Pachymeningitis spinalis externa : recovery complete / by F. W. Langdon... and by Albert H. Freiberg.

Langdon & Freiberg.
Pachymeningitis spinalis externa.

With the Authors' Compliments

PACHYMENINGITIS SPINALIS EXTERNA.

RECOVERY COMPLETE.

BY F. W. LANGDON, M.D.

Professor of Nervous and Mental Diseases, Laura Memorial Woman's Medical College; Clinical Professor of Nervous Diseases, Miami Medical College; Neurologist to the Cincinnati Hospital.

AND

BY ALBERT H. FREIBERG, M.D.

Professor of Surgery, Laura Memorial Woman's Medical College; Orthopedic Surgeon to the Cincinnati Hospital.

CINCINNATI, OHIO.

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RECOVERY COMPLETE.

J. X., aged 19, an American of Hebrew parentage, single, a salesman and clerk, was referred to the Cincinnati Hospital, neurological service, by Dr. A. V. Phelps, February 28, 1898.[1]

His chief complaint was "loss of power in arms and legs" of six weeks' duration. A few days before his admission he developed fever and a feeling of general illness which led him to apply for admission to the hospital.

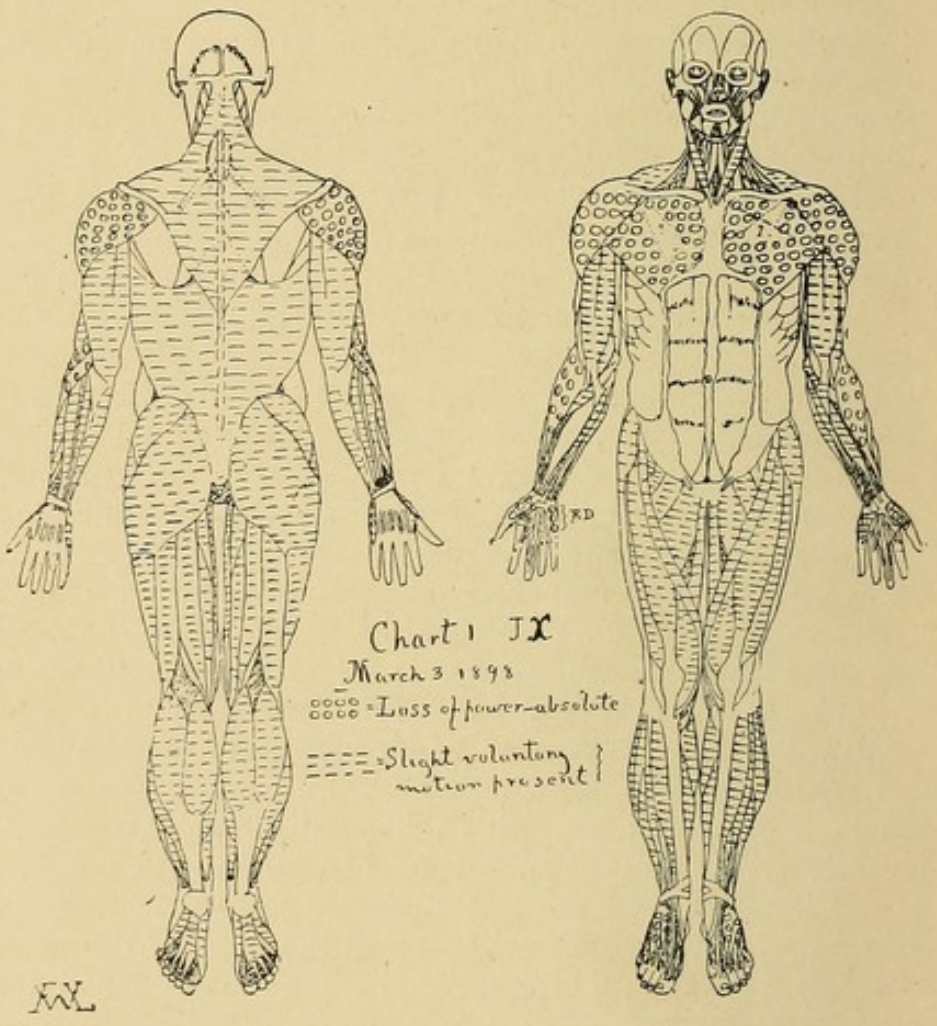
Family History.—His father died of "throat consumption," at the age of 49, having been ill for one year following a wetting and severe cold. His mother is subject to acute rheumatic attacks. One brother died in infancy; one sister has inflammatory rheumatism; three brothers and one sister are living and well.

