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A NEW FORM OF ENDEMIC PERIPHERAL NEURITIS

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INTRODUCTION:

Beyond the North West Frontier of India and separating it from the Pamirs, lies a broad belt of country commonly spoken of by the people of the Punjab as "Yaghistan", or Independent Territory. Until a comparatively few years ago this was practically a "terra incognita", and almost nothing was known of the people, their customs, or their diseases. In 1895, for the first time the veil was lifted, when a large expedition made its way through this region to the relief of Chitral. At the outset, there was a good deal of opposition, but, as matters quietened down, the natives of the country began to seek relief in large numbers from our field hospitals, for wounds and disease. A little later, the Government of India established a special hospital for their care, which was placed under my charge. At the end of the year, when the force withdrew to India, at the request of the people the civil hospital was allowed to remain, and for a period of eighteen months, I lived among them and had them for my sole care. In that interval over 11,000 patients attended for treatment, coming from a11/

all parts of the Swat Valley, and from the adjacent states of Buner, Dir, Bajaur, the land of the Utmankhel and Kohistan, with some even from Afghanistan. I thus came in contact with all the diseases of that region, and among many interesting cases seen, were 23 of special interest, which I am making the subject of discussion at this time.

These were instances of a peculiar form of peripheral neuritis which appears to be endemic in the trans-border region, and is, I believe, different to any of the varieties of poly-neuritis already de-Since leaving the frontier, I have been for four and a half years physician in the large Mayo Hospital of Lahore, to which the sick of all the Punjab come for treatment; and, although I have seen some hundreds of cases of multiple neuritis in that period, I have only met one instance of this affection, and that, curiously enough, came from the trans-frontier region on the west. variety of peripheral neuritis that this condition most nearly resembles is leprous neuritis, but, from a fairly large experience of that disease, I believe it to be quite different from the affection I describe. It is of interest too, that the people of the border, to whom leprosy is very familiar, do not/

not regard it as such, for to this affection they give the name "Spuna", (really Leucodermia), while tubercular leprosy is called "Juzam", and the anaesthetic variety "Pisak." They too, make no distinction with patients the subject of this disease, while sufferers from leprosy are driven out of the villages and compelled to live apart.

Before proceeding to a description of this affection, it will be well to give a rough idea of the geography of this part of the trans-border region so that references afterwards may be understood. "Yaghistan" is not one place, but a generic name given to a number of independent tribal states beyond the Indian frontier. The part with which I deal, lies due north of the Punjab, between the Indus river on the east and the Afghan border on the west; south of it is the Punjab and to the north, Chitral, Hunza, Nagar and Gilgit, - all inhabited by an entirely different race of people. Roughly speaking, "Yaghistan" is about 200 miles from north to south, and 150 miles from east to west; while it is crossed diagonally from north-east to southwest by two large rivers, the Swat and Panjkora, which conjoin in the south-west corner. Separating these rivers from one another, and from the Indus and/

and Kunar rivers respectively, are huge mountain ranges which vary in height from 3000 to 15,000 feet, - generally speaking those tending to be highest which are furthest north. In the low-lying grounds in the bed of each river, there is a swampy zone of rice-fields, which extends up either valley for about 100 miles and varies in breadth from one to three miles. On the higher ground, away from the river, the different cereals are cultivated, and the same cultivation goes on up to a height of about 9,000 feet. Beyond 9,000 few crops are grown, but flocks are grazed right up to the summer limit of the snows - some 13,000 or 14,000 feet. valleys are more or less densely populated throughout their length, but the density is greatest in the lower reaches.

This tract of country is inhabited by a warlike race of people called "pathans", who conquered the country nearly 400 years ago and drove out the original inhabitants. It is split up into large tribal states, named Buner, Swat, Dir, Bajaur and the country of the Utmankhel. Buner lies to the east of Swat, between it and the Indus. Swat comprises the greater length of the valley of the Swat river above its junction with the Panjkora, while from the confluence/

fluence down to the Punjab border is the land of the Utmankhel. Dir, for the most part, embraces the Panjkora valley; and, west of the Panjkora, between it and Afghanistan is Bajaur. Kohistan is the high mountainous region at the head of the Swat and Panjkora rivers, and its people are the original inhabitants of Swat, who were driven out by the Pathans when they conquered the country. state, in its turn, is subdivided into districts, which bear the names of the original leaders - the terminations being "Zai", land, or "Khel", tribe, as "Shamozai", the land of Shamo, "Utmankhel", the tribe of Utman, and so on. There is also throughout the whole region a hardy race of hill shepherds called "Gujars", who graze their flocks beyond the limits of cultivation right up to the snow line, and who lead a nomadic gipsy kind of life.

