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Contribution to the Study of Anaesthetic Leprosy,

WITH

SPECIAL REFERENCE TO PARTIAL SENSORY DISORDERS.

BY

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REPRINTED FROM

THE JOURNAL OF NERVOUS AND MENTAL DISEASE,

June, 1889.

M. J. ROONEY & Co., PRINTERS,
Corner Broadway and 35th Street.
NEW YORK.

CONTRIBUTION TO THE STUDY OF ANÆSTHETIC LEPROSY, WITH SPECIAL REFERENCE TO PARTIAL SENSORY DISORDERS.[1]

By DR. GEO. W. JACOBY.

IT is very far from my purpose to even refer, before this society to the work and investigations which have been so energetically devoted during the last few years to the study of leprosy in its general aspect. The literature of this subject is so large, and the names of investigators in this field so renowned, that it would seem almost superfluous to endeavor to add anything to our present knowledge. But these remarks are true only from a dermatological standpoint; if we examine the question from its neuropathological aspect we still find questions which have been insufficiently ventilated, and upon which, notwithstanding the recent attention of Schultze and others, our knowledge could easily be enlarged. The points which are still *sub judice*, and which should be carefully noted in every case of so called anæsthetic leprosy are: the electrical excitability of the affected muscles; the condition of the reflexes; the absence or presence of fibrillary twitchings and, above all a thorough examination of the sensory symptoms should be made.

These data are important for the purpose of deciding two main questions; firstly, whether in a given case the leprous changes affect the central nervous systems, and secondly, whether we are actually dealing with a case of leprosy; for if in a case of supposed anæsthetic leprosy, in addition to the usual symptoms, progressive atrophy of muscles and trophic disturbances, such as ulceration and gangrene, we also find partial sensory disorders instead of the usual anæsthesia, we can easily understand how the the differential diag-

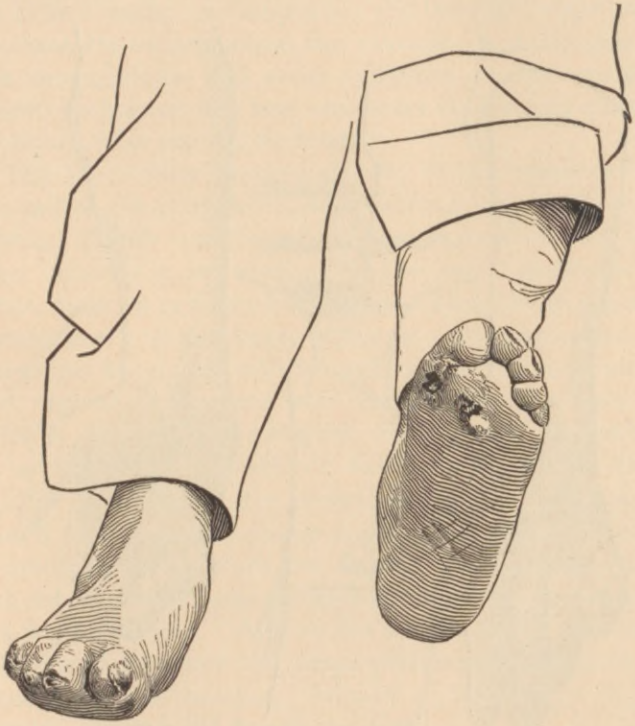
[1] Read before the American Neurological Association, Long Branch, N. J., June 27, 1889.

nosis between leprosy and syringomyelia may cause considerable difficulty. As a proof of the fact that these assumptions are not purely gratuitous ones, but that such a combination of symptoms as here mentioned does occur, I will describe the following case, the opportunity of examining and reporting which I owe to the kindness and courtesy of Dr. F. C. Valentine of New York.

C. S., male, æt. 18. Birthplace, Cuba. When eight months of age he was removed to Cartagena, U. S. of Colombia. His parents are alive and in good health. They have had eight children, five boys and three girls. The girls are all dead. One of the boys, during several years had some skin disease, but is now well. No history of heredity in collateral branches. Grandparents were over seventy when they died.