Personal History.—He has been generally healthy and remembers no illness up to 14 years of age, when he had "sore throat," lasting two days. Syphilis can positively be excluded. He had gonorrhea eight months ago followed in two weeks by pain and stiffness in the back of the neck, which he called "rheumatism." He recovered and returned to his business within month, and was well for the succeeding five months with the exception of occasional pains in the back of the neck which did not disable him. There were no paralytic symptoms during this time.

The onset of the paralysis was gradual, beginning about six weeks before his admission to the hospital and being accompanied by some pain and rigidity in back of the neck. Motor weakness began in the left arm, and

[1] Acknowledgements are due to Drs. Victor Ray and John S. Boggess, internes, for their careful examination and history of the case, of which the following is an abstract, and also to our colleague, Dr. Herman H. Hoppe, for kindly permitting the use of notes made during his term of service.

gradually increased until in one week the entire limb was helpless. During this period the right arm became weak and he gradually lost the use of it. The legs were affected last; presumably simultaneously. Within a month—probably within three weeks—he was completely disabled in all four extremities. At no time previous to admission has he had any pain in his limbs,



or any bladder or rectal defect. He has had no cough; has had some headache and fever for two or three days before admission. Temperature shortly after admission was 101.6—rising to 104.2 within twenty-four hours—pulse 116, respiration 30.[2]

[2] This pyrexia and its accompaniments are apparently due to an intermittent tonsillitis, so far as can be seen unconnected with his spinal lesion and subsiding within a few days. See temperature chart appended.

Present State.—February 29, 1899. Height is 5 feet, 8 inches, weight 138, of medium build, dark complexion; black curly hair, dark eyes.

His general nutrition is good. Part of the thorax and abdomen is covered with a growth of pityriasis versicolor, which patient states has been present for six years. His neck is rigid, its tissues apparently infiltrated and indurated posteriorly. There is not much tenderness on pressure and manipulation. The left tonsil is swollen, its follicles being distended and filled with grayish-white secretion. The uvula and soft palate are bifid; no edema of larynx or pharynx. Patient can swallow and talk without much discomfort.

His mental condition is good; speech not impaired; cranial nerves not affected.

Trunk and Extremities.—Patient can not stand nor walk; quadruplegia is present, practically complete below elbow and knees; he can flex all fingers feebly. The paralysis is moderately rigid, almost "waxy" in type. Foot and wrist drop are marked on both sides. He can flex and extend both elbows feebly. *Extension at elbows is notably stronger than flexion.* Grasping power to dynamometric test R, 0. L, 0.

Sensation.—Tactile sensibility is somewhat diminished in acuity at the ends of the fingers, elsewhere apparently normal. Pain and temperature senses are not accurately tested at this date. Note change in cutaneous sensibility four days later, as shown by charts appended.