STUDY OF THE DISEASE:

The affection I am now about to describe may be defined as a specific form of peripheral neuritis which is endemic in the trans-border regions of northern India, and which runs an exceedingly chronic course lasting over many years. It has a very slow and insidious onset, and begins with purely/

purely sensory symptoms which may continue alone for months or years, and to which the patient attaches little importance; but, sooner or later, these are followed by wasting of muscles and motor changes, and ultimately, there develop white anaesthetic patches in the skin, resembling those of leprosy.

Twenty-four cases in all of this disease have been observed by me which all came from the transfrontier district, and I shall, after giving a general description of the main features of this form of peripheral neuritis, review some of the more typical cases in detail, and use them to illustrate the course of the disease, leaving its Aetiology and Pathology to be considered later.

CLINICAL FEATURES:

The early symptoms are purely sensory, and of the nature of tingling and pricking in the fingers and toes, with the feeling of insects creeping over, and boring holes into, the skin - graphically described by the Pathan as "megi megi kegi". These sensations are first felt in the winter time, and continue till the summer, when they pass away, but recur with greater severity the following cold weather, and are usually first noticed in the fingers and toes/

toes and along the inner border of the hand. For a long time, may be for months or more, nothing further is experienced, but with the ensuing cold weather they augment in intensity, and there is superadded burning in the palms of the hands, and soles of the feet, "sawe kegi", as the patient describes it. Pain is as a rule seldom felt, although now and again a case a more acute commencement with pain in the knee, ankle, wrist or other joints, or in the hands and feet. Sometimes in the winter, patients complain of feelings almost amounting to pain, but these pass away in the summer, and almost always the onward course of the disease is devoid of pain.

symptoms, there develops a feeling of itchiness in the hands and feet and the skin in these regions begins to become dry, crinkled and greyish white. Then the patient will complain that the parts feel numb and dead, and are no longer sensible of outside temperatures. They now feel cold to him and he has constantly to rub them to keep them warm, while it not infrequently happens that they become accidentally injured by snow or fire without the patient's cognisance, and ulcers and scars form on the fingers and toes.

Slowly/

Slowly and gradually the condition spreads from the fingers to the hands, and from the toes to the feet, and insidiously continues creeping up, getting worse and worse each winter and improving somewhat with each warm weather. As the tingling and formication feelings spread up, the burning and numb feelings follow, until, may be, the disease reaches in the arms to the elbows and in the legs to the knees, or even higher. In this way, the disease may continue for years, slowly and insidiously increasing and gaining in extent each winter after its summer of quiescence. Yet, all this time, the symptoms have been merely sensory, and the patient has gone about his work as usual.

When such a case is examined in this sensory stage, there is little to observe. Generally, the patient is a strong, healthy-looking man, and when stripped, merely shows a symmetrical greyish white scaliness of the hands and feet which are dry and cold to the touch. There are usually white linear marks and excoriations from scratching, for these whitened areas are very irritable and itch a great deal, and now and again a sore or scar may be seen on the hands or feet from accident, or the loss of the end of a finger or toe from frost bite. When the/

the cutaneous sensibilities are tested a point of great interest is brought out, for the first sense which seems to suffer is that of temperature, and especially the one for the perception of heat. If tested with hot and cold tubes, both cold and heat perceptions are usually dulled, but the latter to the greater extent. Generally the sensation of a hot tube cannot be perceived at all, or is called cold, while a cold tube may be quite readily recognised as such. As the condition advances cold perception also is lost, so that after a time there is in some or other area absolute anaesthesia to both sensations.

The pain sense is at first little affected, and, very often, while there is anaesthesia to temperature, the pain sense is normal, or hyperalgesic, as tested by pricking with the point of a pin. As the case progresses, the perception of pain is dulled, but only in very advanced cases is there absolute anaglesia. More usually, the pain and touch senses are only dulled, while that of temperature is quite wanting. The tactile sense is still less affected, and at first is quite good, even when the temperature sense is much impaired, and pain somewhat so, but as the disease ages, localization/

isation of touch becomes defective, and in the end, touch cannot be perceived at all. When anaesthesia and anaglesia do occur, the commonest sites are the inner side of the palmar surface of the hands, and the inner sides and dorsal aspects of the feet, while the loss is always symmetrical.

Besides these alterations in cutaneous sensibility, the only other objective signs at this stage are the loss of the palmar and plantar reflexes. The knee and elbow joints are at first unaffected, but as the disease creeps up, they become slightly exaggerated and ultimately lost. For a long time there is no ataxia, and the patient can stand quite steady with the feet together and the eyes shut, but in the advanced cases Romberg's sign develops. The muscular sense is at first quite good, but later on becomes impaired, but the muscles are never sensitive to pinching as in the acute forms of multiple neuritis. The stereognostic sense goes fairly early, and objects placed in the hands can only be recognised with difficulty, or not at all.

