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Nerves in the Arm in Leprosy

I. Clinical, Electrodiagnostic and Operative Aspects[1, 2]

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Every leprosy patient suffers from peripheral nerve involvement as a result of the disease. This may vary from the involvement of intradermal nerves in a cutaneous patch to a major lesion in the nerve trunk. There is, hence, no nonneural leprosy. On the basis of the WHO estimate of 10 to 15 million patients suffering from this disease in the world today, leprosy presents one of the largest problems in diseases of the peripheral nerves.

Despite the magnitude of the problem and the fact that about 20 per cent of leprosy patients suffer from major sensory and motor deficits due to involvement of the nerve trunks of the extremities, detailed clinicopathologic studies of nerves in leprosy have been few, represented primarily by the reports of Dehio (9), Woit (21), Klingmüller (14), Dastur (6, 7), Weddell *et al.* (20), Antia *et al.* (1) and Dastur *et al.* (8).

Some recent studies of larger nerves, for example by Thomas (19), deal chiefly with physiotherapeutic assessments and observations on limited operative exposures. Perhaps the first most comprehensive report on the clinical aspects of the neurologic disorder in leprosy is that by Monrad-Krohn (16), although Barraquer (2) had preceded him in a consideration of "lèpre nerveuse" against the background of other peripheral nervous disorders. Cochrane (4) has summarized some aspects of neuritis in leprosy.

The present study presents comprehensive clinical, electrophysiologic, operative and pathologic observations, with follow-up on patients suffering from peripheral nerve trunk involvement in leprosy. In this first of two papers the clinical, electrodiagnostic and operative findings will be presented and discussed. The gross pathologic changes observed at operation will be included here, but the histopathologic findings, clinicopathologic correlations and a discussion of pathogenesis will be given in the second paper.

MATERIALS AND METHODS

Clinical. The 22 patients were selected chiefly from the Chembur Beggars' Home for Leprosy Patients, and the Acworth Leprosy Hospital in Bombay (four of the patients came from outside Bombay). All presented the polyneuritic form of leprosy, this being a precondition for their selection. All volunteered to participate in the investigation, including surgery which was performed primarily for nerve pain. All were males, ranging in age from 18 to 41 years with an average age of 25 years, who had had leprosy for from one to 27 years, the average being eight years.

Twenty-seven nerve exploration procedures were conducted on the 22 patients, five of them undergoing the operation and the investigation on both arms.

The type of leprosy in these patients was difficult to assess. Their classification was based on three criteria: (1) assessment by leprologists of the Acworth Leprosy Hospital; (2) assessment by Dr. R. G. Cochrane

[1] Received for publication 24 March 1969.

[2] The present investigation stemmed from the collaboration between the Tata Department of Plastic Surgery and the Neuropathology Unit, then of the Indian Council of Medical Research, at the J. J. Group of Hospitals, Bombay, and from support by Grant OVR-IND-22, from the Social and Rehabilitation Services of the Department of Health, Education and Welfare of the U.S. Government. The project was begun in February 1963 and this study of the nerve trunks of the upper extremity constitutes the major investigation.

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during February to March 1965 (as a consultant leprologist); and (3) assessment by us on the basis of the histologic findings in the nerves and, at times, in the skin, together with evaluation of the clinical histories. On these bases there were three lepromatous, 12 dimorphous and seven tuberculoid cases in this study.

Electrodiagnostic. The tests were carried out using a single channel R.A.F. Type IIIc electromyograph. The electrical stimulus in the nerve conduction studies was delivered through an isolated transformer, and had a duration of 300 μsec. and a maximum strength of 200 volts. Conventional concentric needle electrodes were used for electromyography. Surface electrodes (in 32 instances) or concentric needle electrodes (in the remaining) were used for recording the response. As far as possible, both operated and unoperated arms were examined. Some patients were examined more than once postoperatively. Electromyographic studies were carried out on 28 ulnar innervated muscles (generally the abductor digiti minimi, and in many cases the flexor carpi ulnaris and interosseous muscles), and on 17 median innervated muscles (generally the abductor pollicis brevis muscle). The presence of spontaneous activity and insertional activity at rest was noted. The motor unit potentials associated with submaximal contraction were studied with respect to their morphologies and dimensions. The interference pattern on resisted effort was studied.

Thirty-nine ulnar and 38 median nerve conduction records were obtained. The muscles of reference were the abductor digiti minimi and the abductor pollicis brevis respectively. The method described by Hodes *et al.* ([11]) was used to calculate the motor conduction velocity between the elbow and the wrist. The ulnar nerve was stimulated supramaximally at the wrist just proximal to the pisiform bone and at the elbow just proximal to the epicondyle; the median nerve was stimulated at the wrist as it lay between the tendons of the flexor carpi radialis and palmaris longus, and just proximal to the elbow crease, medial to the brachial artery. The latency of the muscle response was measured from the stimulus artefact to the onset of the deflection. The shape of the action potentials obtained at the wrist and elbow were compared in each case.

Surgical. All cases, except one who was operated under brachial plexus block, were operated upon under general anesthesia. The surgical procedure was generally carried out under a tourniquet, consisting of the inflated cuff of a sphygmomanometer wrapped around the upper arm, for 1½ to 2½ hours.

For maximal exposure of the ulnar nerve, a longitudinal incision extending from midarm to the nerve at the wrist, running anterior to the medial epicondyle, was found adequate. The incision was staggered transversely at the wrist and extended into the palm up to the distal palmar

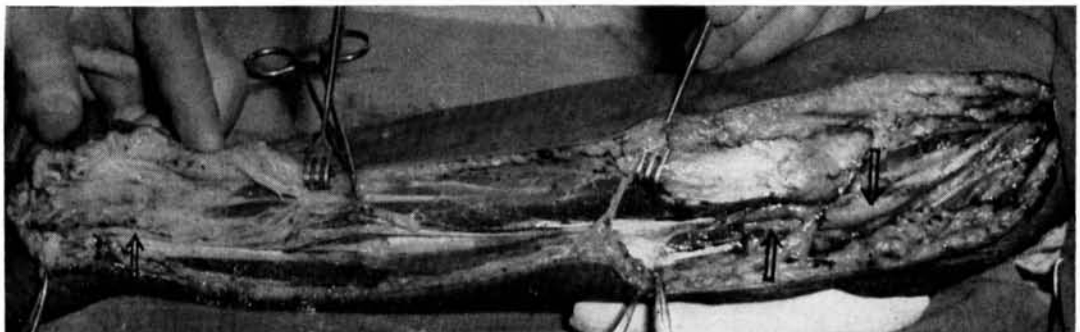
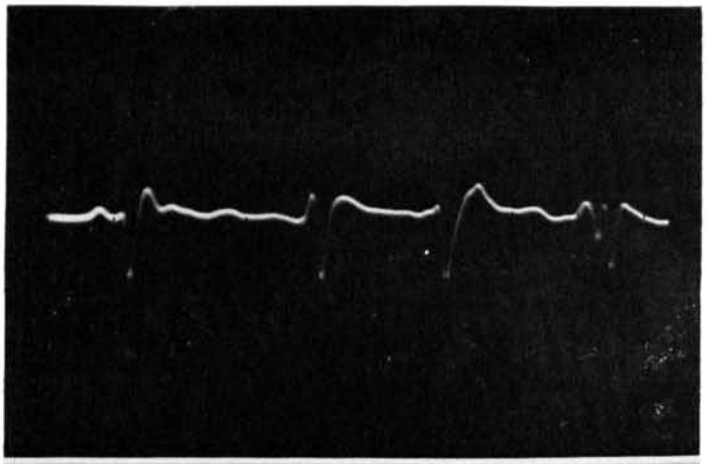


FIG. 1. General view of surgical exposure of nerves and tissues from the middle of the palm to the middle of the arm. Note thickened ulnar nerve above and below the medial epicondyle (arrows) and of normal thinness at the wrist (arrow) (P.S.U. 486; N.P.C.-412).



