

ULNAR AND MEDIAN NEURITIS DUE TO LEPROSY

REPORT OF ONE HUNDRED CASES TREATED SURGICALLY¹

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Although leprosy is a systemic disease, it affects only certain tissues to a significant degree. Those chiefly involved include the skin, nasal mucous membranes, and the peripheral nerves. Involvement of peripheral nerves is always present in some measure; it leads to the majority of permanent sequelae and stigmata of the disease.

During the past eight years surgical procedures upon peripheral nerves have been employed at the United States Public Health Service Hospital at Carville, Louisiana, to relieve unbearable paresthesias or intense pain, and to prevent progressive paralysis of muscles supplied by the affected nerves. More than a hundred operative procedures have been carried out, from which pathologic material has been obtained, yielding precise correlations of the state of the disease in the nerves with the clinical aspects of the patient's progress.

Thus, a survey of the status of leprosy within the nerves during the period of activity is made possible. Leprosy affects the ulnar, median, peroneal, and tibial nerves in descending order of frequency and in a predictable manner. Virchow, in 1864⁽³⁾, observed that the nerve swellings occurred unevenly, especially where the nerves pass close to the bony prominences of the hand and elbow. The ulnar nerve is involved at the elbow just proximal to the medial epicondyle of the humerus. Intense localized pain may radiate distally into the pathways of its peripheral distribution, with enlargement of the nerve and progressive paralysis of the distally innervated muscles.

Often the first presenting sign of leprosy is an insidious onset of hypesthesia along the ulnar aspect of the forearm⁽²⁾. Hypesthesia may be present for several years before the disease is recognized. Leprosy should always be kept in mind in the differential diagnosis of this finding. Neural involvement may become more extensive, with anesthesia developing to involve the ulnar aspect of the hand, ring, and little fingers. Even though there may be no associated objective findings in the area of the anesthesia, a dermal biopsy specimen from this region will show fibrosis of cutaneous nerves or an active leprosy neuritis.

Certain objective findings of early involvement of the ulnar nerve are well recognized. These include the following:

1. Loss of insensible perspiration at the fingertips of the 4th and 5th fingers.
2. Flattening of the rugae of the fingertips with resultant loss of traction on palpation by the examiner.
3. Weakness of the *opponens digiti quinti*. This is one of the first signs noticed

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in early ulnar motor involvement. It is evident in inability of the patient to bring the tip of the little finger into direct opposition to the tip of the thumb.

4. A noticeable flattening of the hypothenar eminence due to the loss of the abductor and short flexor of the little finger, following progressive involvement of several muscles.

5. Weakness of the flexor digitorum profundus, resulting in inability to stabilize the distal interphalangeal joint of the little and ring fingers in flexion. This finding, along with evidence of paralysis of the flexor carpi ulnaris, indicates involvement of the ulnar nerve at a higher level, i.e., the elbow or above.

6. Clawing of the ring and little fingers in late stages of ulnar palsy. These result from hyperextension at the metacarpophalangeal joints, and from flexion contractures of the interphalangeal joints. Also, with loss of the dorsal and volar interosseous muscles, there is diminution (or loss) of ability to abduct and adduct these fingers. Flexion contracture of the tip of the thumb results from loss of the actions of the deep head of the short flexor and abductor brevis (median), which contribute to extension of the distal phalanx in conjunction.

The median nerve is usually involved at the level of the wrist, just proximal to and as it passes beneath the volar carpal ligament. Enlargement, with accompanying pain and tenderness, is seldom encountered prior to involvement of the ulnar nerve. Recurrent episodes of median neuritis are eventually followed by a low median palsy; they are manifest clinically by the carpal tunnel syndrome. The primary effects of median nerve involvement include sensory loss in the peripheral distribution of the nerve to the skin, loss of the lumbrical actions to the index and long fingers resulting in clawing, and loss of the action of the abductor brevis and opponens of the thumb. Involvement of the median nerve alone is rare. It is almost always associated with ulnar involvement at the level of the elbow.