Motor symptoms do not supervene until after months or years, but, sooner or later, there is noticed weakness of muscular movement and the inability/

ability to perform fine movements with the hands, while the toes tend to catch and trip over ruts and stones, and the gait becomes unsteady. By this time there is visible evidence of muscular wasting, and the hands and feet begin to acquire a deformity. The first muscles to waste are those of the hypothenar eminences of the hands, the extensors of the little and ring fingers and the abductor indicis. Following upon those, the muscles of the thenar eminence, the lumbricales and the remaining interossei waste, but it is long before the atrophy extends to the muscles of the forearms. The feet usually suffer in the same way as the hands, but to a lesser extent, and corresponding muscles to those of the hands become implicated, and, in time, when the loss of muscular power extends to the legs, the tibialis anticus, extensor brevis digitorum and peroneal muscles particularly suffer.

When the stage of wasting is reached the hand takes up a peculiar deformity which is characteristic of the disease. The palm becomes completely flattened out from atrophy of the thenar and hypothenar eminences, while the inner border of the hand/

hand loses its rounded contour and becomes straight from the pisiform bone to the base of the little finger. Owing to the loss of power in the extensors of the little and fourth fingers, these fingers begin to curl up from flexion at the two interphalangeal and metacarpo-phalangeal joints. deformity commences first in the little finger, which is often completely curled up, while the ring finger shows only the commencement of the roll, being only flexed at the inter-phalangeal joints. As the condition advances the remaining fingers become implicated and roll up more and more in this characteristic manner, while the thumb becomes bent and folded into the palm and encircled by the contracted fingers. Once this is accomplished the deformity cannot passively be rectified.

The feet do not as a rule show so characteristic a deformity as the hands, but the toes sometimes curl up in the same way as the fingers. When the affection spreads to the legs and involves the anterior tibial and peroneal muscles, foot drop develops, and then the knees have to be raised higher in walking to bring the feet forward and a "steppage gait" results, such as is seen in other forms of peripheral neuritis.

Cases/

Cases when examined at this stage show sensory anaesthesia to pain, touch and temperature on the inner sides of the hands and feet, or even over larger areas, with impaired sensibility about the hands and feet generally. The knee and elbow jerks are now usually lost and there is distinct inco-ordination of movements of the hands and feet, with swaying of the body when placed in Romberg's The grasp of the hand and muscular power as tested by the dynamometer, are always extremely weak, and opposition, adduction, abduction and extension of the fingers and thumbs cannot be performed. I cannot speak clearly of the electrical reactions of the paralysed muscles as I had only a small Faradic battery with me, but as tested with the Faradic stream the affected muscles gave only feeble reactions, or did not contract at all.

The pupils always react to both light and accommodation, but now and again cases are seen with local paralyses in the way of ptosis and strabismus. The organic reflexes are seldom interfered with,unless the neuritis has mascended to the sacral nerve roots, in which cases there may be frequency of micturition, or other bladder or bowel trouble. The general health so far as the affection itself is/

is concerned remains quite good, but sometimes secondary disease becomes superadded. Cord symptoms in the way of girdle sensation, or spinal pain, are almost always wanting, while the intellectual functions are never interfered with. Percussion of the spine and the hot sponge test yield negative results.

Such is the clinical description of a great number of the cases, but, in many instances, patches with loss of pigment and anaesthesia develop in the skin as in ordinary nerve leprosy cases. These patches occur in a less symmetrical manner than the whitening of the skin in the affected parts, and, although they may occur anywhere on the body, they have a special preference for the paralysed areas. They are not present from the commencement of the disease, but develop later than the sensory symptoms, and in several instances, looked as if they were more an accidental complication than a part of the disease.

Each patch is at first small and rounded and slightly raised, and generally occurs on the arm or leg, and once formed it grows rapidly in size in comparison with the other phenomena of the disease, which are essentially slow in their progress. As the/

the patch enlarges, it loses its rounded shape and becomes oval or irregular, while neighbouring patches tend to coalesce. While it grows in size the centre becomes pale and dry, while the skin thickens and is thrown into scaly folds like a chronic eczema and the surface yields a white scaly powder when scraped. . Later on, as the area becomes paler and paler, and more and more devoid of pigment, it smoothens out, and the surface becomes perfectly white and glossy like a patch of true In other cases, instead of becoming leucodermia. smooth, it becomes thickened, hard and horny. These patches, as they increase in extent, and unite, may reach a huge size, and the border has then often a distinctly serpiginous outline and is darker than the surrounding skin, and I have seen instances of them extending from the legs up to the thighs and round on to the buttock. When the sensibilities of these whitened areas are tested, they are always ultimately found to be anaesthetic, and in them, as in the paralysed areas, first the temperature sense disappears, then that of pain, and last of all, that of common sensation. Many of these patches greatly resemble the white anaesthetic patches of leprosy, while others again, are indistinguishable/

distinguishable at sight from those of leucodermia.