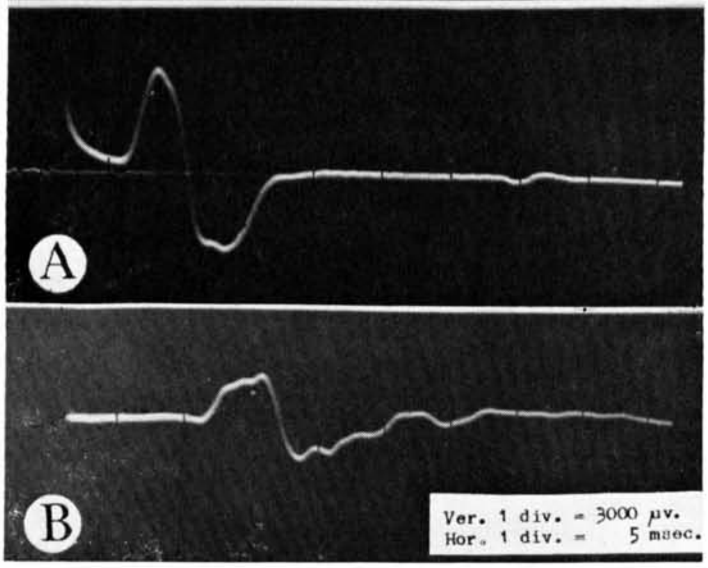


FIG. 2. Right abductor digiti minimi electromyogram (using concentric needle electrodes) of maximal contraction, showing single oscillations, with only two units. This marked drop-out of motor units suggests denervation. Amplitude 3,000 uv, duration 6 msec.

FIG. 3. Same muscle as in Figure 2. Ulnar nerve conduction (surface electrodes). (A) Stimulation at wrist and (B) stimulation at elbow; distal latency (4.5 msec); elbow-wrist conduction velocity (48.1 m/sec) is minimally slowed; in addition, there is attenuation of the elbow response (amplitude 4,000 uv) as compared to that obtained from the wrist (amplitude 9,900 uv).

crease, in order to provide exposure of the median nerve in the lower forearm and of the palmar branches of both the ulnar and the median nerves (Fig. 1). Details of further exposure of the two main nerve trunks are noted under OBSERVATIONS. Care was invariably taken not to lift a nerve trunk completely off its bed. All cutaneous branches in the arm and forearm were carefully dissected, as also were the motor branches to various muscles in the forearm and the palm, notably those to the flexor carpi ulnaris and to the thenar and hypothenar muscles.

Two times magnifying spectacles were employed wherever necessary during the dissection and while undertaking nerve biopsy. A Zeiss binocular operating microscope with magnification up to 32x was also utilized to observe nerve details and their environment in selected areas as well as to visualize the intraneural structure on incising the epineurium and after biopsy.

Generally after release of the tourniquet (and occasionally before), the nerves, dissected bundles, motor branches and muscles were stimulated by a pulse of 0.01 milliseconds from a R.A.F. Type V stimulator. The pulse strength was variable from 0-200 volts and the lowest voltage required for a smallest muscular twitch was recorded.

Intravital staining of the flexor carpi ulnaris near its motor point, using methylene blue for staining nerve fibers, was carried out in a few cases. Biopsy specimens were taken of selected cutaneous nerves, bundles from nerve trunks, muscles and deep fascia.

In three patients, who preoperatively showed total loss of sensory and motor function of the nerves as assessed clinically and electrically, total excision biopsy was undertaken of the ulnar nerve in the arm and the forearm, and of the median nerve in the forearm distal to its last motor branch.

OBSERVATIONS

Clinical. The duration of nerve symptoms in the operated arm, usually expressed as pain, varied from three months to 12 years with an average of two years and two months.

No patient noted any form of motor deficit as the initial defect. All patients gave a history of onset with a cutaneous patch, generally with impaired sensations, but not all of them were certain about the latter. Six began with a small or large patch on the arm that was eventually operated, eight with an anesthetic patch on a leg, seven with diffuse lesions on the face and one with an acroteric anesthetic lesion on a leg.

On direct questioning the majority of the subjects gave a history of pain in the ulnar nerve, a few giving a history of pain in the lateral popliteal or median nerve. There was one very chronic case from whom no history of nerve pain could be elicited.

Clinical findings related to nerve trunks. ***Ulnar nerve.*** This was invariably the nerve maximally affected, the site of greatest involvement being the lower third of the arm; in 23 cases it was tender, and in 20 it was swollen in this region, while in the ulnar groove at the elbow it was swollen or tender in only 12 cases. Some tenderness and/or thickening were detected in the ulnar nerve at the wrist in only two. In the same subjects on the unoperated side, the ulnar nerve was found thickened in nine cases in the lower third of the arm, and in four in the groove.

Median nerve. This was the next most affected nerve in the arm, being tender in nine cases in the operated limb and enlarged in seven of these.

TABLE 1. *Assessment of motor function in muscles examined.*

Muscle or muscle group	Total examined	Weakness: Severe (grade 0–1)	Weakness: Moderate (grade 2–3)	Weakness: Mild (grade 4–5)
Abductor digiti minimi	19	6	7	6
Digital abduction and adduction	27	10	10	7
Intrinsic action	26	18 (medial)	3 (medial)	5 (medial)
		7 (lateral)	4 (lateral)	13 (lateral)
Flexor carpi ulnaris	27	9	5	13
Flexor digitorum profundus	27	—	2 (medial)	25 (medial)
		—	—	25 (lateral)
Wrist flexion and extension	27	—	—	27
Thumb-abduction opposition	27	4	5	18
		5	4	16

FIG. 4. Closer photograph of very thickened ulnar nerve in the lower arm, showing a tear in the deep fascial sheet covering it and including some irregular blood vessels. Note surrounding adhesions (P.S.U. 655; N.P.C.-761).

The radial nerve in the groove was not thickened to palpation in a single case.

Lateral popliteal nerve. This was thickened in 23 of 40 limbs examined, and there was tenderness in 11 of these, while the posterior tibial nerve was so involved in only nine.

Clinical findings related to cutaneous nerves. The great auricular nerve was found thickened on one or both sides in five of these cases but was tender in only two.

The radial cutaneous nerve at the wrist was found thickened in a third of the cases.

The ulnar cutaneous nerve at the wrist was palpable in only two instances.

Cutaneous sensations in the operated arm. The sensation of heat (about 65°C) and of pain from light pin prick were found moderately impaired in 14 of the 27 limbs tested in the ulnar territory and in about 10 in the median territory. The same two modalities were severely affected in only eight instances for both these nerve territories. On the other hand, there was moderate impairment of superficial tactile sense (using a wisp of cotton wool) in seven of 27 limbs tested but severe impairment in 16 of 27. Thus testing for thermal and pain sense seemed to reveal more cases of moderate cutaneous sensory loss while testing for light touch appeared to bring out more cases with severe loss in these acroteric lesions.

Deep sensations. One clinical sign of value appeared to be the tenderness evoked by firm pressure in the middle of the palm. This was later correlated with extensive diffuse intrapalmer fibrosis compressing the nerves there. Rarely, in a few more severe cases, pain was elicited even on full extension of the fingers.

In about half of the cases position sense of fingers and phalanges was tested. While even small movements at the metacarpophalangeal joints were found preserved, small excursions of the distal phalanx of the fifth finger went unnoticed in three cases. All these were patients with chronic total loss of sensory and motor function of the ulnar nerve, on clinical and electromyographic examination. Subsequently, from all three patients the greater part of the ulnar nerve was excised and from two of them segments of the median nerve also.

Clinical findings in relation to muscles. ***Operated limb.*** Table 1 indicates the rough grade-wise involvement of motor function in seven muscles or muscle groups tested.