The patient himself was always delicate. He was not nursed by his mother, but by a Cuban woman, who subsequently had an illegitimate child, which at the age of six years developed a similar disease to that with which our patient is now afflicted. From his third to seventh year the patient was perfectly healthy. At this time an eruption appeared upon the buttocks, consisting simply in a reddening of the skin, which being observed by the mother, she asked the boy whether he had been struck. This was not preceded by pains. Similar spots hereupon appeared on the thighs, arms and forearms, lasted about a year and then disappeared. All of these spots looked as though he had been struck a slight blow sufficient to redden the skin. They did not have any definite form and and varied in size, some being very small, others half an inch in diameter. Just prior to the appearance of the first spot, patient found a place upon one of his legs, near the tibia, into which he was able to insert pins, without producing pains. He did this purely "for fun," because some of his playmates could also do it. When eight years of age the spots disappeared, and remained absent for a year. They then again became apparent, and extended to the face, involving the chin, face and ears; they again disappeared without leaving any mark. During this time he was under treatment. These

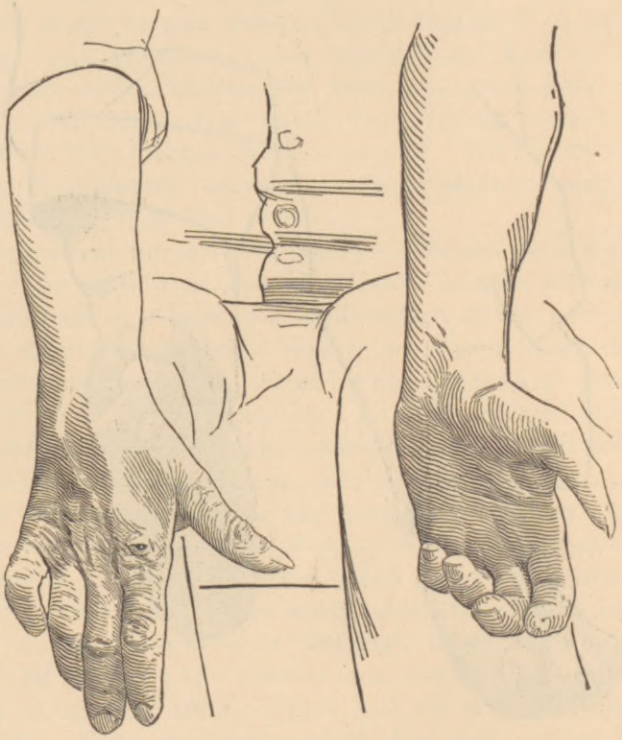
spots, when the patient was ten years old again reappeared, began to swell and became elevated and glossy. Coincidently his ears assumed a dark brownish red appearance, and became swollen and deformed. The hairs on the spots upon the body, fell out. The spots showed no distinct line of demarcation from the surrounding territories. When eleven years old, he suffered from restlessness at night, not



being able to find a comfortable position for his limbs ; also suffered from frequent cramps in the calf muscles. At the age of twelve, "a corn" as he calls it, appeared at the ball of the great toe on the right foot, ulcerated and made a deep sore. The same then occurred on the left foot. From this time until his fifteenth year, he was occasionally better, occasionally not so well. At fifteen, his hands became swollen, also the skin of the thorax, the lobes of the ears

and the nasal mucous membrane. The hands were not discolored, but the chest, chin and ears were brownish red. The joints of the fingers were enlarged but painless.

On the fourth finger of the right hand at the second joint, a swelling appeared which suppurated and left an ulcer. During all this time the ulcers on the feet persisted. At the age of sixteen he was attacked by some form of fever



and remained in bed for a month. During this time, all symptoms of disease again disappeared, only to reappear soon after getting out of bed. It was at this time that he noticed that he could not grasp small objects, could not button his clothes, and frequently burned his hands without being aware of it. Now, also the small muscles of the hands began to waste ; wasting proceeded slowly. Fingers continued to swell and finally began to contract in a pos-

ition of flexion. Previous to this, he had a large number of "corns" upon his fingers. At various times pieces of bone came from the ulcers on his feet.

Status præsens, Aug. 8, 1888.—Complains chiefly about the ulcers on his feet, and of the appearance of his hands. He says that his hands do not incommode him much, although he has had to give up his trade of watchmaking. Medium height, weight 125 lbs. Head and face normal; eye-brows sparse; no alopecia. On the right upper part of the chest 1½ inches below the clavicle, is a circular scar with central della and striæ radiating to the periphery. Several such scars on chest and scars of different character (probably from setons) on arms.