I will now briefly enumerate the features of a few typical cases of the disease so that its course may be more readily followed.

CASE I. Hazrat Gul, male, aged 22, from Swat Kohistan. He complained of tingling and numbness in the toes and of being able to feel nothing with them; there was also formication, but no burning nor loss of power. The symptoms had been present for two months. To the touch the toes were cold, and heat sensations were not well appreciated, while pain and touch sensations were also slightly defective. The plantar reflexes were gone, but the deep reflexes were natural and there was neither wasting of muscle nor trophic change in the skin. This was a case in its very infancy.

CASE II. Kach Kol, male, aged 34, from Dardial Nikpikhel in Upper Swat. He complained of a feeling of ants creeping over fingers and toes, of sensations of burning in the feet, and of a "pins and needles" feeling below the knees. The duration was a month. Testing the temperature sense with hot and cold tubes, he could not distinguish in the hands and feet when the tube was hot, and always/

always called it cold. The appreciation of cold was better, but not quite good, and at times he was uncertain. The touch and pain senses were natural. The grasp of the hand was good and there was no loss of muscular power either in the hands or feet. There was no wasting of muscle or skin change. The palmar and plantar reflexes were lost, but the knee jerks were natural and there was no ataxia. This case was much the same as the first being only in its initial stage, but the defect in temperature appreciation is very characteristic.

CASE III. Alam Gul, male, aged 30, from Deolai Nikpikhel in Upper Swat. He complained of formication in the hands and feet, tingling and numbness in the legs and arms, and of them feeling dead, with difficulty in walking. The affection had lasted eight months. From the history he gave, the trouble had begun first in the feet and later on had implicated the hands.

He was a small man and quite like a woman in appearance, and his hands and feet were as small as a girl's. There was a suspicion of syphilis in his history. Tactile and pain senses were quite natural; the temperature sense was bright in relation/

lation to cold things, but hot tubes were often erroneously interpreted as cold. The plantar and knee reflexes were absent. The pupils reacted well both to light and accommodation. The flexors of the ankle and extensors of the feet were weak, and there was foot-drop. The calves were soft to palpation but were not wasted. The thenar and hypothenar eminences had disappeared and the palms were quite flattened out. The abductor indicis on either side was much atrophied and the other interossei all slightly implicated. The little and ring fingers were curled up in the characteristic manner already described, and the inner border of the hand was flattened. The grip of each hand was weak and co-ordinative movements with either hand were defective.

He stood quite well with the feet together and the eyes closed, and rose naturally from a lying posture. When he walked the feet were raised high above the ground to compensate for the foot drop and the knees were acutely flexed. The muscles of the feet and front and outer sides of the legs would not react to the Faradic current. The forearm muscles contracted well under stimulation, but the thenar and hypothenar muscles not at all. The abductor indicis and other interossei reacted slightly, while the/

the skin of the palms was insensitive to the strongest stimulation.

CASE IV. Shah Rasul, male, aged 35, from Gogdarra in Babozai, Upper Swat. For some years he had felt formication in the hands and feet and below the elbows and knees, and had experienced a "dead feeling" in the feet and hands. As with all the former cases there was no pain, but the hands and feet were muscularly weak. Temperature, pain and touch sensations were all wanting below the knees, and were all impaired in the hands. was no wasting of the calf muscles, or of those of the feet, but the palms were flattened out from the disappearance of the thenar and hypothenar elevations; the inner margin of the hand was straight and the fourth and fifth fingers were bent up in the characteristic manner. The grasp of either hand was weak, and the abductor indicis of both sides was wasted. The skin of the hands and forearms was dry, scaly, and whitish in appearance, while a similar change was seen over the feet and legs. A patch of true leucodermia was present on the inner side of the right wrist towards the front.

The various functions of the body were natural. The knee and plantar reflexes were absent. He walked/

walked quite naturally, and did not drag the feet, while with the feet together and the eyes closed, he remained firm.

CASE V. Sheikh, male, aged 56, from Manglaor in Babozai, Upper Swat. He complained of pains in the ankles and knees, a "dead" feeling of the upper surface of both feet, the outer side of the left leg and in the hands, a burning sensation in the soles, and a feeling of insects crawling on the front of the feet. The duration of his complaint was two years.

Tactile sensation was quite good everywhere, but the outer side of the left leg and the dorsal surfaces of both feet were insensible to pinching. When pricked with the point of a pin he could appreciate the sensation of pain, but could not localise it properly below the knees, although he could in the hands. He could not distinguish a hot tube at all below the knees, and only now and again would recognise a cold one. The knee and plantar reflexes were absent, but all the organic ones were normal. The grasp of the hands was very good, but the little finger of the right hand was curled up. There was a very slight enfeeblement of progression but no ataxia. The palmar and interesseal muscles were not wasted.