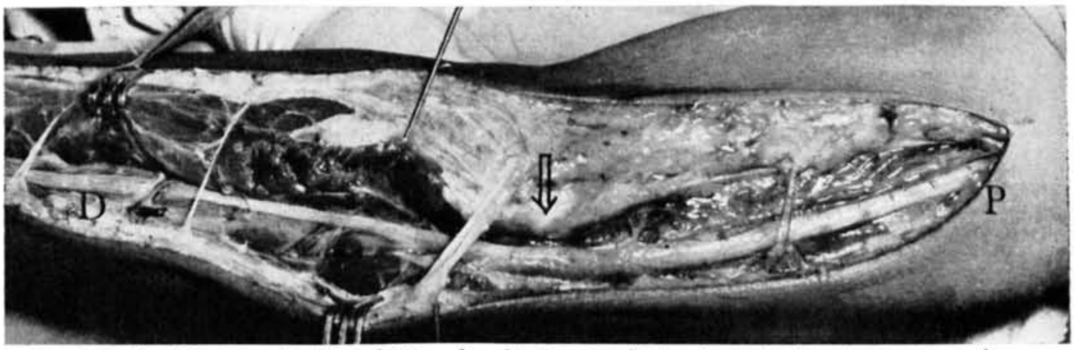


FIG. 5. (P = proximal, and D = distal) Lesser thickening of the ulnar nerve than in the previous pictures and restricted to the condylar (arrow) and supracondylar regions. Note smooth narrow course of the nerve in the forearm; some thickened cutaneous nerves (P.S.U. 266; NP-B-688).

As expected, the muscles supplied by the ulnar nerve were more often and more severely involved than those supplied by other nerves in the arm. The palmar muscles supplied by the ulnar nerve and the flexor carpi ulnaris were most frequently affected. Clinically detectable wasting accompanied severe loss of motor power, this being most noticeable in the region of the first dorsal interosseous muscle, the thenar eminence and the hypothenar eminence respectively.

Electrodiagnosis. ***Electromyography.*** As expected from the clinical features, all 28 ulnar-innervated muscles examined showed abnormalities suggestive of denervation. The number of active units during increasingly strong contraction was reduced in all records; partial interference, single oscillations or no voluntary activity being seen (Fig. 2). The shape and dimensions of the surviving units were also abnormal, polyphasics of long duration (over 10 msec.) or giant units being frequent. The latter (which were over 4,000 uv in amplitude) were reliable evidence of nerve regeneration. Spontaneous activity in the form of fibrillations was noted in only nine instances for two possible reasons: (a) a real absence reflecting a chronic slowly-progressive denervation, and (b) an apparent absence due to technical difficulties in

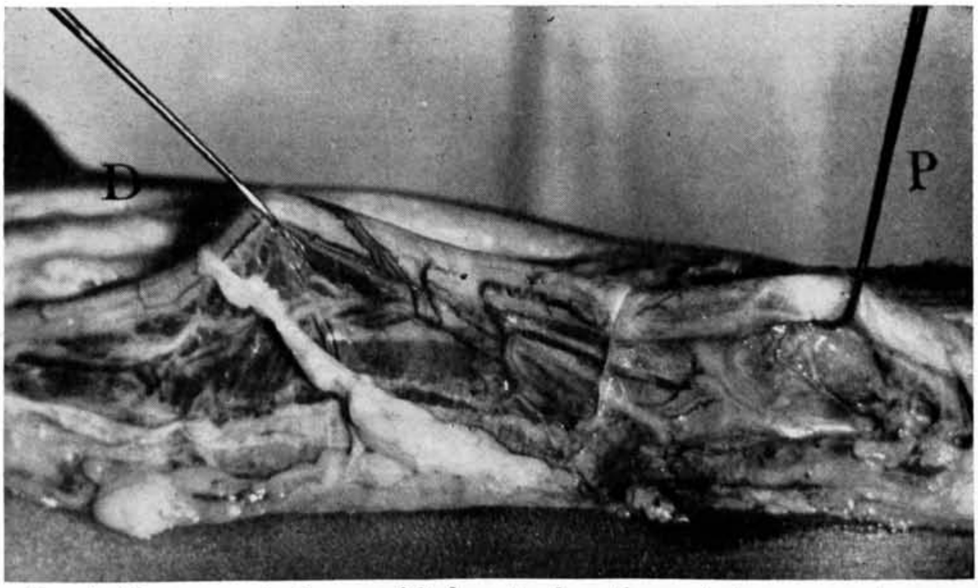


FIG. 6. Ulnar nerve in the arm lifted up to show the posterior mesoneurium providing blood supply to it, mostly in the form of arcades. (P.S.U. 645; NP-C-758).

TABLE 2. *Conduction velocity.* (a) *Ulnar nerve.*

No.	Name	Time of examination	Wrist-muscle latency (m/sec)	Elbow-muscle latency (m/sec)	Conduction velocity (m/sec)
1	P.S.A.	Preoper.	N.R.[a]	N.R.[a]	0 [b]
2	P.S.A.	Preoper.	N.R.[a]	N.R.[a]	0
3	G.B.B.	Postoper.	5.5	15.0	30 [b]
4	I.F.D.	Preoper.	N.R.[a]	N.R.[a]	0 [b]
5	I.F.D.	Preoper.	N.R.[a]	N.R.[a]	0
6	T.G.	Postoper.	5.0	15.0	24 [b]
7	T.G.	Postoper.	5.1	11.5	39.1[b]
8	V.V.J.	Preoper.	N.R.[a]	N.R.[a]	0 [b]
9	V.V.J.	Preoper.	N.R.[a]	N.R.[a]	0
10	B.H.M.	Postoper.	2.2	7.8	52.1[b]
11	J.G.M.	Postoper.	2.5	7.9	45.7[b]
12	A.R.M.	Preoper.	2.3	7.2	59.2[b]
13	A.R.M.	Postoper.	2.5	6.5	55.2[b]
14	A.R.M.	Postoper.	3.1	6.7	65.3[b]
15	A.R.M.	Preoper.	2.8	7.5	60.8
16	A.R.P.	Postoper.	5.9	12.5	41.6[b]
17	A.R.P.	Postoper.	1.7	5.0	79.1
18	C.J.P.	Preoper.	N.R.[a]	N.R.[a]	0 [b]
19	C.J.P.	Preoper.	N.R.[a]	N.R.[a]	0
20	A.G.P.	Postoper.	2.5	8.5	47.5[b]
21	A.G.P.	Postoper.	3.8	10.0	50.0[b]
22	A.G.P.	Postoper.	3.3	8.6	59.4[b]
23	K.S.R.	Preoper.	5.3	12.5	36.1[b]
24	K.S.R.	Postoper.	4.0	11.5	36.6
25	K.S.R.	Preoper.	4.2	9.5	48.1
26	K.S.R.	Postoper.	4.0	10.0	40.0
27	K.H.S.	Preoper.	4.5	12.5	46.7[b]
28	K.H.S.	Preoper.	3.1	12.0	42.7
29	B.S.	Postoper.	N.R.[a]	N.R.[a]	0 [b]
30	B.S.	Postoper.	N.R.[a]	N.R.[a]	0
31	R.M.T.	Postoper.	2.1	7.5	45.9[b]
32	R.M.T.	Postoper.	3.3	8.3	55.6
33	R.M.T.	Postoper.	1.9	5.8	68.7
34	K.J.T.	Preoper.	2.7	7.3	66.1[b]
35	K.J.T.	Postoper.	2.3	6.1	76.3[b]
36	K.J.T.	Preoper.	2.4	8.6	46.1[b]
37	K.J.T.	Postoper.	2.3	8.1	47.6[b]
38	P.S.Y.	Postoper.	3.3	7.1	61.6
39	P.S.Y.	Postoper.	3.3	10.3	41.4

[a] No response.
[b] Values obtained pre- and/or postoperatively on the *operated* side.

recording these low-amplitude potentials. No pseudo-myotonic high frequency discharges were recorded at rest in the muscles. No myopathic records were obtained in this series. Eight of the 10 tracings of the voluntary contraction of the median-innervated abductor pollicis brevis were normal, the remaining two showing abnormalities similar to those in the ulnar innervated muscles. Spontaneous fibrillations were present in three.