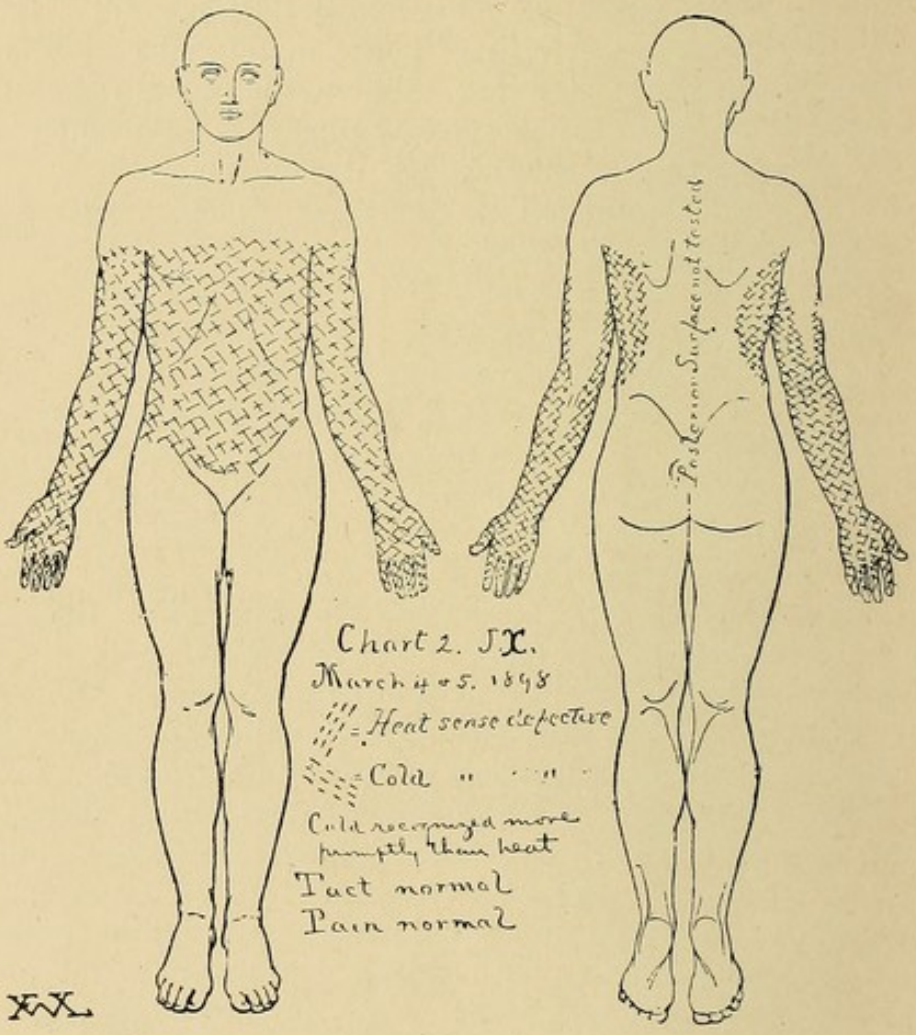
Reflexes.—Organic. No defects of deglutition, defecation or micturition. Tendon: Elbow-jerks present and equal; wrist-jerks present, active and equal. Knee-jerks present, exaggerated and equal; rectus and ankle-clonus present and equal on both sides. Cutaneous: Not observed. Vasomotor system: Patient sweating freely. Trophic: No muscular atrophy observable to ordinary examination. No trophic ulcerations.

Urine.—Reaction acid, barely; sp. g. 1030; phosphates in excess; albumin and sugar absent.

Blood-count shows a moderate leucocytosis (16,500.) This was probably due to the intercurrent tonsillitis.

July 10.—A tuberculin test, with m. xv of a 1-250 solution was followed in two hours by headache, and chill

and sweating in twenty-six hours. Two days after admission the acute tonsillar inflammation subsided, the temperature dropped to normal, and for the next four weeks fluctuated between 98 (a. m.) and 100 (p. m.), only once during this time rising 2 degrees above the 100 mark. After the two weeks following this period, the temperature varied between 98.4 (morning) and 99.4 (evening). (See chart appended.)



Re-examination four and five days after admission. (See chart No. 1.)

Motion.—Quadruplegia, of waxy, rigid type is still present. The pectorals, deltoids, supinators, small thenar muscles, and short extensors of toes seem absolutely powerless on both sides. Elsewhere the muscular power is barely sufficient to flex and extend joints, ex-

tension seeming rather stronger than flexion at the elbows.

The tongue protrudes in the median line, is longitudinally fissured, and a general fibrillary tremor of the entire organ is present.

Sensory defects of a "dissociation type" have appeared as per Chart No. 2. These consist practically of diminution and loss of appreciation of heat and—to a less degree—of cold, with preservation of tact and pain over thorax, abdomen and upper arm anteriorly, and on forearm and hands anteriorly and posteriorly. (See Chart 2.) Tests were made in the ordinary manner with test-tubes of decided warm and cold water for temperature, cotton and pin for tact and pain. A week later (see Chart 3) these sensory defects were increased by addition of an area of *analgesia* over the thorax anteriorly. At this date power in legs has apparently increased, so that he can move both feet and legs with considerable freedom as he lies in bed.