The hands show marked atrophy of the interossei muscles and of the eminences, most marked on dorsal surface between thumb and finger. There is contracture of the flexor tendons, most marked in the small finger of both hands, main en griffe on both sides. There is no scar at the contractures. Joints freely movable. The thickened ulnar nerves can be plainly felt through the skin in the epitrochlear regions. Pressure over the nerves produces pain, which is also felt in the fingers. The skin of the palms is dry and shining. Growth of nails retarded. Wasting of muscles of forearms, also of deltoids. The toes are malformed. The feet are plump and stumpy and show but little motion. The arch of the foot is lost. On the ball of each foot is seen a deep suppurating ulcer of about the size of a five cent piece. Also in the middle of the plantar surface of each foot, over the carpo-phalangeal joint is an ulcer in the process of healing. Motion of the thighs is good. (See photographs of hands and feet).

Electrical examination of the affected muscles shows a reduced excitability to both currents. Distinct reaction of degeneration could nowhere be obtained, but in the left hand, the muscles supplied by the ulnar nerve show ACC=KCC. The reaction of the apparently normal muscles is normal.

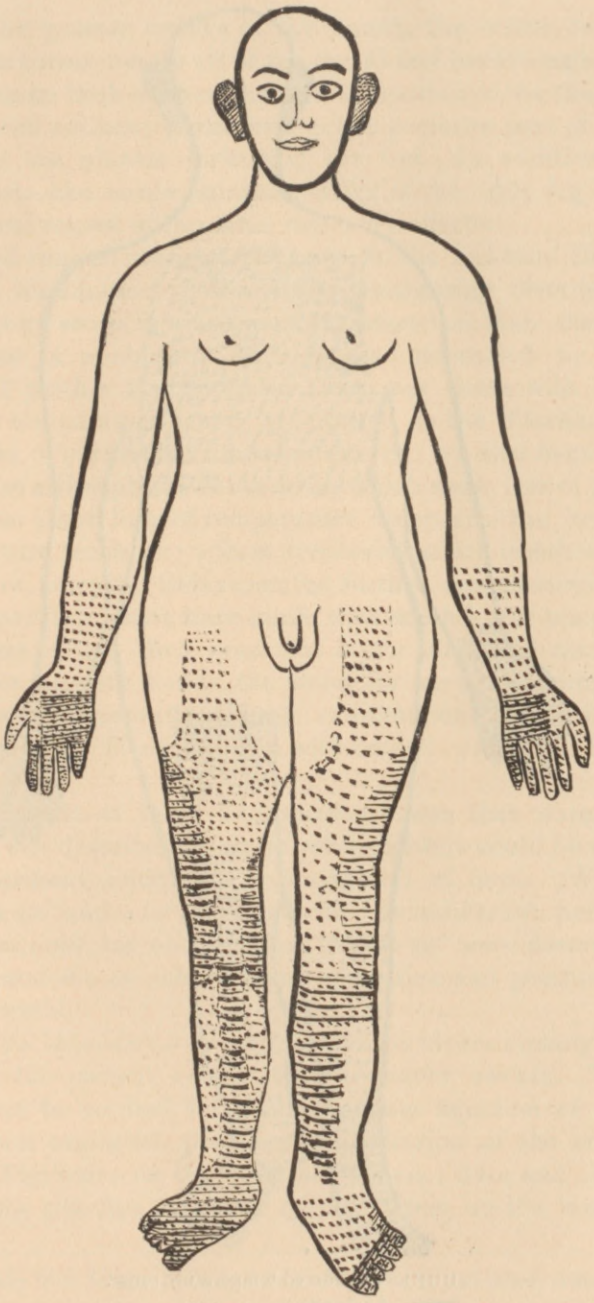
Mechanical excitability of muscles normal. *Reflexes:* the patellar tendon reflexes are much increased on both sides. Cremaster and abdominal reflexes present.

Sensation—Tactile sensibility.—The examination was made with a pin, with cotton and by pressure with the finger. Both arms, shoulders, and back showed reduced sensation (hypæsthesie v. Rentz) in irregular patches. In these places the perception for all strong applications was perfectly clear. Weak applications were also noticed and correctly localized, but the differentiation by the patient of the various modes of proceedure was not plain ; thus, if only lightly touched, the head of the pin could not be distinguished from the point, and cotton not from the finger. In the entire remainder of the body, the sense of touch is perfectly normal.