Nerve conduction studies. These are reported in Table 2 for each of the cases where it was done. Reduced elbow-wrist conduction velocities of ulnar nerves were present in 26 of the 39 records (normal

TABLE 2. *Conduction velocity.* (b) *Median nerve.*

No.	Name	Time of examination	Wrist-muscle latency (m/sec)	Elbow-muscle latency (m/sec)	Conduction velocity (m/sec)
1	P.S.A.	Preoper.	4.0	7.3	64.2[b]
2	P.S.A.	Preoper.	2.6	6.5	57.1
3	G.B.B.	Postoper.	3.1	7.7	59.7[b]
4	I.F.D.	Preoper.	N.R.[a]	N.R.[a]	0 [b]
5	I.F.D.	Preoper.	3.3	7.0	64.6
6	T.G.	Postoper.	3.0	7.5	51.1[b]
7	T.G.	Postoper.	2.6	11.0	26.2[b]
8	V.V.J.	Preoper.	N.R.[a]	N.R.[a]	0
9	V.V.J.	Postoper.	9.5	19.5	23.6
10	B.H.M.	Postoper.	2.6	6.8	51.4[b]
11	B.H.M.	Postoper.	2.7	6.0	64.7
12	J.G.M.	Postoper.	2.0	5.8	57.9[b]
13	J.G.M.	Postoper.	2.8	6.4	62.2[b]
14	A.R.M.	Preoper.	2.2	5.7	61.4[b]
15	A.R.M.	Postoper.	3.1	6.1	78.7[b]
16	A.R.M.	Postoper.	3.6	6.6	77.3[b]
17	A.R.M.	Preoper.	3.5	7.0	70.8
18	A.R.P.	Postoper.	2.2	6.0	67.1[b]
19	A.R.P.	Postoper.	2.4	5.5	78.7
20	C.J.P.	Preoper.	15.0	23.5	26.0[b]
21	C.J.P.	Preoper.	2.0	8.0	37.6
22	A.G.P.	Postoper.	3.5	9.5	42.6[b]
23	A.G.P.	Postoper.	3.6	8.1	53.3[b]
24	A.G.P.	Postoper.	4.2	8.2	53.0[b]
25	K.S.R.	Preoper.	3.2	6.5	62.1[b]
26	K.S.R.	Postoper.	3.7	7.3	63.8[b]
27	K.S.R.	Preoper.	3.1	6.8	51.3
28	K.S.R.	Postoper.	4.4	7.5	65.1
29	K.H.S.	Preoper.	4.2	9.6	51.8[b]
30	K.H.S.	Preoper.	3.5	9.0	45.1
31	R.M.T.	Postoper.	2.4	6.9	52.2[b]
32	R.M.T.	Postoper.	2.9	6.7	61.5[b]
33	R.M.T.	Postoper.	2.7	6.2	71.4
34	R.M.T.	Postoper.	3.4	7.0	67.2
35	K.J.T.	Preoper.	3.5	7.4	64.1[b]
36	K.J.T.	Postoper.	2.6	6.5	62.1[b]
37	K.J.T.	Preoper.	3.0	7.1	56.6[b]
38	K.J.T.	Postoper.	2.6	6.3	66.5[b]

[a] No response.
[b] Values obtained pre- and/or postoperatively on the *operated* side.

range 50-73 m/sec.). This figure includes 10 nerves which were totally nonconducting (i.e., velocities of 0 m/sec.). A wrist-muscle latency above the normal maximum of 4.5 m/sec. was noted five times, and was in each case associated with slowed elbow-wrist conduction (Fig. 3). A feature of the ulnar nerve conduction records was the polyphasic appearance of the response when the nerve was stimulated at the elbow as compared with the response obtained at the wrist. This did not appear to be due to repetitive firing. In 10 patients this feature was present in conjunction with a reduced conduction velocity in the elbow-wrist segment of the nerve. Attenuation and polyphasia of the elbow response coexisted eight times. There was no

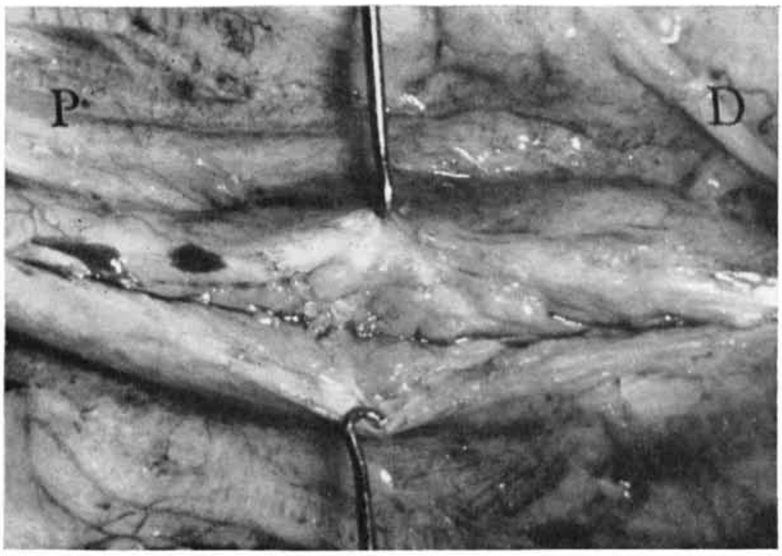


FIG. 7. Closer view of swollen totally degenerated ulnar nerve in the lower arm, with an incision along its center showing absence of funicular pattern and fibro-fatty replacement (P.S.U. 590; NP-C-664).

instance of anomalous innervation of the hand muscles in this study.

Reduced elbow-wrist velocities of median nerves were present in nine instances (including two unresponsive nerves). A wrist-muscle latency over the normal maximum of 5.0 msec. was seen twice (9.5 msec. and 15.0 msec.). Each time, as in the case of the ulnar nerve, this was associated with markedly reduced conduction velocity proximally in the elbow-wrist segment, values of 23.6 and 26.0 m/sec. being obtained.

Surgical. ***Ulnar nerve.*** In the upper arm, in 17 cases the deep fascia of the anterior medial compartment was thickened, congested and opalescent and found to compress the enlarged nerve lying underneath. It was divided longitudinally and, when adherent to the epineurium of the underlying nerve, had to be dissected off (Fig. 4). A compressing effect of the fascia on the underlying nerve was then demonstrated by the flattened nerve assuming a more rounded shape. The nerve was found to be thickened in the lower quarter of the arm in only two cases. The thickening varied from minor enlargement (Fig. 5) to gross thickening of up to 2 cm. (Fig. 4) in diameter. The swelling was generally spindle-shaped, the maximum thickness being about 5 cm. above the epicondyle. The thickening was generally restricted to the lower third of the arm but in several cases extended up to the middle of the arm and downwards into the ulnar groove and between the two heads of the flexor carpi ulnaris (Fig. 1). The thickened portion of the nerve felt firm but palpation of the nerve above the swelling generally revealed a normal, soft consistency. Leashes of blood vessels approaching the nerve through its posterior mesoneurium and forming arcades on the epineurium were best seen by lifting up the nerve in the lower arm (Fig. 6).