Reflexes.—Pupils are moderately dilated when at rest, respond well to accommodation and contract to light, but do not dilate farther when light is excluded. Organic: Has to be catheterized for a day or two. Vasomotor: Well marked "tache" over thorax and abdomen. Trophic: No "bedsores" or other ulceration.

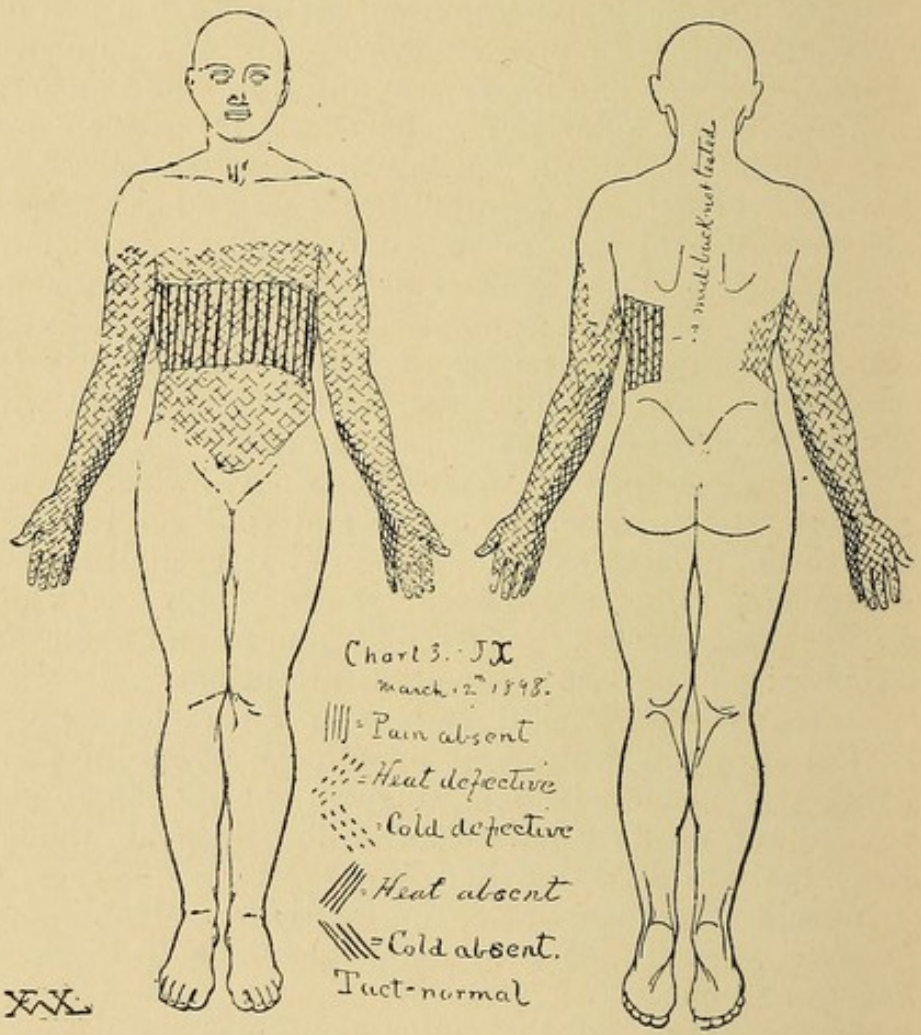
March 17.—Electrical tests now and later showed partial R. D. in muscles of hypothenar group of right hand, as evidenced by very sluggish contraction to galvanism and nearly equal responses to both poles, though K. C. is slightly greater than A. C.

Muscles elsewhere react normally to galvanic and faradic currents. There is slight improvement in power of legs and arms.

Twelve days later the motor symptoms were practically unchanged, the defects of heat and cold sense were somewhat diminished in area, but persisted on thorax and a longitudinal strip along the inner surface of the right arm, forearm and hand; also over left forearm and hand posteriorly. (See Chart 4.)