CASE VI., Khaperai, female, aged 27. a Gujarani from Jabba in Adinzai, Lower Swat. She had suffered for three years from formication over the back of the neck, and between the shoulders, from burning feelings in the scalp, palms and soles, and from flushings of heat and cold in various parts of the body. Along with those sensory phenomena there was a very characteristic deformity of both hands, for the thumbs were flexed and turned in over the palm while the fingers were bent and curled over the thumb. The tactile sense was natural and so was that of pain. Cold was well appreciated, but heat perception was impaired in the palms. The feet and hands were scaly and white, as in other cases, but no other trophic lesion was present. The plantar reflexes were lost, but the motor system otherwise natural.

CASE VII. Pir Wali, male, aged 25, from
Manglaor, in Babozai, Upper Swat. He complained
of giddiness in the head, tingling and formication
in the tips of both elbows, and the feeling that
these parts were "dead". The duration of the symptoms had been four months. His home was among the
snows and his drinking water was derived from the
snow/

snow itself. On the back of either elbow there was a white anaesthetic patch of skin, about six inches long by four inches broad, and which extended more above than below the joint. The skin of this area was absolutely white and very much thickened, while the surface was scaly and thrown into folds. These patches were almost symmetrical in site and appearance and were insensible to all forms of stimulation. The general condition otherwise was normal.

CASE VIII. Moab, male, aged 49, from Pashat in Bagaur. He complained of his hands and feet being dry and asleep, and of the feeling of insects crawling over them and boring holes into them. There was no pain. He had been a sufferer for three years. Touch, pain and temperature were all impaired below the knees, and somewhat in the hands. The clasp of either hand was weak and the abductor indicis and interessei were wasted. The thenar and hypothenar eminences were atrophied and the palm was perfectly flat. Its inner margin was straight and the little and fourth fingers were bent up. The knee reflexes could hardly be elicited, and the plantar reflexes were absent. The feet were white, dry and scaly, but there was no other trophic change. The right eye was the seat of cataract and there was a slight internal squint.

CASE IX. Gayan, male, aged 39, from Nalkot in Shamezai, Upper Swat. His complaint was burning sensations in the palms and fingers and below the knees, with the feeling of insects creeping over the skin. The duration was three years.

The grasp of the hands was weak, especially on the left side. The forearms were somewhat wasted, the left being the more so. Heat and cold sensations were not appreciated here and there, and pain and touch could not be localised correctly. bowels were slightly constipated, micturition was frequent and the calls urgent. The arms and legs were whitened and the surface of the skin was rough and scaly. The whiteness and scaliness extended up the thighs, and above was limited by a distinct line of demarcation. Here and there over the body, roughly symmetrical on the two sides, were whitish patches, characterised by scaliness and roughness and a perfect anaesthesia. In some places these patches were round or oval, in other places irregu-The centre was always more lightly coloured than the surrounding skin, while the edges were distinctly darker. Some of the older patches were of large size, with irregular outlines, and their centres were white, smooth and glossy, presenting an appearance like patches of leucodermia. of/

of the whitened areas were smooth, others roughened and scaly. On the back of the right heel was a large irregular patch of true leucodermia. The knee reflexes were much exaggerated, but the plantar were absent. The patient stood quite well with his eyes closed and the feet together. The abductor indicis and the palm muscles were wasted; the palm had its characteristic flattened appearance on either side, and the second, third, fourth and fifth digits had the typical folded up appearance. There was an ulcer on the thumb of the left hand and a large callous ulcer on the dorsum of the right foot, while a few months previously there had been necrosis of one of the phalangeal bones.

CASE X., Hashim, male, aged 29, a Gujar from Ghorband, Buner. He had been eight months ill, and complained of feelings of heat in the limbs, and tingling and numbness in the arms below the elbows, and in the legs below the knees. His home was among the snows and he drank snow water. At first there had been a good deal of pain in the knee, ankle, wrist and elbow joints, but that soon passed away. He said where he lived a great number suffered from the same disease.

The skin of the legs from the knees downwards was/

was glossy and white, and there was a trophic ulcer on either great toe. When walking the feet did not drag, and in Romberg's position there was no ataxia. The grasp of either hand was weak, the palms were flat, their inner borders straight and the thumb was bent inwards on the palm with the fingers curled about it. The wrists were straight, but drooped a little when weary. The knee jerks were unimpaired, but the plantar reflexes were lost. The perception of both heat and cold was absent in the whitened area. Touch and pain sensation were very slightly impaired, but lost here and there in small patches.