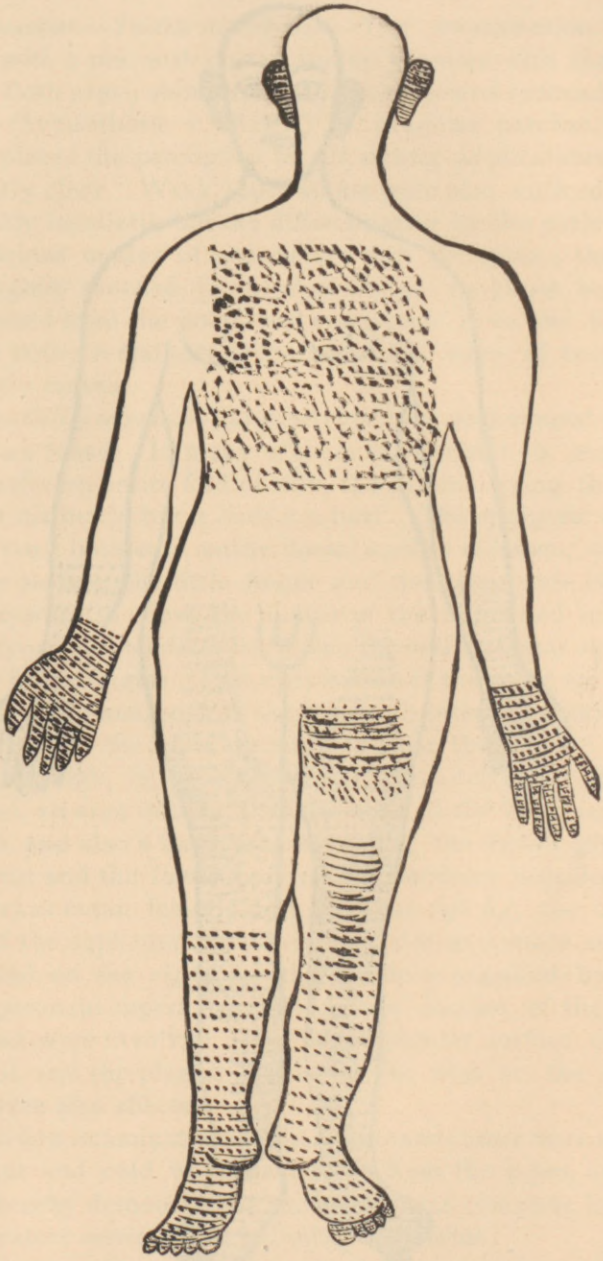
Sensibility to pain.—Large territories of pronounced analgesia are found. In many of these territories it is possible to transfix an entire fold of skin, the patient saying that he feels it distinctly but it does not hurt. The analgesic territories are : both ears, entire dorsal surface of hands; on the palmer surface, the little finger and the ulnar side of the palm upward to above the middle of the hand and inward just beyond the medium line; also the entire palmar surface of the index finger. This distribution is the same for both hands. Also analgesic is the left thigh internally and externally, from the knee upwards to near the middle. On the right thigh, in addition to the same territories being affected, an area internally at the bend of the knee was involved, and also a large area involving the entire gluteus maximus and the lower part of the territory supplied by the nervus cutan. femoris post. On the *left leg*, the upper half of the saphenous territory and dorsal surface of the toes, and on the *right leg* the territory supplied by the nerv. peroneus superf., and the entire dorsum of the foot and toes were involved. The entire plantar surface of the left foot, and the plantar surface of the toes on the right foot, were also affected.

The first examinations of the *temperature sense* were made with hot and cold water as it came from the pipes, and it was thereby demonstrated that there was complete loss of temperature sense in the following territories :

The upper part of both ears, the dorsal surface of both



:::::::: Disordered temperature sense
═══ Analgesia.



::::::: Disordered temperature sense.
═══ Analgesia.

hands, the palmar surface of the hands, the wrists, lower half of forearms, nearly all of the crural and lumbo-inguinal territories on both sides of the anterior surface of the thighs, both dorsal surfaces of the feet, entire posterior part of the legs and the plantar surface of the feet. In addition to these parts, the entire anterior surface of the right leg and the gluteal region of the same side were affected.