March 29.—About this time I asked my colleague, Dr. Freiberg, orthopedic surgeon to the hospital, to see the patient, and it was decided by his advice to make extension on the entire vertebral column in the hope

of relieving the pressure which was presumed to exist on the upper cervical cord. The effect of the extension and counterextension on the sensory symptoms was startling. Twenty-four hours after application of the apparatus, it was difficult to detect any sensory loss over thorax or abdomen, and when found it was in such



irregularly scattered patches as to make its accurate charting impracticable. By April 10—twelve days after application of extension—the only remaining defects were a loss of temperature—heat and cold—sense over right hand, palmar and dorsal surfaces; and over the left hand, dorsal surface only. The case was now transferred to the care of Dr. Freiberg, who furnishes the following:

SURGICAL HISTORY.

On March 28 I examined the patient whose condition has been accurately described by Dr. Langdon. I found present, exclusive of the nerve symptoms already described, a considerable, firm, diffuse swelling immediately below the occiput and extending downward to the fourth cervical spine. Without any distinct boss it was sufficient to render indistinct to the touch the vertebral spine. There was considerable tenderness on pressure complained of in the whole swollen area.

There was no torticollis whatever, but the patient was unable to rotate the head with freedom. Likewise it was impossible for him to bend the cervical spine backward to the normal extent. There was some interference with the power of approximating the head toward either shoulder. The nodding motion was not interfered with. Examination of the pharynx failed to show anything abnormal.

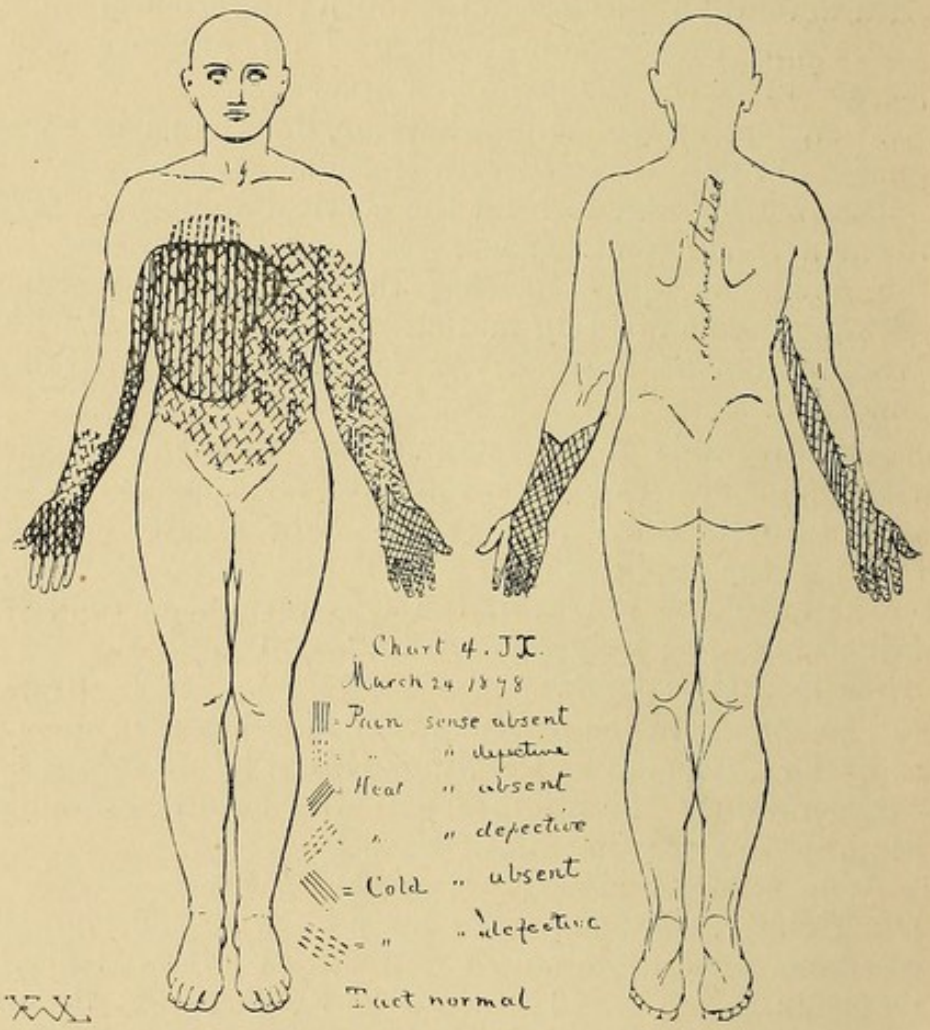
The diagnosis of tubercular disease of the upper cervical spine was made with some reserve, especially with regard to its exact localization. The treatment consisted in the application of weight and pulley extension to the head, the weight of the body serving as counter-extension. The weights were increased gradually from three to twelve pounds.