In eight cases an incision about 2 cm. long was made in the thickened and adherent epineurium of the area of maximal swelling to permit visualization and dissection of the nerve bundles. In three instances separate funiculi could be identified though they were pale, edematous and difficult to dissect individually even under magnification. In five cases the bundles were ill-defined, being pale, matted together and fibrous. In one of these only yellowish fatty tissue was seen along the center of the nerve (Fig. 7). Wherever justifiable, on the basis of the preoperative motor and sensory assessment as well as electrical stimulation of the nerve at the time of the operation, a long narrow wedge-shaped biopsy specimen of the

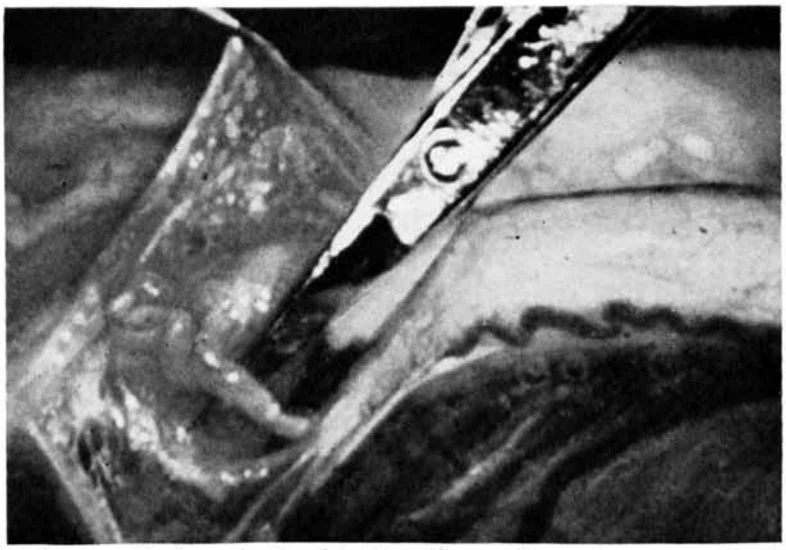


FIG. 8. Photograph through 8x objective of operating microscope showing single dissected nerve bundle on the tip of the scissors and one small vessel on the surface of the nerve. The perineurium is reflected and held up.

nerve was removed from the site of maximum thickening, including a few of the matted funiculi. This was undertaken in 14 cases. In a few early cases single nerve bundles could be dissected out, using the magnifying spectacles or the operating microscope (Fig. 8). While in these cases a fine continuous network of capillaries was observed over the funiculi, in the more advanced cases this was irregular and absent over some areas. No intraneural abscess was found in any of the nerves studied in this series. In one case an abscess was found in relation to an enlarged epitrochlear lymph node, with an associated thrombosis of a subcutaneous vein in that area and with adhesions to a thickened and tender ulnar nerve. Biopsy of the extraneural fascial sheet was also obtained.

In 11 of the 21 ulnar nerves exposed at the elbow there was visible and palpable thickening within the groove, this being an extension of the spindle-shaped swelling in the upper arm. In eight cases there was adhesion of the nerve to the inner surface of the fibro-osseous tunnel (Fig. 9). In seven instances there was downward extension of the thickening and induration into the proximal part of the forearm under the flexor carpi ulnaris for a distance of about 5 cm. Division of the overlying tendinous fibers of the origin of flexor carpi ulnaris provided a release of the nerve from the constricting effect produced by these fibers.

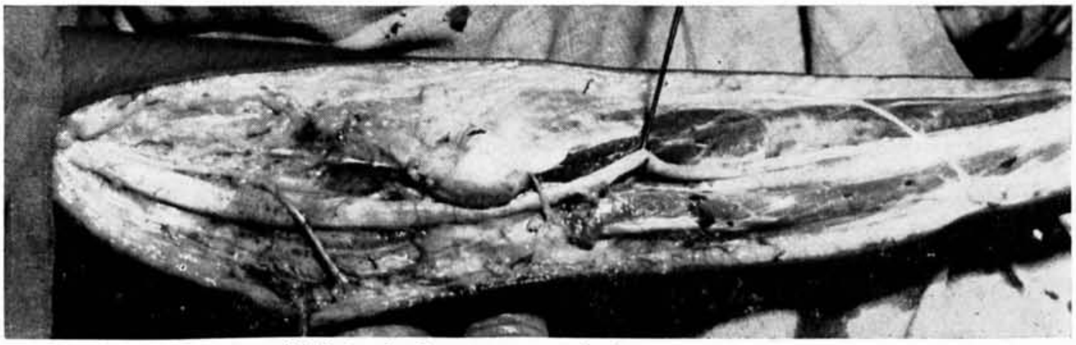


FIG. 9. Operative field in the lower arm and the upper forearm showing compression of the ulnar nerve in the epicondylar fibro-osseous tunnel, and its proximal swelling (P.S.U. 500; NP-C-527).

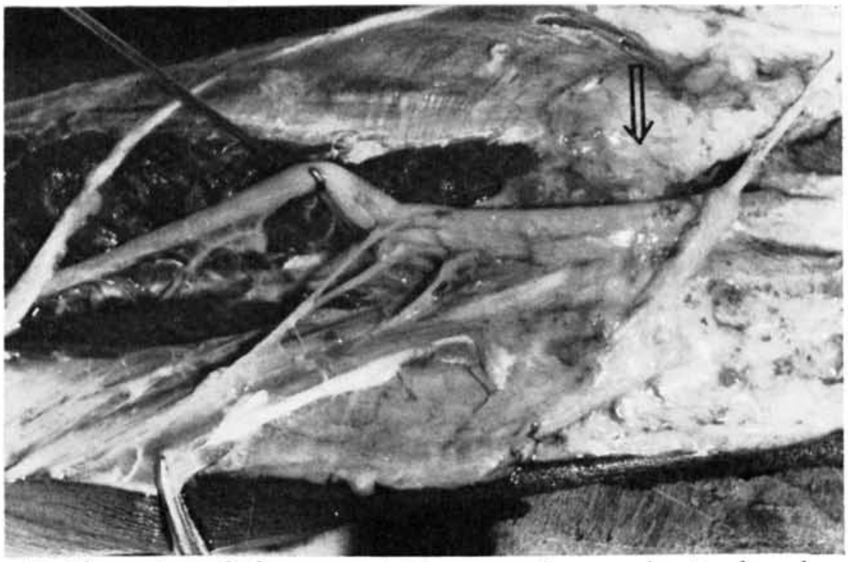


FIG. 10. Closer view of ulnar nerve in the upper forearm showing branches going to the flexor carpi ulnaris and flexor digitorum profundus muscles (arrow on medial epicondyle) (P.S.U. 505; NP-C-490).

A dumb-bell effect was sometimes produced by the thickening of the nerve above and below the fibro-osseous tunnel (Fig. 1). Biopsy of the nerve in the groove was performed in one case only.

The ulnar nerve, once it entered the deep compartment of the forearm, after passing between the two heads of the flexor carpis ulnaris, lay under cover of the overlying flexor carpi·ulnaris muscle belly. The nerve in this site was generally thin and normal looking, though firm.