CASE XI. Ghulam Sarwar, male, aged 54, from Balogram in Babozai, Upper Swat. He complained of pain in the arms below the elbows every cold weather, a feeling of cold in the hands, the sensation of insects crawling on the skin, and of a sleepy feeling in the fingers. The duration of these symptoms was three years.

The disease had begun with tingling and numbness in the hands in the winter time, associated with feelings of formication and burning, and these at first had passed away when summer came. The temperature sense was impaired in the hands, but those of pain/

pain and touch were more or less natural. There was some loss of power in the clasp of the hand on either side, and the palmar eminences were commencing to atrophy, and the little fingers to curl up. With the other symptoms there was associated a whitened patch on the front of either wrist, over which the skin had become thickened, and which desquamated a white scaly powder when scraped. In these areas there was a loss of all kinds of sensibility. The deep reflexes were natural, and there was no ataxia.

CASE XII. Shahdad, male, aged 25, from

Manglaor in Babozai, Upper Swat. He complained of
a numb feeling over the body generally, and was unable to say whether it felt hot or cold, for the
skin surface seemed dead to him. There were also
burning sensations in the palms and a feeling of insects creeping about the hand. The duration of his
illness was ten years.

The condition first began in the hands with the sensory phenomena complained of above, and it was eight years before muscular symptoms were superadded. He stood well, with the feet together and the eyes shut, but in walking dropped the foot on either side, and walked with the typical "steppage" gait. The left/

left foot appeared to droop less than the right.

The penis also was numb and felt dead, but both the bladder and bowels acted naturally.

Tactile sensation throughout was fairly good, and pain less so, but the temperature sense was much impaired and hot and cold tubes could not be recognised as such on the hands, nor differentiated from oneranother below the knees. The grasp of either hand was weak. In both hands the palmar eminences were flattened out, and the inner margin straight, while each little finger had the characteristic curl, and each ring finger to some extent as well. Both abductor indicis muscles were quite wasted and the other interessei small. The right calf was smaller than the left, and both plantar reflexes were absent. The knee reflexes were present, but the right was not well marked. In Romberg's position there was no ataxia, and both incoordination and the muscular sense were fairly good. His house was surrounded by snow during the winter and he drank snow water then, but spring water in the summer. He had a very leprous-like face, but showed no other signs of leprosy.

DIAGNOSIS:

In dealing with this question as to the nature of the disease I shall, in the first place, give my reasons for considering it a form of poly-neuritis, and shall then compare it with the known varieties of multiple-neuritis and discuss its right to be called "endemic".

The first point then to settle was, whether the condition was an affection of the cord, or a primary inflammation of the peripheral nerves. The sensory phenomena themselves, in the way of tingling, formication, pricking, burning and numbness, with their symmetrical occurrence, both in the hands and feet, pointed more to a lesion in the nerves than in the cord, and so did the fact that the symptoms had first developed in the most distal parts as the fingers and toes, and had then crept upwards. Further, when the areas of defective sensibility were mapped out, their outline was found to more resemble the cutaneous distribution of peripheral nerves than any of the sensory cord segments of Head. In the advanced cases the symmetrical wasting of the intrinsic muscles of the hands and feet , the trophic changes in the skin, and the paralytic phenomena, along with sensory symptoms, could only be explainable by a very extensive cord lesion, of which there was no evidence, while the/

the absence of special cord symptoms in the way of girdle sensation and interference with the bladder and bowel reflexes of micturition and defaecation precluded the idea of any cord lesion at all.

Furthermore, the whole clinical features of the disease were almost typical of the affection known as multiple neuritis, for there were symptoms of involvement of all the functions of a nerve and preservation of the bladder and bowel reflexes, with a symmetrical affection of both extremities, and the history of commencement at the periphery. There was therefore little difficulty in recognising the disease as a form of poly-neuritis.

The second point to be established was whether this form of peripheral neuritis corresponded with any of the varieties described, or not. The known varieties may be shortly grouped together as

- (a) Certain acute forms, due to exposure, the poison of alcohol, or to the toxines of specific infectious diseases, as Influenza, Diphtheria, Septicaemia, etc.
- (b) Certain chronic forms, due to the metallic poisons of lead, arsenic or mercury; or to the organic poison of syphilis, and
- (c) <u>Certain endemic forms</u>, due to Beri-beri, malaria and leprosy.

The acute forms were speedily eliminated from the/

the diagnosis, for they all run an acute course with a tendency to spontaneous cure, while this disease is exceedingly chronic and slowly progressive.

Further, the varieties due to influenza, diphtheria and septicaemia are not manifestly peripheral, and diphtheria and influenza are almost unknown in these regions. Alcoholic neuritis was soon also disposed of, for the people were strict Mohammedans, who are forbidden by their religion to take wine in any form, and the cases themselves bore no resemblance to alcoholic neuritis.

