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Harrington, Archibald W. Teacher, John H. 1869-1930. University of Glasgow. Library

Publication/Creation

[Glasgow]: [Printed by Alex. Macdougall], [1911]

Persistent URL

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CASE OF PERIPHERAL NEURITIS OF OBSCURE ORIGIN, WITH SECONDARY CHANGES IN THE SPINAL CORD.

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(Reprinted from the "Glasgow Medical Journal," August, 1911.)



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This case was remarkable for the slowly advancing but steadily progressive involvement of nerves, those of the limbs being first affected, and, finally, both recurrent laryngeal, and the right sixth, nerves. In addition, extensive changes, resembling those found in tabes dorsalis, were present in the

spinal cord.

The patient was a woman of careworn appearance, who had never enjoyed good health. The cause of her illness was rather obscure. She denied alcohol, but drank tea to excess. Until three years prior to admission to hospital she had been in very comfortable circumstances. During these last years she had not been so prosperous, and, in consequence, had worried a good deal. It is extremely probable, too, that her food had been insufficient.

A considerable amount of mental dulness and apathy was present, so that estimation of the sensory disturbance was difficult. In the earlier stages, however, there was some anæsthesia of the legs and arms. Marked deficiency of the muscular power of the limbs was present, but not specially of any muscle or group. The pulse was small and frequent on admission, and remained so throughout. Clinically, the most remarkable feature of the case was the involvement of the laryngeal and pharyngeal muscles. Except in cases following diphtheria, the occurrence of such palsies is rare in peripheral neuritis. The laryngeal palsy only became noticeable four and a half months after the onset of the illness, and at the

same time the right external rectus muscle became paralysed. For a few days prior to death the pharyngeal muscles were evidently paretic, saliva and food débris accumulating about the posterior pharyngeal wall. The soft palate was completely paralysed. The diaphragm and intercostal muscles escaped. Such symptoms, with the persistent acceleration of the pulse, indicated that the vagus nerves were affected. This involvement could not be attributed to a relapse, as the patient showed practically no signs of improvement at any time. She eventually died of pneumonia, probably due to insufflation.

Very marked changes were found in the nerves and spinal cord. The lesions in the cord were not unlike those found in tabes. Clinically, however, there is no doubt that this was not a case of tabes. The general history of the case is quite opposed to such a diagnosis. The arm-jerks disappeared while the patient was under observation, and when seen by one of us, prior to admission, the knee-jerks were present. There was no inco-ordination, but simply tremor. Argyll-Robertson pupil nor optic atrophy was present. Pathologically, while there was an acute degeneration of the posterior columns, most of the nerve lesions were evidently older than the change in the posterior columns. In our opinion, the latter were secondary to degeneration of the cells of the posterior root ganglia, which, in turn, was the result of the very complete destruction of the peripheral nerve trunks which had taken place.

The exciting cause of the neuritis in this case was, as has already been pointed out, obscure. It is interesting to note that Gowers states that the cases with most widely distributed symptoms are those that result from obscure toxæmic states, from cold and from alcoholism, and that it is in these, especially in the first, that the nerves of the heart, larynx, and of the muscles of respiration most frequently suffer.

Mrs. T., aged 58, was admitted to the Glasgow Royal Infirmary on 6th August, 1909, complaining of pains in the limbs of about seven weeks' duration.

Patient states that she has never enjoyed robust health. During the past seven weeks she has been much troubled with pains in the hands, fore-arms, feet, and legs. These pains are almost constantly present, and are becoming more severe. They are "sharp and jumpy" in character, and are worse at nights. They prevent her walking any distance or working with her hands. A very disagreeable sensation is

present on putting the hands into water. Sometimes there is a feeling of "pins and needles" in the fingers, while at other times they are numb. She has difficulty in walking, and sometimes feels very dizzy for a day at a time. The appetite has been poor, and the bowels are irregular, tending to be constipated.

Previous health.—Measles and whooping-cough in childhood and "rheumatism" as a girl; bronchitis for the past thirty years. Menstruation ceased thirteen or fourteen years ago. She used to have "bilious" headaches frequently, but these

have ceased since the onset of the pains in the limbs.

The family history is unimportant. Patient has had no

children or miscarriages.

She has always been in comfortable circumstances. During the last three or four years, however, she has not been so well off, and has been considerably worried. She drinks tea to

excess, but denies alcohol.

On admission, patient is a very thin, small woman, with flabby muscles and very little subcutaneous fat. She looks anxious, and complains a good deal in rather a querulous The temperature is normal. The left pupil is slightly larger than the right. Both pupils react normally, and the ocular movements are good. The grip of the hands is very poor, and the power of the arms and legs is impaired. The flexor-jerks in the arms are very active. The tricepsjerk cannot be elicited, nor can the knee-jerks, but it is doubtful whether the latter are entirely absent. The plantar reflexes are present, equal and flexor. The epigastric reflexes are present but not active. Umbilical reflexes are very There is probably no sensory disturbance. Some enlarged glands are present in the right groin. The mouth is practically edentulous. The tongue is clean, and the fauces and pharynx normal. The lumbar and lower dorsal regions of the spine are very stiff.