It will thus be seen, by reference to the annexed chart, that the distribution of the territories showing disordered temperature sense is much more symmetrical than that of the areas of analgesia. We are also impressed by the fact that neither the analgesic areas, nor those with disordered temperature sense, are limited to the distribution territories of particular cutaneous nerves; we also must remark that although the same areas which show loss of pain sense also show loss of temperature sense, still the loss of temperature sense also affects territories which in other respects are normal. Subsequently, further examinations of the temperature sense were made with much lower temperature, and it was then seen that in the ulnar and median territories of both hands, the sense for very low temperatures was preserved. These examinations were made with test tubes filled with ice, with cooled water and with hot water.

The result of these examinations was that water at 110° F. was described as warm, while blisters could be produced without eliciting any complaint of heat. Water between 50° and 110°, was described as indifferent, neither warm nor cold, but temperatures below 50° were described as cool and ice as cold.[2] Sense of posture and position of limbs normal.

Trophic disorders.—In addition to the ulcers already described, the patient asserts that he never sweats. This could not be verified, for a subcutaneous injection of 0,02 pilocarpini muriatici, produced perspiration of the entire body. The hairs on the body are sparse; here and there very long silk-like hairs are found. Thus, on the middle

[2] Normally from 86 upward is considered warm and 75 and below, cold.

anterior surface of the right arm is one of about 8 inches in length.

No fibrillary twitchings, no spontaneous pains. Eyes, sense of taste, speech, smell, and hearing, normal.

Internal organs normal.

If now we briefly summarize the case before us we must lay stress upon the following features :

1. Marked atrophy of both hands, principally of the small muscles, but also affecting the forearms and deltoids.

2. Increased tendon reflexes.

3. Sensory paresis, consisting chiefly of analgesia and disordered temperature sense, with preservation of tactile sense.

4. Trophic disorders.

If in addition to these points we consider the previous history of the case, together with the etiological data, we can, I think, arrive at no other conclusion than that we are dealing with a case of leprosy, of the so-called nerve or anæsthetic form.

The symptoms presented by this case do not esentially vary from those described by other writers ; certain changes to which little attention has heretofore been paid, have been carefully examined, and these changes, judging from the few cases in which they are referred to at all, do not appear to be of constant occurrence in this disease. Thus, Breuer, Rosenthal,[3] Vallin and Rosenbach[4] also found the tendon reflexes increased, while Schultze[5] did not find any abnormal deviation. Leloir[6] mentions the presence of fibrillary twitchings in one of his cases, while this symptom was absent in mine, and as far as I know in all other reported cases. Reaction of degeneration was not found by Rosenthal, Rosenbach, or myself, but was present in cases of

[3] M. Rosenthal. Zur Klinischen Characteristik der Lepra Anaesthetica. Viertelyahrschrift fuer Dermatologie, p. 425, 1881.

[4] Rosenbach. Ueber die Neuropathischen Symptome der Lepra. Neurol Centralblatt., p. 361, 1884.

[5] Schultze, F. Zur Kenntniss der Lepra. Deutsches Arch. f. Klin. Medizin., vol. 43, 1888, p. 496.

[6] Leloir. Traite de la Lepre.

Dehn [7] and Schultze. Loss of temperature sense was observed in the cases of Rosenthal and Rosenbach, but could not be found by F. Müller [8] nor Schultze. The symptoms here mentioned are of particular value in deciding upon the probable anatomical location of the disease, and the fact that these symptoms vary in the different cases would of itself lead us to suppose that the location of the pathological process is not always the same, but that this also varies according to circumstances. In one case, therefore, it is certainly admissible, if we consider the symmetrical distribution of the atropy and of the trophic disorders, the increased tendon reflexes, and the partial loss of sensation, to doubt that the assumption of a peripheral neuritis constituting the entire anatomical change is warranted, notwithstanding the fact that Leloir,[9] Schultze, and the majority of writers take this view of the matter.