The chronic forms due to mercury, lead, and arsenic too, needed little consideration, for in no case was there a history of any of those poisons. Besides, since 1897 I have seen a number of cases of mercurial and arsenical neuritis in Lahore, from unfaithful wives slowly poisoning their husbands, and can definitely say that such cases do not at all resemble this affection. Further, the drop wrist of lead palsy, or the tremors of the mercurial and arsenical forms, were never present among my cases. Syphilis was easily excluded from the want of history of a chancre, or external evidences of infection, but apart from that, syphilis very rarely causes peripheral neuritis.

There then only remained the three endemic forms due/

due to beri-beri, malaria, and leprosy. Beri-beri is not a disease of these parts, and the only cases which have ever occurred were a few among our troops on the Malakand in 1899, the cause of which was traced to India. Moreover, none of my cases presented the heart conditions, or showed the oedema and acute onset of beri-beri. Malarial neuritis was easily excluded, for none of the patients came from the malarial zone of the low valleys, where malaria is most fearfully rife, but all from high and malaria-free districts, while of the many thousands of cases of malaria which come to hospital none of them showed peripheral nerve symptoms.

I have left the consideration of leprous neuritis to the last, for it is hard to separate the two affections from one another, and, from want of definite pathological observations, I cannot conclusively say that the two diseases are distinct—although I believe they are. Although there are many points in common in both affections, I shall be able to point out many features of difference, and to show that, if not a separate disease, it is an exceedingly rare form of leprous neuritis. It is also possible that in some patients the two affections may co-exist, and in one or two instances I have thought that to be the case. The points of resemblance/

resemblance between the two diseases are:-they are both endemic in the same regions, and both
run an exceedingly chronic course; they both show
white anaesthetic patches in the skin, and are both
associated with peripheral neuritis.

Leprosy however occurs in India as well as beyond the border, while apparently this disease does not, and during my seven and a half years in India Inhave had a fairly large experience of leprosy in all its forms, but cannot remember having seen a case with features exactly the same as these. leprous neuritis there is, too, generally a history of fever at the onset, with the presence of a transitory macular eruption, while the anaesthetic pale patches develop with or before the sensory symptoms, and sooner or later the superficial nerve trunks show thickening. In my cases there was no history of initial fever or macular eruption; the anaesthetic white patches developed later than the sensory changes, while many cases showed no white anaesthetic areas at all, and in no instance was a superficial nerve trunk enlarged. In anaesthetic leprosy, again, the phenomena are not necessarily symmetrical, and usually at some or other time other features of leprosy are present. The symptoms of my cases were/

were always symmetrical, and in no patient did I observe leprous tubercles under the skin, although one patient had a somewhat leprous face. It, further, is rare for leprous symptoms to confine themselves to the extremities, and very commonly the face is involved, while I found it rare to have changes other than in the limbs. A minor point of difference is that in the leprous patches pain and temperature both disappear together, but in my cases temperature was always affected first. Further there is the fact that the people themselves do not regard the disease as leprosy.

These facts and my own experience make me incline to believe that this form of peripheral neuritis is distinct from ordinary nerve leprosy, and as I have only observed it in the confines of a definite region, and have not heard of or seen cases outside that region, I believe I am justified in calling it "endemic"

AETIOLOGY.

I have seen twenty-four instances of this affection in all. Of these twenty-three were males and one a female. Twenty-one were trans-border Pathans, two Gujars, or cowherds, and one a Hindu living over the frontier. All the cases were adults, the youngest/

youngest being 22 and the oldest 60. Twenty-one of the patients were cultivators by occupation, 2 were cowherds, and 1 a baker.

The disease appears to be confined entirely
to the trans-frontier region and to be endemic
there, for cases of the disease do not occur in the
plains of India, and patients removed from the endemic area always have their symptoms benefited.

So far as I can judge the specially endemic areas
are the Babozai and Nikpiknel valleys in Upper Swat,
with the adjacent Ghorband region on the Buner side,
from whence most of my patients came. A few isolated
cases came from Bajaur, Dir, Kohistan and the Utmankhel country and one from Baluchistan, but in these
regions the affection is more uncommon.

The disease, for the most part, seems to affect a high elevation and one of the most constant features in the history of the cases was association with intense cold. The lowest elevation from which my patients came was 3000 feet, but some were living at greater altitudes, even up to 9000 feet or more. Throughout all this region the hills are more or less under snow from November till March down to about 4000 feet, and it was usually during this period that the symptoms began. It is hardly possible though/

though, that extreme cold can be other than a predisposing cause, for the after history points more to some toxic or toxaemic poison continuing to act on the peripheral nerves. Whatever the poison may be, it must be of low virulence from the slowness with which it produces symptoms and spreads, or, it may be, if there is some localised chronic poisoning, that re-infection after re-infection occurs. New-comers into these regions do not develop the disease, for in all the varied expeditions which have occurred beyond the border, I have never heard of any of our troops developing the affection. A prolonged residence in these areas thus appears to be necessary for its inception.