The pulse is small and regular, numbering 104 per minute. The systolic blood-pressure is 160 mm. Hg. The arteries feel distinctly thickened. The cardiac sounds are pure but somewhat distant. The second aortic sound is relatively loud. At the left base there is comparative dulness, with some diminution of the breath sound but no râles. Otherwise the lungs seem normal. Nothing abnormal can be detected on examination of the abdomen. The bowels are rather

constipated.

16th August, 1909.—Condition is much the same, the pains being slightly less. To-day there is some anæsthesia of the

outer sides of the legs, more marked on the right. Tactile sensibility is distinctly deficient on the right leg, and painful sensation is diminished. Perception of heat and cold is dulled, and a painfully hot test-tube is only felt as "warm." The patient's answers, however, are rather confused, and there is

probably a good deal of mental disturbance.

30th August, 1909.—Patient is rather worse. Tremor is considerable. Pains and tenderness of the nerve trunks are still marked in the arms, fore-arms, thighs, legs, and dorsa of Considerable atrophy of the muscles is present, marked in the fore-arms, interessei, and thenar eminences. Common sensation is somewhat deficient in the hands and fore-arms. Heat and cold are appreciated fairly well. anæsthesia is more marked in the legs, chiefly on the outer sides, and it extends a handbreadth above the knees. Tactile, painful, and thermal stimuli are either not appreciated or delayed. The patient can localise fairly well. Only slight jerks can be elicited in the arms, and the knee-jerks are The plantar reflexes are present and flexor. skin generally is rough and dry. From the elbows downwards it is of a distinctly brownish colour, which is said to have been of recent development. The pulse is varying between 108 and 128 per minute. The cardiac sounds are feeble and toneless. A few coarsely crepitant râles are present at both bases.

7th September, 1909.—Patient is sleeping better, complaining less, and taking her food fairly well. All movements of the extremities are possible, and the limbs can be handled freely without complaint. Tremor is considerable, and sensation is still defective. The pulse varies from 116 to 132 per minute,

1st October, 1909.—Patient says she does not feel so well,

but objectively nothing can be made out.

14th October, 1909.—No improvement has occurred, and there is probably some loss of weight and strength. The patient is absolutely listless, and takes no interest in anything. She says she has no pain, but the soles of the feet are tender when pressed upon. The mental dulness makes accurate examination impossible, but there seems to be no actual palsy, although all the muscles are tiny, flabby, and of slight power. Sensation is probably impaired in the arm below the elbow, and in the legs below the knee. The reflexes are all diminished or absent. She sleeps fairly well, but does not take her food well. Pulse numbers between 108 and 132 per minute.

16th October, 1909.—Paralysis of right external rectus is

present.

23rd October, 1909.—There is no perceptible change. Paralysis of the right external rectus persists. The right pupil is slightly smaller than the left. The reactions to

light are very trifling.

31st October, 1909.—Patient's progress is very unsatisfactory, and there has been no gain either physically or mentally. Recently, swallowing has been difficult and she has been having choking fits. The pulse is still frequent. Dorsiflexion of the left foot is associated with inversion. The degree of flexion is probably equal. The right grip is probably less than the left. Palsy of the right external rectus persists. The left palpebral fissure is larger than the right. The left eyebrow is more curved than the right, and the left frontal furrows more marked. The knee and arm-jerks cannot be

elicited. The soft palate is inactive.

1st November, 1909.—The choking attacks have continued. Some are definitely related to swallowing, but others have occurred apart from this. In an attack this morning, patient, who was lying quietly, suddenly sat up in bed, with croupy and stridulous inspiration and easy expiration. She became slightly cyanosed, and the pulse was frequent. After a few minutes the attack passed off, and the breathing became easy and free. On examination there is notable paralysis of the soft palate. There seems to be some paresis of the pharyngeal muscles. Saliva and food débris are accumulating about the posterior pharyngeal wall, chiefly on the right side. epiglottis moves. The right vocal cord moves very little, and lies almost in the middle line. The left cord moves deficiently and with a somewhat tremulous motion. The cords do not come completely together on phonation, and only a small aperture is present between them on deep inspiration.

4th November, 1909.—There has been slight fever since 29th October, and great restlessness with extreme general weakness. The facial and pupillary conditions are unaltered. There is slight comparative dulness at the right base with absence of breath-sound, and a few râles are audible at the left base. The "choking" attacks have been less troublesome for the last twenty-four hours. The diaphragm is acting.