The change in the patient's condition has been spoken of by Dr. Langdon as startling; this is by no means an exaggeration. The improvement was, however, steadily progressive from this time forward.

June 4.—The appetite is good; no fever; no pain; less rigidity. He can move his neck with considerable freedom; also all joints of extremities. Grasp: dynamometer R. 50, L. 46; knee-jerks exaggerated; R. and L. ankle-clonus present R. and L.

The patient was kept in bed with the same weight attached until Aug. 4, 1898. At this time an examination showed an apparently complete return to the normal in every regard save one—the power to rotate the head laterally. The extension was therefore removed and a Sayre jury mast applied. On August 22 the patient expressed a desire to leave the hospital and was permitted to do so, wearing no apparatus, and he walked out of the

hospital without assistance. About six weeks ago I had an opportunity of examining the patient, and his condition remains the same. A very slight interference with rotation of the head and the induration about the upper spinous process, which has never disappeared entirely, are all that remained of the conditions found upon the first physical examination of the neck. The patient



asserted that his muscular power had returned in full degree.

ADDITIONAL COMMENT AND SUMMARY.

Since leaving the hospital the patient has been under our separate and joint observation at intervals of a few weeks. His health remains good and he attends regularly to his duties as salesman and clerk in a store.

Neurologic examination May 19, 1899, by Dr. Langdon: Height, 5 ft. 8 inches; weight, 138 pounds; general nutrition good.

Temperature, 99.2, after dinner and active exercise; pulse 72, regular. A slight fullness is apparent about the fourth or fifth cervical spines. Head movements are free as regards rotation and extension; somewhat limited as to flexion forward—can not touch manubrium sterni with chin. There is no muscular atrophy; no defect of gait or station. Power is good in all extremities. Grasp: dynamometer R. 105, L. 98—normal average about 80. The same dynamometer was used as in former tests.

Sensation.—No subjective sensory symptoms; no defect of tact, pain, temperature or muscle sense.

Reflexes.—Organic: No bladder or rectal defect, no dysphagia. Myotatic: Elbow and wrist-jerks present; right rather more active than left; knee-jerks somewhat hyperactive and equal. Rectus-clonus and ankle-clonus absent. Cutaneous: Palmar absent; epigastric, hypochondriac, abdominal and cremasteric present; normal. Plantar present, giving marked "flexor[3]" responses in both feet.

Diagnosis: Having in view the quadruplegic type of paralysis, the possibilities to be considered were: 1, cerebral diplegia; 2, poliomyelitis anterior; 3, multiple neuritis; 4, cervical myelitis; 5, spinal tumor; 6, hematomyelia; 7, hematorrhachis, external or internal; 8, leptomeningitis spinalis; 9, pachymeningitis spinalis interna; 10, pachymeningitis spinalis externa.—primary, *a,* rheumatic; *b,* gonorrheal; secondary *c,* tubercular (vertebral caries.)

Cerebral diplegia was readily ruled out by the history, the gradual onset, and absence of mental, aphasic, or cranial nerve symptoms. Poliomyelitis is excluded by the rigid type of paralysis and the presence of marked sensory defects. Multiple neuritis was not indicated by the history, and was further eliminated by the absence of pain, tenderness and muscular atrophy. Cervical transverse myelitis would have presented sphincter defects and trophic lesions, which were absent. Spinal

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[3] See Collier, "Brain," 1899.

tumor is conspicuous for the presence of root pains, which were absent. Gumma was excluded by lack of evidence of syphilis; and as already stated, syphilis is absolutely excluded in the case by recent developments of a most convincing nature.