Schultze explains the increased tendon reflexes, when they occur, by assuming an abnormal irritability of the muscles, and also believes that the symmetrical distribution of the disease in the hands, feet, and face is due to the greater exposure of these parts to direct infection. That partial sensory disorders have been found in anæsthetic leprosy, Schultze does not deny, but states positively that he has never found any. All such assumptions and negative statements go for nothing in the face of positive facts, and these facts all tend toward corroborating the statement already made, that the assumption of a peripheral neuritis alone is insufficient to explain the symptoms encountered.

It will also at once become evident that this question of the central or peripheral localization of the pathological process in the anæsthetic form of leprosy, must assume practical interest, on account of the relationship which this class of cases may bear to cases of syringomyelia.

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[7] Dehn. Deutsche Medizin. Wochenschrift., 1887, No. 45, p 988. Discussion of a paper by Dr. Arning.

[8] Muller, F. Ein Fali von Lepra. Deutsches Archiv. fur Klinische Med., vol. 34, p. 205.

[9] Leloir. Gazette des Hopitaux, p. 575, 1888.

Were it not for the etiological factors and previous history of my case, we would be in a quandary as to the differential diagnosis between leprosy and syringomyelia, provided, of course, that the symptomatology of the latter affection as to-day accepted, may be considered sufficient for diagnostic purposes. Attention has also been called to the possibility of mistaking these two affections, by other writers, and in practice this difficulty is illustrated by the case of Steudener[10] and Langhans.[11] In both of these cases the diagnosis of leprosy was made *intra vitam*, and the autopsy disclosed cavities in the cord.

In Steudener's case the following was shown on Autopsy. Spinal cord. A fissure like cavity, filled with a mucous sticky fluid, extends with various interruptions from the medulla to the lumbar enlargement. A loss of substance is found in the pyramid and a larger one near the vagus nucleus. A horse-shoe shaped, fissure like defect is also found above the cervical enlargement. Peripheral nerves of right upper extremity show thickening of the neurillemma; foci of small granulation cells and spindle-shaped swelling of the right radial nerve.

In Langhans' case, the autopsy showed an immense focus of softening, implicating the posterior horns, Clarke's columns and the grey commissure. Here is found a cavity which traverses the cord in a transverse direction. The peripheral nerves showed thickening of the peri and endoneurium.

These cases then, which from their clinical history were looked upon as cases of leprosy showed upon autopsy in addition to changes of the peripheral nerves such as we find in leprosy, changes in the spinal cord such as we find in syringomyelia.

Even assuming that these cases were not cases of leprosy at all, but were true cases of syringomyelia, as Schultze contends, there is still sufficient evidence from

[10] Steudener, F. Beitrage zur Pathologie der Lepra Mutilans, Erlangen, 1867; Obs. I., p. 7.

[11] Langhans, Th. Zur Casuistik der Ruckenmark's affectionen. Virchow's Archiv., 1875, vol. 64, p. 175.

other sources furnishing the proof of implication of the cord in leprosy. It is true that in the majority of anatomically examined cases the central nervous system was found normal and the peripheral nerves always affected, but on the other hand, the cord has been found to be the seat of congestive and hyperplastic changes, the membranes showing signs of inflammation and the cord itself being found thickened and sclerosed. Tschirjew[12] found the central canal of tne cervical region filled with round cells and also atrophy of the cells of the posterior horns and of the columns of Clarke.

Danielssén and Boeck,[13] in autopsies frequently found a toughening and atrophy of the cord with discoloration of the grey substance and diminution of the ganglia. On the posterior spinal surface meningitis was observed which often implicated the posterior nerve roots. They always found the cord to be more or less diseased. In consideration of these facts therefore, I can understand how in some cases the anatomical change may be in the cord, (perhaps a leprous new formation, with subsequent softening, and final formation of cavities) in others in the peripheral nerves, and in still others in both the nerves and cord. The sharp lines which have thus far been drawn around anæsthetic leprosy as an affection of the peripheral nerves, will, in view of the cases showing partial sensory disorders, have to be modified, or it must be acknowledged that partial sensory disorders as such are not characteristic of syringomyelia.

As well as I believe that the first position, that of considering all the symptoms in anæsthetic leprosy due to a peripheral neuritis, is untenable, so also do I believe that statements regarding the occurrence of partial sensory disturbances as a pathognomonic symptom of central cord affection, have been entirely too categorical, as shown by the following citations from Schultze and Starr.