The two branches of the nerve to the flexor carpi ulnaris and the single branch

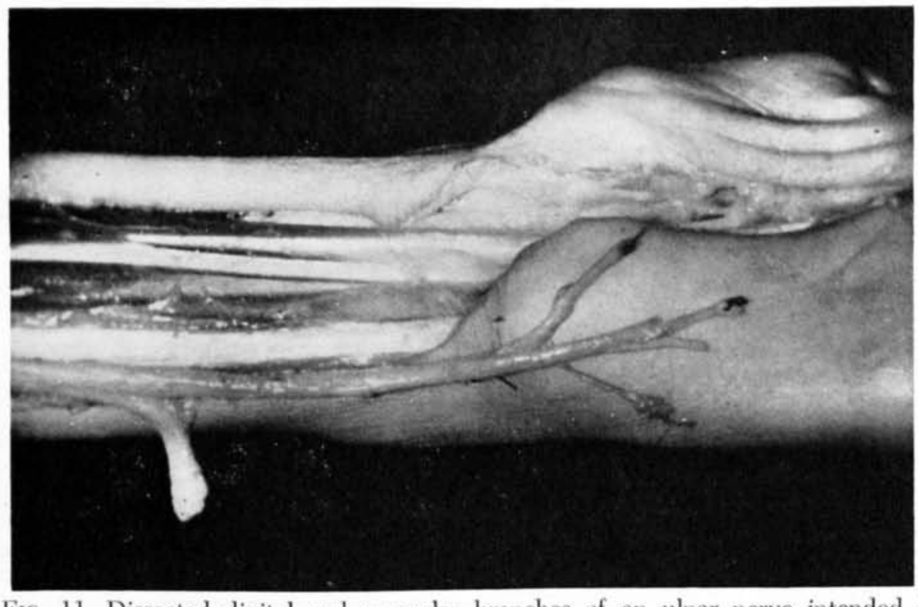


FIG. 11. Dissected digital and muscular branches of an ulnar nerve intended for total resection; (the branches are placed on the skin of the palm for the sake of clarity) note the markedly thickened dorsal cutaneous branch of the ulnar, hanging down (P.S.U. 770; NP-C-953).

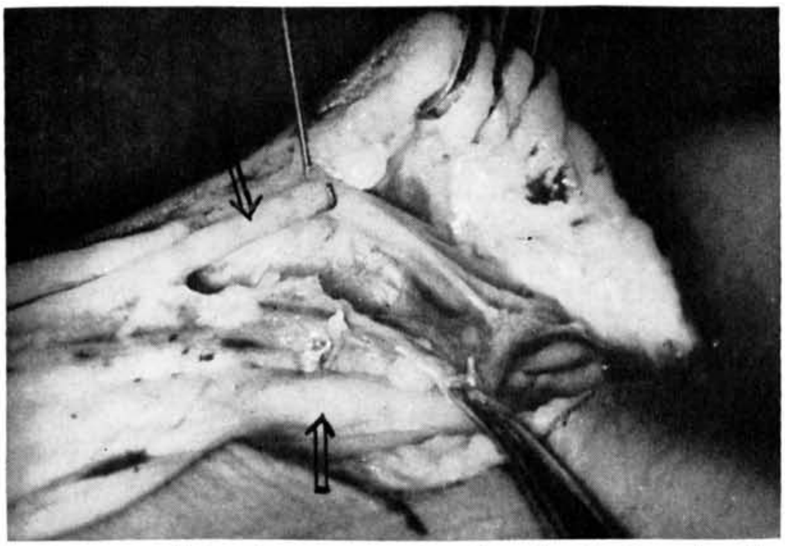


FIG. 12. Dissection of the ulnar nerve and its branches in the palm, showing the ulnar nerve hooked up (upper arrow) and its deep branch passing without adhesions through the fibrous tunnel which leads it to the deeper muscles. Note also the thickened median nerve (lower arrow) passing upwards through the now opened flexor carpal retinaculum (P.S.U. 212; NP-B-537).

to the ulnar part of the flexor digitorum profundus were traced to their entry into the muscles (Fig. 10) stimulated, and muscle biopsy taken from near the motor point. The flexor carpis ulnaris muscle was pale in 11 cases and the flexor digitorum profundus in three cases.

In the lower third of the forearm where it is not covered by the flexor carpi ulnaris belly there was some thickening of the ulnar nerve and its dorsal cutaneous branch (Fig. 11) which enters it about 7 cm. above the wrist crease. This branch which was dissected in all cases up to the point where it winds around the lower end of the ulna, was found thickened in 13 cases, the enlargement in some being so marked as to make it almost as thick as the parent nerve. A few bundles of this branch were biopsied in 11 cases.

The palmar and terminal branches of the ulnar nerve were generally normal in size. There was pallor of the hypothenar muscles in all cases where they were adequately exposed. The deep branch was seen to pass into the deep compartment of the hand through a dense fibrous tunnel (Fig. 12). This unyielding tunnel was divided with a pair of scissors as it seemed a potential source of compression. Actual adhesions between the swollen deep branch and the wall of this tunnel were noted in five cases (e.g., in Fig. 13). Biopsy of the hypothenar muscles was undertaken in two cases.

Median nerve. The fascial extraneural sheet covering the median nerve in the lower third of the forearm was thickened in six cases. The median nerve trunk and its palmar branch along its lateral aspect were carefully inspected, the former showing thickening and firmness for a distance of about 7 cm. above the wrist crease in 18 cases. The swelling was spindle-shaped and tapered to normal dimensions and consistency proximally under the flexor digitorum sublimis muscle belly and distally at the distal border of the flexor carpal retinaculum (Figs. 12, 13). Examination under magnification revealed findings similar to those in the ulnar nerve in the lower arm. A deep wedge biopsy was taken in three cases.

The nerve tapered to normal proportions at the wrist in 18 cases and some thickening persisted in three others. There was no adhesion to the tendons or to the retinaculum.

During the dissection of the trunk into its terminal thenar and digital branches, difficulty was noted in identification and separa-

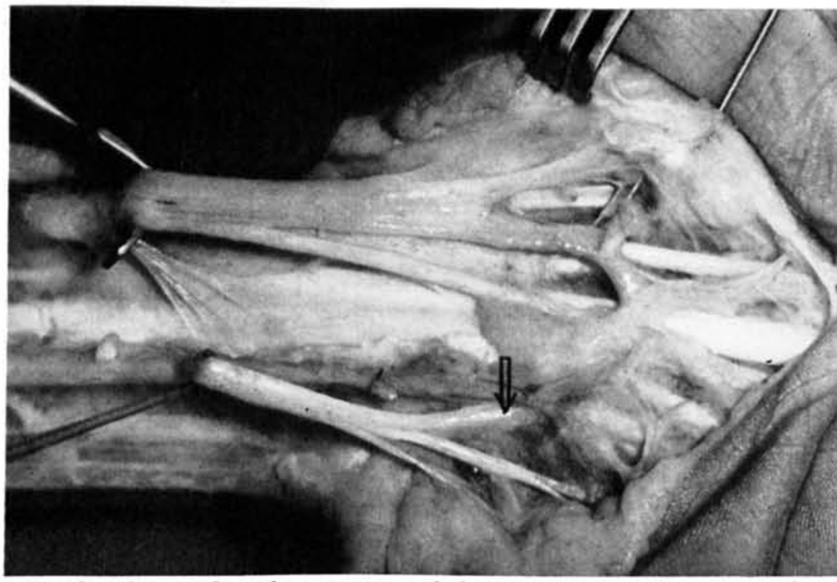


FIG. 13. The ulnar and median nerves and their branches in the wrist and the palm showing adhesions around the deep branch (arrow.) The thickened median nerve is released from the flexor carpal retinaculum. Note adhesions around digital branches and vessels in distal part of palm (P.S.U. 500; NP-C-527).

tion of the branches in 11 cases due to adhesions of the nerve and its branches to the surrounding palmar connective tissue (Fig. 13). Sharp dissection was required to free them.

Thenar and lumbrical muscle atrophy was noted and biopsy was obtained in five cases.

Anomalies. In three cases a thin triangular muscle was noted in the lower forearm. It had its apex in the region of the pisiform bone and the lower end of the hypothenar muscles, and it fanned out to its insertion into the deep fascia superficial to the palmaris longus tendon in the lower quarter of the forearm (Fig. 14).

In one patient, where both arms were explored, an anomaly was found in relation to the median nerve. On one side a large aberrant branch descended independently from the forearm into the hand to supply the digital branches to the middle and fourth fingers. On the other side, the aberrant branch arose in the forearm and fused with the main trunk above the wrist.