A high ulnar and low median paralysis presents the composite clinical manifestations of an unopposable thumb, hyperextension of the metacarpophalangeal joints, and flexion contractures of all interphalangeal joints. Under these conditions, the patient learns to develop a compensatory mechanism by flexing the wrist to relax the flexor digitorum sublimi and profundus tendons, whereby tension is placed upon the extensor tendons. By obtaining increased extension of the fingers, the patient is able to grasp and pick up objects more easily. Rehabilitation, particularly in the form of reconstructive hand surgery, is less successful at this stage than if instituted after the patient has learned to substitute for lost function. The "main-en-griffe" deformity of the hand should pass into well-deserved extinction by supplementing medical therapy with early institution of physical therapy and surgical measures.

SURGICAL PROCEDURES

In a series of one hundred cases of leprosy neuritis involving the ulnar and median nerves, surgical decompression by incision and removal of a portion of the neural sheath at the site of enlargement was found to be paramount in relieving paresthesias and excruciating pain, and in delaying or preventing progressive paralysis. The swelling of the neural sheaths at the site of involvement is due to varying degrees

of edema and thickening. The fibrosis and adhesion of the enclosed nerve are proportional to the number of acute neuritic episodes previously experienced. Grossly, the affected nerve has a dull yellow color in contrast to the glistening white appearance of normal nerves. Its diameter is increased from two to four times normal. Enlargement is due to edema produced by the extra- and intraneural infiltration by *M. leprae* and the accompanying inflammatory response. Intraneural fibrosis occurs in the more advanced cases. Abscess formation is encountered less frequently. When present, abscesses consist of frank suppuration revealing leucocytes and acid-fast bacilli; no growth, however, is obtained on artificial media.

The surgical procedures do not alter the basic disease process; rather they allow it to continue under a more favorable environment by releasing the nerve from entrapment. The edema accompanying recurrent acute episodes of neuritis is believed to lead to strangulation of the nerve, especially when its sheath is inelastic as a result of fibrosis from previous inflammatory episodes. In the case of ulnar neuritis due to involvement at the elbow, the nerve is freed of its adventitia and mobilized so that it may be brought anterior to the medial epicondyle without tension. The flexor muscles of the forearm are divided at the epicondyle and reflected laterally until the median nerve is recognized in the antecubital fossa. The thickened sheath is resected and the nerve is placed anterior to the medial epicondyle, deep to the flexor muscles of the forearm. This places the enlarged nerve in a location preventing its subjection to trauma, which usually occurs at the site of its original position. Placing the nerve beneath a muscle mass seems desirable, since such locations are rarely the site of leprous neuritis. Usually an articular branch of the ulnar nerve is sacrificed at the elbow to facilitate its relocation. The procedure serves also to interrupt the association of the nerve with other structures that may be involved, viz., the vascular and lymphatic vessels.

The median nerve cannot be transposed at the level of the wrist, where it is commonly affected. The surgical procedure, therefore, consists only of dividing the volar carpal ligament in its entire width well into the palm as it overlies the nerve. Careful dissection is essential to avoid the interruption of important motor branches. Specimens of small sensory branches and the sheaths of larger nerves are thus made available for histologic study.

PATHOLOGY

Seventy nerve specimens were obtained during surgery, for pathologic study, in this series of 100 cases. They consisted of small nerves, usually articular branches of the ulnar nerve, obtained in the region of the elbow. The study of these nerves indicated two distinct phases of leprous involvement, an infiltrative and a granulomatous. In the infiltrative phase there is an expanding disease process consisting of infiltration of bacilli both intra- and extracellularly, within or along

nerve elements, such as axis cylinders, myelin, and endoneural connective tissue, and within Schwann cells. The latter observation, coupled with the conspicuous absence of the process in the central nervous system, optic nerve, and nonmyelinated autonomic nerves, has led to the plausible theory that the acid-fast bacilli of leprosy may have a primary affinity for the Schwann cell⁽¹⁾. The granulomatous process differs in no way from the lesion in the skin, and is the lesion in the larger nerves leading eventually to healing with dense fibrosis. The chronicity of the disease process is observed microscopically as well as clinically by constant simultaneous regressions and advances, often within cells adjacent to one another, bacilli proliferating in one while disappearing from its neighbor. This process is intermittent and unstable, and the factors that determine its progress, favorable or unfavorable, are not known. Trauma, stretching, compression, inflammation, edema, and temperature, all have been cited as possible factors.

