental Diseases*,[1] although he acknowledges "that we are on the right track and that we shall be able to make a diagnosis in the early future." The fact is, however, that the diagnosis has been repeatedly made *intra vitam* and verified *post mortem*. Charcot[2] stated in 1889 that Schultze had twice and Kahler once found the diagnosis justified at the autopsy. Moreover, Déjerine,[3] in 1890, and again in 1891,[4] and with Sottas,[5] in 1892, reported cases in which the diagnosis of syringomyelia, which, during life, had been based on the muscular atrophy and the sensory dissociation, was corroborated after death. Verified diagnoses have also been recorded by Gyurman,[6] Critzmann,[7] and Bernstein.[8] In a report of a case of syringomyelia by Hughlings Jackson and Galloway[9] mention is made of an instance in which a diagnosis of Ferrier's was proved to be right at the necropsy. In Hoffmann's[10] Cases IX and XIV the disease had been recognized during the lifetime of the patients. Finally, the reviewer of Dr. Gray's book in the *American Journal of the Medical Sciences* for

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[1] Pp. 267 and 268.

[2] Leçons du Mardi, 1888–1889, p. 488.

[3] Société de Biologie, February 8, 1890.

[4] La Semaine Médicale, 1891, No. 6.

[5] See Neurolog. Centralblatt, 1892, p. 710.

[6] Abstract in Neurolog. Centralbl., 1891, p. 504.

[7] Essai sur la Syringomyélie, 1892; Neurolog. Centralbl., 1892, p. 708.

[8] Neurolog. Centralbl., 1893, No. 3, abstract.

[9] Lancet, Feb. 20, 1892.

[10] Deutsche Zeitschrift für Nervenheilkunde, Bd. iii.

March, 1893, states that two cases of syringomyelia have been diagnosticated *intra vitam* in Philadelphia, and that the autopsy confirmed the diagnosis; the cases had, however, not yet been published. One of them is, perhaps, identical with that very recently recorded by J. H. Lloyd.[1]

This list makes no pretensions to being exhaustive, but it will suffice to show that the diagnosis may be ventured with some degree of certainty. Not all of the cases conformed to the most frequent type, but the pathognostic value of progressive amyotrophy with impairment, and generally dissociation of sensation, was sufficient to guide the observers to the recognition of the disease. The assumption of syringomyelia explains the course and the symptoms of the case with which we are dealing. The process of gliosis, taking its origin in the gray substance behind the central canal in the lower cervical region and extending into the posterior cornua, produced the subjective and objective disturbances of sensation in the upper extremities; ascending in the left posterior horn into the central regions of the occipital nerve and of the sensory part of the trigeminus, it impaired sensation in the left side of the head and face, and, descending into the dorsal part of the cord, rendered sensation on the trunk abnormal; the invasion of the right and left anterior cornua, successively, in the lowest cervical and first dorsal segments, gave rise to atrophy of the small hand-muscles in the same order, and, from the wasting of the muscles of the left fore-arm, it is evident that the destructive process is extending

[1] University Medical Magazine, March, 1893.

into the anterior horn higher up. The distressing jerking in the left lower limb may possibly be due to irritation of the lateral pyramidal tract on the same side. It may be mentioned that a similar symptom was noted in one of Schultze's[1] cases.

The loss of the left conjunctival reflex is due to the sensory disturbance in the distribution of the fifth nerve, and the slight narrowing of the left palpebral fissure is ascribable to the implication of sympathetic fibers in the lesion at the lowest cervical and uppermost dorsal segments. There is no concentric limitation of the field of vision. According to Hoffmann,[2] limitation of the visual field has been observed by Déjerine and Tuilant in seven, and by Morvan in five cases of syringomyelia, without hysteria; by other authors it has been found in five patients with syringomyelia in whom hysteria was either certain or at least probable; and it was absent in 26 cases. Including the present instance, therefore, its absence has been noted in 27 out of 44 cases of syringomyelia. Its comparative frequency in the French cases is conspicuous.

The opaque nerve-fibers in the retina merit some attention. This anomaly is rare. Manz,[3] of Freiburg, states that he found this opacity mentioned only 154 times in the records of half-a-million patients with ocular diseases. But among 113 psychopathic male inmates of a Freiburg institution opaque nerve-fibers were observed in the eyes of four. Manz had

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[1] Virchow's Archiv, Bd. 102.

[2] Deutsche Zeitschrift für Nervenheilkunde, iii, p. 16.

[3] Berliner klinische Wochenschrift, 1890, p. 955.

already previously[1] called attention to the possible value of this, as well as other congenital anomalies of the eye, as indicating a neuropathic predisposition, although they are not necessarily pathologic; and he suggested that they were likely to be found associated with certain diseases dependent on congenital abnormalities of the central nervous organs. Now, according to the prevailing theory, the origin of syringomyelia is to be sought in a defect in the developmental tendency of the spinal cord, or, as Hoffmann more definitely expresses it, in a defective closure of the embryonic canal, whereby germinal cells are retained in the line of closure which subsequently proliferate and lead to primary gliosis. In support of the view that a defect in the original constitution of the central nervous system is responsible for the development of syringomyelia Hoffmann adduces some cases in which that disease was combined with other nervous diseases generally considered as arising from a congenital predisposition, and in others in which at the autopsy abnormal anatomic features were found that must be regarded as developmental errors. Among the latter are mentioned an abnormal strand of white fibers in front of the anterior commissure, heterotopia of gelatinous substance in a posterior column, heterotopia of gray matter, etc. The presence of medullated (opaque) nerve-fibers in the retina is submitted as an addition to this list.

In conclusion, the peculiar form noted in some of the spinous processes may be alluded to as perhaps indicating some developmental irregularity.

[1] Arch. f. Psychiatrie, 1884, xv, p. 837.