Long excision biopsies. The nerve was excised in continuity from below the elbow to the palm in two patients with total ulnar paralysis of four years' duration. In one case of total paralysis of one and a half years' duration the ulnar nerve was excised from the level of the anterior axillary fold to the palm (Fig. 11). In two instances the median nerve was also excised from below the level of its last motor branch in the forearm to its digital and thenar branches. The digital branches in the fingers were also biopsied in one case through separate incisions in the fingers.

Follow-up. Twenty of the 22 patients were seen at least once, between six months and two and a half years postoperatively. There was no instance of operative trauma to motor or sensory nerves. Some degree of relief of nerve pain was present in 16 of the 25 limbs, usually occurring one to two weeks after operation. The pain either ceased completely, or was less frequent and less severe. A subjective feeling of increase in muscle strength was reported by three patients, who were thereafter able to return to their old occupations of tailor, barber and compounder respectively. One patient said he had not burnt his fingers since he was operated, suggesting improvement in sensory function.

Those patients who did not benefit from the extensive decompression were generally dimorphous or lepromatous cases with a history of several reactions.

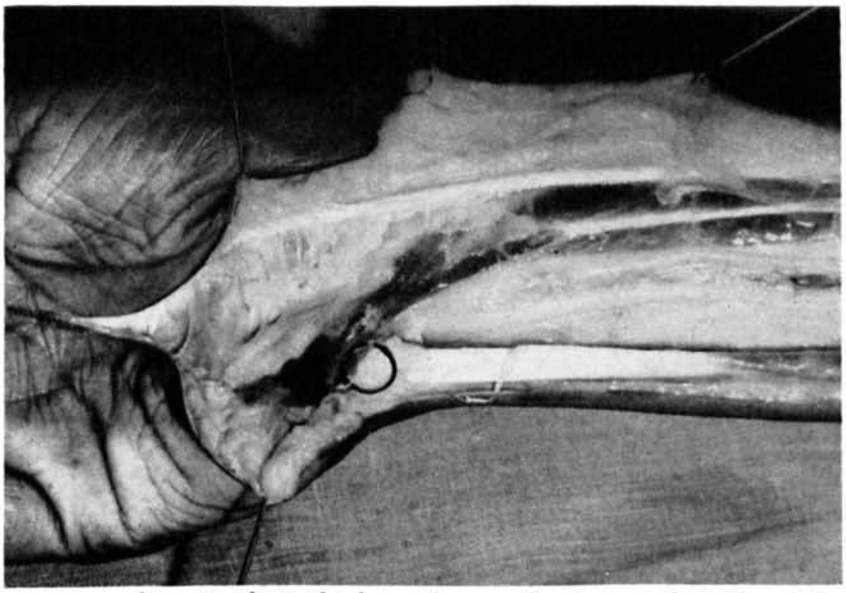


FIG. 14. Anomalous muscle in the lower forearm fanning out from the pisiform region to the tendon of the palmaris longus. Circle is placed around site of pisiform bone where the terminal of the tendon of the flexor carpi ulnaris is seen (P.S.U. 538; NP-C-577).

DISCUSSION

This is perhaps the first report of a comprehensive multidisciplinary investigation of nerves in leprosy, combining an extensive operative exploration with preceding clinical and electrodiagnostic observations, succeeding histologic and follow-up studies.

This study was conducted jointly by both investigating officers (plastic surgeon and neuropathologist) and the Assistant Research Officer at all stages of preoperative investigations, operative procedures and postoperatively. Biopsies were often taken by the neuropathologist himself at the operation at which he was the first assistant.

Clinical features. A dual purpose was served by these extensive nerve exploration studies, both investigative and therapeutic. While the main objective was to see the nerves *in extenso* and to examine their surroundings, relief of pain was also a factor indicating such exploration. In fact, as the study progressed, patients volunteered for explorations on a second limb when relief from pain had been obtained in the first limb. Postoperative assessment also showed no additional sensory or motor deficit. No objective clinical improvement was detected in these functions.

The above objectives naturally demanded a cautious selection of relatively advanced cases with established polyneuritic leprosy, especially in the earlier phases of the study. With the confidence gained from experience, cases of relatively earlier stages of polyneuritis were accepted for exploration and, with the help of magnification, bundle biopsies of nerves were also undertaken.

The preponderance of dimorphous cases in our material probably reflects the most prevalent type of leprosy in this part of India. The greater immunologic instability of patients with dimorphous leprosy could also perhaps account for the greater incidence of nerve pain amongst them and their higher incidence in our material.

The retention of deep sensation of various types, for example, deep pressure and joint sense, was noteworthy even in the presence of gross cutaneous sensory loss, muscle weakness and wasting.

Ulnar nerve involvement was found to be more frequent and more severe than that of the median nerve, as has been generally found to be the case.

Electrodiagnostic features. The general pattern of the electrodiagnostic abnormalities appeared to relate more truly to the severity of the nerve involvement than to the type of leprosy.

The ulnar innervated muscles of the hand and forearm were, as expected, more frequently and more severely, affected as determined by electromyography, than those supplied by the median nerve. The presence of giant units would strongly suggest reinnervation, by collateral sprouting of denervated muscle fibers with a consequent increase in the size of the motor unit. Such sprouting has been demonstrated by Dastur ([7]) in leprosy in the flexor carpi ulnaris muscle, and recently during experimental reinnervation by Karpati and Engel ([12]).

Although myositis, either granulomatous or nonspecific, was noted histologically ([5]) no electromyographic records were obtained to suggest primary muscle damage, in contrast to what Magora *et al.* ([15]) have demonstrated.

The nerve conduction studies confirm the more extensive involvement of the ulnar nerve, abnormalities such as reduced conduction velocity, polyphasia and attenuation of the elbow response being more frequently obtained than in the median nerve.

The fact that in both nerves prolonged wrist-muscle latency was always associated with reduced elbow-wrist conduction velocity, suggests that at least in patients with well developed neuritis there is no evidence of selective compression or inflammatory fibrosis at the wrist. The converse finding of a normal distal latency with slowed conduction in the forearm in as many as ten instances is consistent with the histologic and gross observation of maximal involvement of the ulnar nerve behind and proximal to the medial epicondyle, and of the median nerve in the lower one-fourth of the forearm proximal to the flexor retinaculum. The nerve stimulation findings in the median nerve in leprosy would then contrast with the report of Thomas *et al.* ([18]) in a large number of median nerves in the carpal tunnel syndrome. They found delay in distal conduction (in wrist-muscle conduction) in 67 per cent of symptomatic cases while elbow-wrist slowing of less than 49 m/sec. was found in only 17 per cent. These authors emphasize that the distal latency was prolonged out of proportion to the proximal slowing.

It is significant that although all patients had symptoms and signs of neural involvement, impairment of the conduction velocity was not invariably found even with clearly neuropathic electromyographic records in the reference muscle. It would appear, therefore, that the electrodiagnostic application of nerve conduction velocities in leprous neuritis should be limited to the relatively advanced cases. This should be compared with the observations in other chronic polyneuropathies, e.g., peroneal muscular atrophy and related syndromes ([10, 22]) where impairment of conduction in the long peripheral nerves is marked in symptomatic cases, and even some mildly affected or asymptomatic cases of Charcot Marie-Tooth disease. This will be discussed further in a second paper ([5]).

In performing these studies care was taken to reduce skin resistance (by use of alcohol or acetone) and to adjust the stimulus strength till no further increase occurred in the size of the response. The latter precaution is of special importance in leprosy since the effectiveness of a stimulus applied to an unevenly fibrosed, edematous and infiltrated nerve cannot be assured, and high-tissue resistance could result in a shift of the effective point of stimulation. Both factors would contribute significantly to errors in computation of conduction velocity. In this study, no tissue or room temperature measurements were made. It is possible, however, that no significant errors were introduced by this omission in view of the warm, equable, moist climate of Bombay.