7th November, 1909.—Patient is much worse. The temperature is higher and the pulse and respirations more frequent. She is noisy, sleepless, and restless. The tongue is very dry. She can swallow semi-solid food fairly easily. There is occasional incontinence of urine. A systolic murmur

is present at the apex, and signs of consolidation are present at the left base, with a few râles at the right base. Speech is very indistinct.

The patient gradually sank, and died on 8th November.

Abstract of pathological report (post-mortem No. 6,156).— The body was extremely emaciated, and muscular wasting was very marked. The internal organs showed general atrophy. There was a slight cirrhosis of the kidneys. The cause of death appeared to be broncho-pneumonia. There were numerous calcareous lymphatic glands in the root of the neck. The thyroid was considerably enlarged, soft, and red. It consisted to a large extent of young thyroid tissue showing signs of active proliferation. The condition appeared to be a simple hypertrophy. The brain showed no pathological condition, except slight atheroma of the arteries at the base. Changes in the spinal cord were not recognised at the post-mortem examination.

The following nerves were examined, and portions taken for microscopic examination, viz., the median, peroneal, recurrent laryngeal, and vagus nerves above the origin of the recurrent laryngeals on both sides. The nerves were much less easily dissected out of the surrounding connective tissue than normally, being evidently adherent to it.

The pons varolii, medulla oblongata, cord, and nerves were examined by the methods of Marchi, Weigert-Pal and van

Gieson.

The cord showed pronounced degeneration of the posterior columns. This was evident by all three methods. There was a large amount of blackening in sections stained by Marchi's method. The other two methods showed a definite sclerosis, but there was no shrinkage of the columns, which were full and rounded on the posterior surface (see Fig. 1). degeneration extended the whole length of the cord, involving the columns of Goll and Burdach about equally. It could not be traced above the nuclei of the medulla oblongata. The pia mater appeared to be slightly thicker and denser than normal. The posterior roots showed advanced degeneration and interstitial sclerosis, but less than the peripheral nerves. anterior nerve roots appeared normal by the Marchi and Weigert methods, but with van Gieson's stain one had the impression that the amount of interstitial tissue was in excess of the normal. The same might also be said of the interstitial tissue in the white matter of the cord generally, apart from the definite sclerosis of the posterior columns. The conditions

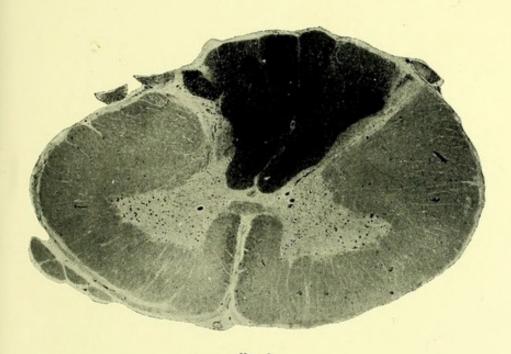


Fig. 1.

Section of the spinal cord showing degeneration of the posterior columns and nerve roots (Marchi's method).



Fig. 2.

Sections of the vagus nerve showing comparatively slight and of the median nerve showing very advanced degeneration (Marchi's method).

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were not favourable to examination of the cells of the grey matter, but they certainly showed an excess of pigment, and displacement of the nuclei to the side, and probably chromatolysis. Through a misunderstanding, the spinal ganglia were not taken.

The nerves.—The nerves of the leg and arm and the recurrent laryngeal showed very advanced degeneration, the intensity of which was best shown by van Gieson's method. sheaths of the nerves and the septa between the nerve bundles were greatly thickened and sclerosed, consisting of dense ibrous tissue, with very few cells. The nerve bundles, nstead of presenting the normal rounded form, were shrivelled and angular. This was distinctly less advanced in the recurrent laryngeal nerves. In sections by Marchi's method they showed much blackened material, partly in nerve fibres, but principally in the form of large masses not definitely related to fibres. Van Gieson's method showed very advanced interstitial sclerosis, with only a very few nerve fibres remaining embedded in dense connective tissue. The vagus nerves, on the other hand, showed very little degeneration by Marchi's method, but distinct interstitial fibrosis with van Gieson's stain (see Fig. 2). The root of the right sixth nerve gave a well-developed Marchi reaction and considerable interstitial sclerosis; the left, a much slighter degree of both changes. There was also distinct early degeneration (Marchi) in the roots of the fifth nerve, and in some nerve roots about the middle of the medulla oblongata. The only lesion made out in the pons varolii was some early degeneration in the neighbourhood of the root of the sixth nerve.

The condition may be summarised thus:—There was wide-spread interstitial neuritis, and this was of a very advanced degree in the nerves of the limbs, less in the laryngeals, and still less advanced in the sixth nerve and vagus. The degeneration in the posterior nerve roots and columns, while pronounced, appeared also to be very much less advanced than that in the peripheral nerves, and clearly of much more recent development. Pathologically, therefore, as well as

clinically, the case is one of peripheral neuritis.