In this series the earlier infiltrative lesion was most commonly seen, only 20 per cent of the cases being of the granulomatous variety. This may be accounted for by the acute neuritic symptomatology that often prompted surgical intervention. In addition to the findings indicating neuritis, in 27 specimens infiltration of perineural tissues was noted, in which the lepromatous process was of sufficient extent to account in part for the consistent finding of enlargement of the involved nerve to two to four times its normal size. Eight specimens of nerves were positive for acid-fast bacilli in cases in which the skin was not positive. In three cases epitrochlear lymph nodes were removed adjacent to the ulnar nerve. In these, the bacilli stained much more poorly in the node than in the nerve branch. This suggests that the acid-fast bacilli in the nerve are perhaps afforded some protection from the systemic treatment of the disease, and this could account for the frequent finding of activity within peripheral nerves long after the patient has been pronounced bacteriologically arrested on the basis of skin scrapings. This lends support to the idea that treatment for life is indicated, to maintain a state of relative inactivity once this stage is reached.

A composite picture of leprosy as it affects the radial, median, and ulnar nerves at various levels of the upper extremity has been gained through the dissection of a series of autopsy specimens. The following are some observations made from this study.

Ulnar nerve.—1. At the wrist the motor and sensory elements are solidly involved in long-standing cases. In mild cases demonstrating only slight hypesthesia clinically, there is moderate infiltration of the superficial branch supplying sensory fibers to the skin of the palm and ring and little fingers. Only a few millimeters away, in the deep motor branch going to the intrinsic muscles of the hand, there is no involvement whatever. This supports the theory that this is an ascending sensory nerve infection.

2. Between the elbow and the wrist there is less infiltration than at the wrist in the very active cases, and perhaps none in the slight or early cases.

3. At the elbow the nerve tends to be more heavily infiltrated with acid-fast bacilli, and there is more accompanying cellular reaction than at the wrist, in both the advanced and the mild cases.

4. Between the elbow and the axilla the process begins to quiet down in the very active case and tends to be absent in the mild case.

5. At the axilla the process is usually absent, at most, very slight.

Median nerve.—1. At the wrist there is heavy infiltration in the advanced cases and moderate involvement even in the mild cases, some bundles being spared completely in the latter.

2. From the wrist to the axilla (where the nerve is continuously deep to muscle), there is relatively little involvement in the heavily infected cases. Curiously, there is little if any involvement at the elbow level in all cases, even though this structure is only a few centimeters away from the universally and heavily involved area in the ulnar nerve.

Radial nerve.—This nerve is usually involved in its cutaneous distribution distal to the midforearm, but seldom proximal to this point.

In summary, all three nerves are involved at the wrist level, but to a diminishing degree proximally, with the notable exception of the ulnar nerve, as described.

CLINICAL RESULTS OF SURGICAL TREATMENT

One hundred neurolyses were performed for ulnar and median neuritis. Eighty-one procedures were carried out to relieve patients from complaints due to neuritis, which included severe pain beginning at the site of involvement and radiating distally throughout the cutaneous distribution of the nerve. In 57 per cent of the series there were objective clinical findings, of varying severity, due to ulnar or median paralysis. Fifteen per cent of the operations were carried out prophylactically and empirically, on the basis of experience gained from patients who had previous involvement of the contralateral ulnar or median nerve. In this entire series of 100 cases, 76 had palpably enlarged nerves before surgery.

In evaluating the clinical results of this procedure, one or more benefits and the percentage incidence are shown in Table 1.

TABLE 1. *Results of 100 neurolyses in 77 patients.*

Effect	Per cent of cases
Relief of pain	99.0
Neurologic changes:	
Halted	62.0
Slowed	14.0
Improved function	30.0
No relief of any kind	1.0

SUMMARY AND CONCLUSIONS

Leprosy is a chronic infectious disease with clinical and pathologic involvement of certain nerves, occurring in a predictable and specific manner. Anesthesia of normal-appearing cutaneous areas is often the presenting symptom and sign of leprosy.

From the studies here reported it appears logical to assume that this process may be the primary lesion of the disease, the skin and other organ systems being involved subsequently and in varying degree.

Enlarged nerves should arouse the suspicion of the examining physician, especially when they are located in the sites of predilection in leprosy. Many cases may be diagnosed in relatively early stages even though they do not present signs of anesthesia and muscular atrophy in relation to the involved nerve.