These observations of the ulnar and median nerves in leprosy accord generally with earlier studies on the facial nerve in this disease ([8]). In individual cases, nerve damage was more accurately detected by electromyographic studies than by the measurement of nerve conduction velocities.

Surgical features. Contrary to our earlier fears, a tourniquet time of up to one hour and forty minutes did not produce any adverse effects even on these damaged nerves.

Similarly, even the extensive dissection and exposure of the nerves from the upper arm to the distal palm did not lead to any adverse effects. This was probably due not only to the delicate handling of the nerves but also to the careful preservation of the blood supply. This supply is derived chiefly from the bed of the nerve through the posterior "mesoneurium," as described by Smith ([17]), and was invariably left intact. Keeping this important anatomic feature in mind, we did not carry out any anterior transposition of the ulnar nerve. We believe this would devascularize an already affected part of the nerve and further jeopardize its function.

While we confirmed prior clinical observations of maximal thickening of the ulnar nerve above the epitrochlear groove in the upper arm, it was observed that this thickening extended downwards into the groove and up to the passage of the nerve between the two heads of the flexor carpi ulnaris. While there was slight thickening of the ulnar nerve in the lower forearm in some cases, its dorsal cutaneous branch was frequently thickened, at times reaching a proportion larger than that of the parent trunk.

The median nerve also showed maximal thickening in the lower forearm above the flexor carpal retinaculum. It invariably narrowed to normal size and consistency as it reached the lower border of the flexor digitorum sublimis. Its lower end, under the flexor carpal retinaculum, was only slightly thickened and not adherent to it.

Slitting and dissection of nerve sheaths demonstrated, even to the naked eye, and more so under the operating microscope, the density of the adhesions of the various sheaths and of the nerve bundles to one another. This precluded adequate dissection of the individual funiculi, except in the few early cases, and also demonstrated the futility of attempting to decompress the nerve by multiple incisions or by desheathing ([3]). In fact, the latter procedure, often going under the misnomer of neurolysis, should be recognized as being physiologically unsound, technically difficult, and damaging.

Extraneural factors. Secondary extraneural factors, compressive or otherwise, acting upon primarily diseased leprous nerves and producing further damage at certain sites, have been enumerated earlier ([7]). Some of these, and other additional sites of nerve damage by extraneural mechanisms, were observed during the present study.

Wherever there were obvious pathologic changes in the nerve there generally were associated perineural adhesions and inflammatory changes. These changes were most noticeable in relation to the ulnar nerve in the arm. The deep fascia was thickened, congested and adherent to the medial aspect of the nerve, and appeared to constitute a form of compression of the nerve. The most significant site of compression of the swollen ulnar nerve was found to be the unyielding fibro-osseous tunnel behind the medial epicondyle. An additional site of compression appeared to be the tendinous arch formed by the two heads of origin of the flexor carpi ulnaris. In the forearm and wrist the ulnar nerve did not seem to suffer any compression. In the palm, the fibrous tunnel through which its deep palmar branch passes into the deep compartment, was a frequent source of compression when this branch was swollen.

The only significant site of compression of the median nerve was in the unyielding flexor carpal tunnel under the carpal retinaculum.

The digital and muscular branches in the palm, of both the median and ulnar nerves, were frequently enmeshed in, and adherent to, the inflammed fibro-fatty tissue of this region.

Any operative procedure aimed at release of the nerves must take into account all these sites and the mechanisms of compression of these nerves.

SUMMARY

A comprehensive clinical, electrodiagnostic, surgical and gross morphologic investigation of peripheral nerves in 22 cases of polyneuritic leprosy is presented.

Detailed preoperative clinical evaluation of the sensory and motor status of these patients revealed findings consistent with leprosy and polyneuritis of long duration.

Preoperative electrodiagnostic studies on 17 patients, including 41 ulnar and 41 median nerves, revealed more severe slowing of conduction velocity in the former than in the latter. Electromyography was a more sensitive indicator of nerve damage in individual cases than nerve conduction velocity studies.

An extensive exploration of the ulnar and median nerves through the palm, the forearm, and arm was carried out in 25 limbs. This revealed gross morphologic changes in both these nerves to be more extensive than observed clinically. These changes were at the classical sites as well as at less recognized positions, and were related to the operation of extraneural factors of compression. These included thickened deep fascia, fibro-osseous tunnels and palmar tissues.

Relief of nerve pain was achieved in most cases, but objective improvement was not observed after operation, on clinical or on limited electromyographic examination.

Correlative histopathologic findings and pathogenesis will be considered in a second paper ([5]).

RESUMEN

Se presenta un estudio completo de tipo clínico, electrodiagnóstico, quirúrgico y morfológico macroscópico de los nervios periféricos en 22 casos de lepra polineurítica.

La evaluación clínica detallada preoperatoria del estado sensorial y motor de estos pacientes reveló hallazgos consistentes con lepra y polineuritis de larga evolución.

Los estudios electrodiagnósticos preoperatorios en 17 pacientes, que incluyeron 41 nervios ulnares y 41 nervios medianos, demostraron que el retardo de la velocidad de conducción es más severo en el primero que en el segundo. La electromiografía resultó ser un indicador más sensible de lesión nerviosa en los casos individuales, que los estudios de velocidad de conducción nerviosa.

Se llevó a cabo una exploración extensiva de los nervios ulnar y mediano a través de la palma, el antebrazo y brazo en 25 extremidades. Esta exploración reveló que las alteraciones macroscópicas en ambos nervios son más extensas que lo observado clínicamente. Estas alteraciones se encontraron en las localizaciones clásicas, como asimismo en posiciones menos conocidas, y estaban relacionadas con la presencia de factores de compresión extraneurales. Estos incluían engrosamiento de aponeurosis profundas, túneles fibroóseos y tejidos palmares.

En la mayor parte de los casos se consiguió alivio del dolor del nervio, pero no se observó mejoría objectiva después de la operación, al examen clínico o electromiográfico parcial.

En un segundo trabajo consideraremos los hallazgos histopatológicos correlativos y la patogénesis. (5).

RÉSUMÉ

Les résultats d'une recherche portant sur les caractéristiques cliniques, electrodiagnostiques, chirurgicales et morphologiques des nerfs périphériques, dans 22 cas de lèpre polynévretique sont présenté.

Une évaluation clinique pré-opératoire détaillée de l'état moteur et sensoriel de ces malades a fourni des résultats qui permettent d'admettre un diagnostic de lèpre et de polynévrite de longue durée.

Les études électrodiagnostiques pré-opératoires menées sur 17 malades, et portant sur 41 nerfs cubitaux et 41 nerfs médians, ont révélées que la vitesse de conduction présentait un ralentissement plus grave au niveau du nerf cubital qu'au niveau du nerf médian. L'électro-myographie a constitué un indicateur plus sensible que les résultats des études de vitesse de conduction, pour mettre en évidence la lésion nerveuse dans des cas individuels.

Une exploration approfondie des nerfs cubitaux et médians, au niveau de la paume, de l'avant-bras et du bras, a été pratiquée sur 25 membres. Cette exploration a permis de montrer que les modifications morphologiques au niveau de ces deux nerfs étaient plus étendues qu'elles n'apparaissent cliniquement. Ces modifications étaient situées tant aux endroits classiques qu'en des sites moins généralement reconnus; elles étaient en relation avec la présence de facteurs de compression extra-neuraux. Ces facteurs comprenaient entre autre l'épaississement d'aponévroses profondes, de tunnels fibro-osseux et de tissus palmaires.

Un soulagement de la douleur nerveuse a été obtenue dans la plupart des cas. Néanmoins, une amélioration objective n'a pas été observée après l'opération, pour autant que l'on puisse en juger par l'examen clinique ou par une exploration electromyographique limitée.

Les observations histo-pathologiques correspondantes, de même que le problème de la pathogénèse seront considérés dans un second article.

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