Alcohol certainly plays no part in its causation for the reasons I have explained, nor apparently do the metallic poisons of lead, mercury, and arsenic. Syphilis again has no place; nor has malaria, for these regions are well above the malarial zone so clearly defined in the valley itself. Leprosy possibly may be a cause, or it may itself develop in the peripheral nerves when their vitality is lowered by this disease. It is also possible that diseased states of grain, or other food, or the consumption of some special article of food, may play a part in causing the disease, but on this point I cannot definitely speak as no white man has up to the present penetrated so far into upper Swat.

PATHOLOGY:

This, I can only speculate upon for I have never seen a fatal case of the disease, nor did I have the requisites with me for making microscopic or bacteriological investigations. The clinical features, however, point to the affection being a very chronic interstitial neuritis or perineuritis of the peripheral nerves. It would appear to commence in, or around the peripheral terminations of those nerves which have the longest distance to travel and whose terminals are therefore farthest removed from their nutritive centres in the cord and the inter-vertebral ganglia. These nerves are those of the fingers and toes, and are also those most readily affected by cold. I believe the affection begins for the most part as a perineuritis of the peripheral nerves of the fingers and toes, and spreads slowly up the nerve sheaths towards the cord. In addition to ascending it would appear to penetrate in to a greater or less extent between the nerve bundles as an adventitial neuritis and in the compression or destruction of nerve fibrils which goes on, the slender, highly specialised, and feebly protected sensory nerves first suffer. Of these the most highly specialised are those which carry temperature sensations, next in order those for pain, and least/

least specialised those for common sensation, and it is interesting to note that the first nerves affected are those most highly specialised and those last affected the more lowly specialised nerves of touch.

As the perineuritis creeps up the nerve sheaths it does not appear to cause nodular thickenings, as in the case of leprous neuritis, or if so the swellings have never been palpable. In some instances the neuritis may ascend even to the spinal nerve roots, and it is possible that in some cases it may extend to the cord itself, combining a secondary myelitis with the primary neuritis.

DIFFERENTIAL DIAGNOSIS:

Leprous neuritis is the only affection which can well be confounded with this disease, and I have already indicated the main points of difference in the two conditions. Syringomyelia somewhat resembles it in regard to the sensory changes, but the distribution of the anaesthesia is different, and in syringomyelia the legs generally show a spastic paralysis, while the disease itself is rare. Progressive muscular atrophy shows rather similar changes in the way of wasting of muscles, but is not associated with sensory phenomena or with trophic changes/

changes in the skin. The ataxia, incoordination, loss of knee jerks, and sensory phenomena of advanced cases may appear to simulate locomotor ataxia, but the visceral crises, lightning pains, interference with the lumbar reflexes, and Argyll Robertson pupil, of that disease are all wanting.

PROGNOSIS:

In the early sensory stage the prognosis is quite good, if cases can be removed from the endemic area and vigorously treated, for recoveries often occur. In the advanced cases all that can be hoped for is to stay the progress of the disease, for the affection itself cannot then be eradicated. The disease, however, is very chronic and does not appear to materially shorten life, and most probably death usually occurs from some intercurrent disease.

TREATMENT:

In the first place it is of importance to remove the patient to a warmer locality, such as the Punjab, or to get him to live lower down in a warmer part of the valley. If that is not possible the body must be protected as much as possible against the severe cold of the winter by thick woollen clothing for the body, woollen gloves for the hands,/

hands, with warm socks and moccasins for the feet.

A change of diet along with change of climate is also advisable.

Local treatment is always most beneficial and the best form is massage with stimulating oils and liniments. Those I have most used are turpentine, mustard oil, gurgun and chaulmugra oils and the acetic turpentine liniment of the British Pharmacopoeia. Electricity is also of great value and I used the faradic current and wire brush in all my cases with great benefit. The subcutaneous injection of strychnine into the hands and feet was also useful in some of the cases.

For the anaesthetic patches and thickened areas of skin I used 4% salicylic acid lanoline and 25% ichthyol lanoline with benefit, while the official ointments of sulphur, creosote, carbolic acid, and the acid nitrate of mercury were all found useful. I know, however, of no remedy which can restore colour to the decolourised patches.

Internally, the remedies used with most advantage were strychnine, the iodide of petash, and the perchloride of mercury, while in some cases benefit was derived from the administration of Burroughs and Wellcome's orchitic extract tabloids in doses of from 5 to 15 grains in the day.

do mint star Satheland, m. 13. Captain, Indian of which I was.