During neurolysis on idiopathically enlarged nerves, a small non-motor branch, together with a portion of the sheath, should be sacrificed for pathologic study with acid-fast as well as H & E stains of the sections.

Excellent relief of pain by surgical decompression of acutely inflamed nerves seems to be obtained in all cases. Results, mostly favorable, have been noted in the effect of these procedures in slowing or halting the progression of neurofibrosis.

Frequently a nerve lesion is positive for acid-fast bacilli long after the patient is considered bacteriologically arrested on the basis of negative skin scrapings. This fact supports the view that treatment for life is indicated, in order to maintain a state of relative inactivity.

RESUMEN

La lepra es una enfermedad infecciosa crónica con envolvimiento de ciertos nervios, que ocurre de una manera específica y predicable. La anestesia de áreas cutáneas de apariencia normal es frecuentemente el presentante síntoma y signo de la lepra. De los estudios aquí comunicados, parece lógico asumir que este proceso pueda ser la lesión primaria de esta enfermedad, siendo envueltos subsecuentemente y en grado variado la piel y otros órganos y sistemas. Nervios agrandados deben despertar las sospechas al médico examinante, especialmente cuando ellos están localizados en los sitios de predilección de la lepra. Muchos casos pueden ser diagnosticados en los estadios relativamente tempranos, aunque ellos no presenten signos de anestesia y atrofia muscular, en relación con el nervio envuelto.

Durante la neurolysis de nervios agrandados idiopáticamente, deben ser sacrificados una pequeña rama no motora junto con una porción de la vaina, para el estudio patológico de las secciones coloreadas para ácido-alcohol-resistentes como así también con hematoxilina-eosina.

En todos los casos parece obtenerse un excelente alivio del dolor por decompresión quirúrgica de los nervios agudamente inflamados. Resultados mayormente favorables se han notado con estos procedimientos, en sus efectos de disminución o detención de la progresión de la neurofibrosis.

Frecuentemente, una lesión del nervio es positiva para los bacilos ácido-alcohol-resistentes, mucho después que el paciente es considerado bacteriológicamente detenido sobre la base de los raspados de piel negativos. Este hecho apoya el punto de vista de que el tratamiento por toda la vida es indicado, con el objeto de mantener un estado de relativa inactividad.

RÉSUMÉ

La lèpre est une maladie infectieuse chronique accompagnée de l'atteinte clinique et pathologique de certains nerfs. Cette atteinte survient de manière spécifique et prévisible. L'anesthésie au niveau de zones cutanées d'apparence normale est souvent le symptôme de lèpre qui attire l'attention.

Des études qui sont ici présentées, il apparaît logique de déduire que ce processus peut être la lésion primaire de la maladie, la peau et les autres systèmes et organes n'étant affectés qu'ultérieurement et à un degré variable.

Les nerfs épaissis devraient éveiller les soupçons du médecin, surtout lorsqu'il s'agit de nerfs situés aux endroits de prédilection pour la lèpre. Beaucoup de cas peuvent être diagnostiqués à un stade relativement précoce de la maladie, même s'ils ne présentent pas de signes d'anesthésie ou d'atrophie musculaire correspondant aux nerfs atteints.

Pendant que se produit la dégénérescence musculaire des nerfs typiquement épaissis, on devrait sacrifier un petit filet nerveux nonmoteur, sans négliger une partie de la gaine, afin de procéder à une étude histopathologique qui devrait comprendre une coloration des coupes pour recherche des acido-résistants ainsi qu'une coloration par H-E.

Un soulagement très net de la douleur semble être obtenu dans tous les cas par la décompression chirurgicale des nerfs atteints d'inflammation aiguë. Des résultats favorables ont été notés dans la plupart des cas à la suite de ces mesures, aboutissant au ralentissement ou à l'arrêt de la fibrose nerveuse.

Souvent, une lésion nerveuse reste positive pour les bacilles acido-résistants longtemps après que le malade a été considéré comme bactériologiquement blanchi sur la base de frottis cutanés négatifs. Ce fait confirme l'opinion qu'il est indiqué de poursuivre le traitement pour la durée de la vie, afin de maintenir un état d'inactivité relative.

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