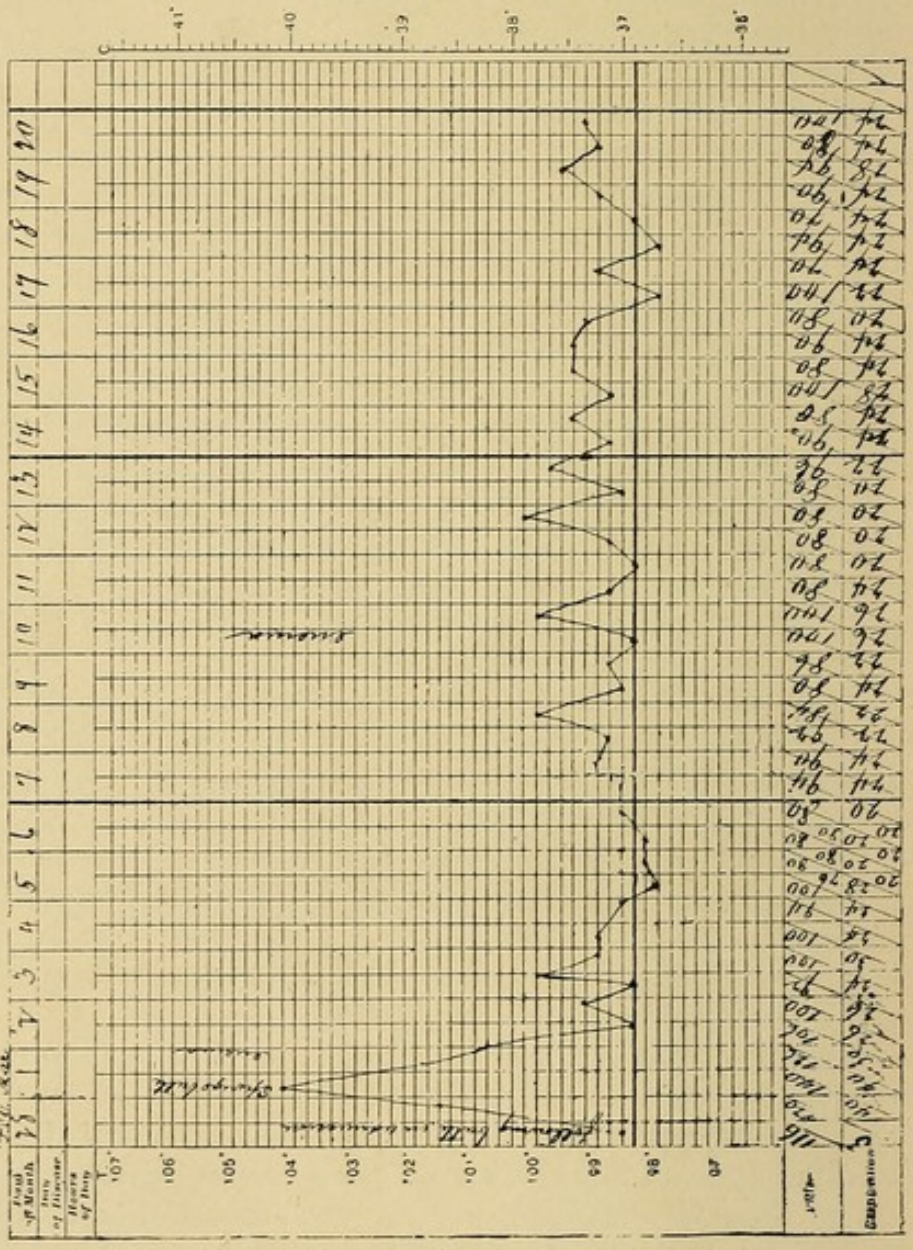

Chart 5, J.X. Pachymeningitis Spinalis Externa.