Schultze[14] writing of the symptoms of syringomyelia,

[12] *Tschirjew* Archiv. de Physiologie, 1879, p. 615-23.

[13] *Danielssen* and *Boeck* Traité de la Spadalsked ou Elephantiasis des grees. Paris, 1850.

[14] *Schultze.* Virchows Archiv. Vol. 102 p. 450.

says: "In multiple peripheral degeneration of the nerves, the peculiar partial sensory paralyses have not yet been observed, particularly the disproportion between sense of touch in contradistinction to sense of pain and temperature." And Starr,[15] writing in 1888, speaking of the differential diagnosis between syringomyelia and neuritis, says: In neuritis all sensations are equally affected, and there is no case on record where pain and temperature sense have been lost with preservation of touch and muscular sense."

About six months ago I instituted a series of examinations of the sensory condition of all cases of peripheral neuritis coming under observation. Of many cases examined only one case showed a partial sensory disorder, and while the form was different from that seen in syringomyelia, it nevertheless will prove of value in supporting my proposition. In this case there was preservation of perception to very low temperature, while all other senses were lost.

The patient was a male, age 46; a brewer, who had his hand and wrist crushed between two kegs of beer.

Open wound, suppuration. Severe degenerative paralysis occurred, implicating the median and ulnar territories. Sensation; anæsthesia and analgesia, particularly marked in the distribution of the ulnar. Sense for temperatures from 40° F. upward lost, but temperatures below that were always felt as cold. A mixture of ice and salt in a test tube, when applied to the skin always produced the same exclamation, "very cold," while a blister could be produced by means of hot water, without causing a feeling of heat or pain. A case very similar to this, has been recently observed by Ziehl[16] and from his article I learn that Nothnagel[17] has reported a case in which after traumatism to the ulnar, reduction of all sensory qualities except that of temperature (heat and cold) occurred. Berger[18] reports a case

[15] *Starr, M. A.* Amer. Journal of Med. Sciences, 1888, p. 4 7.
[16] Ziehl Deutsche Med. W'schrift, p. 835, 1889.
[17] Nothnagel. Deutsches Archiv. f. Klin. Med. Vol. II, p. 296.
[18] Berger. Wiener Med. W'schrift, 1872, p. 786.

of peroneal paralysis due to cold, in which the sense to light touch was preserved and localized, while that to pain and temperature was very much reduced. Pick[19] also has lately reported a case of peripheral loss of temperature sense with preservation of all other qualities. These cases, while they do not present precisely the same sensory disorders as those observed in syringomyelia, at any rate plainly controvert the statements that partial sensory disorders do not occur in peripheral affections.

The strange fact that in the peripheral cases of Ziehl and myself, as well as in certain territories in the case of leprosy, sense of cold was preserved while that of heat was lost, can be explained and understood by the interesting and sufficiently well-known investigations of Blix[20] and Goldscheider.[21] In these three cases this occurred in the ulnar distribution, and in all of them there existed a neuritis of the ulnar nerve; it is therefore fair to assume that this symptom is in these cases of purely peripheral origin. The question then arises is there anything in this symptom, preservation of cold sense with loss of heat sense, which is characteristic of peripheral disorder? Is perhaps the diagnostic point between central and peripheral disorders of temperature sense to be sought in this fact?

These questions cannot be answered, for cases of syringomyelia have not been specially examined with very low temperatures, and therefore the sense of cold may still have been present, where now it is supposed to be absent.

My conclusions from these facts and reflections are that:

1. The differential diagnosis between anæsthetic leprosy and syringomyelia cannot always be made.

2. Partial sensory disorders are not characteristic of syringomyelia, but may occur in anæsthetic leprosy as well as in purely peripheral affections.

3. A differential diagnostic point between central and peripheral loss of temperature sense, may lie in its complete loss in the one case and its partial loss in the other.

NOTE.—Since the above was written, an acticle by Dr. S. M. Suzuki, in the Sei-I-Kwai Medical Journal, Tokyo, May, 1889, has come to my notice: Suzuki, in 17 cases of leprosy found the tendon reflexes exaggerated in all of them, and he also believes that this is due to a central cause.

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[19] Pick Wiener Med. W'schrift, 1888, p. 617.
[20] Blix Zeitschrift für Biologie xxi, p. 143.
[21] Goldscheider Dubois-Reymond's Archir, 1885.