Hematomyelia and hematorrhachis have a sudden onset with rapid improvement if the patient lives. Leptomeningitis was contraindicated by slow onset, the ab-

sence of pain or hyperesthesia on movement of the spine, the evident localized character of the lesion, with little tendency to spread. Pachymeningitis interna was excluded by absence of adequate causes, as syphilis, alcoholism and trauma; also by absence of marked pain, and irritative root symptoms, as well as absence of the later muscular atrophy. Thus, by a process of exclusion a tentative diagnosis of pachymeningitis externa was reached, but whether this was primary, i. e., rheumatic or gonorrheal, as the history might suggest, or secondary to vertebral caries, as the heredity and part of the symptomatology would indicate, is perhaps an open question.

As Dr. Freiberg has stated, the diagnosis of tubercular disease of the vertebra was made with some reserve by both of us. Gowers[4] states on this point: "When clear indications of caries precede the paralysis the nature of the case can hardly be mistaken . . . When the two develop together mistakes are often made, but are usually due to the want of repeated examination of the spinal column. It is when the root or cord symptoms precede distinct evidence of bone disease and when the latter is so slight as to be equivocal that the chief real difficulty in diagnosis occurs." It will be seen that the case here recorded comes under the third condition described above.

To sum up: The symptoms indicated an increasing pressure of exudate with its incidence at the first and second cervical segments of the cord anteriorly, thus compressing the ascending anterolateral tract of Gowers —temperature sense symptoms—and the pyramidal tracts before their complete passage into the lateral columns. In no other situation is it possible to conceive an external lesion causing the motor symptoms here presented, viz.: quadruplegia with waxy rigidity and without muscular atrophy of the arms. The slight electric changes noticed in the hypothenar group of right hand were doubtless due to a nerve-root involvement lower down; as was the absence of pupilary dilatation on removal of light.

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[4] Manual of Diseases of Nervous System, vol. i, p. 251.

Against the presence of primary vertebral disease are the supposed "rheumatic" constitution, the preceding gonorrheal infection, the absence of trauma. In favor of primary vertebral disease of tubercular origin are the hereditary factor, the tenderness on pressure, with thick-

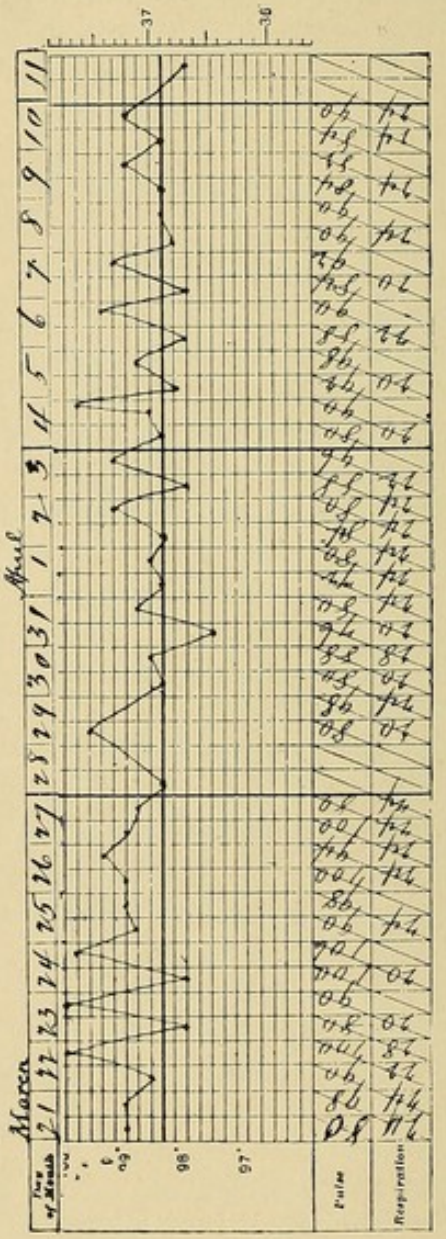

Chart 6, J.X. Pachymeningitis Spinalis Externa.

ening over cervical spine; the reaction to the tuberculin test, which, however, was much delayed; and finally the doctrine of probabilities, as well as the favorable outcome.

Treatment.—In addition to the surgical treatment proper, of extension and counterextension, potassium iodid was given in twenty-grain doses for about one month following his admission to the hospital. Cod-liver oil and hypophosphites were also administered, and the patient was kept in the open air whenever practicable.