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CLINICAL NOTES

Editor's Note: In an effort to increase the utility of the JOURNAL in continuing medical education, it has been suggested that a new feature of the JOURNAL be added on a trial basis to the Editorial Section—Clinical Notes. In this section, we welcome contributions dealing with practical problems in leprosy work. Submissions to this section will undergo minimal editorial changes and may well contain controversial points. Letters to the Editor pointing out other viewpoints are welcome.—RCH

Cervical Perineurial Cyst, A Red Herring (Case Report)

Intraspinal cysts may be located outside or inside the dura mater. They are detected incidentally during myelographic or spinal CT studies and are usually benign. Intradural perineurial cysts are seen in the sacral area and, rarely, in the cervical or thoracic regions.[1-3] Perineurial cysts mostly arise at the site of the posterior root ganglion, and their wall is composed of neural tissue. These cysts are differentiated from meningeal cysts/diverticula by their location along the nerve root, delayed filling with the contrast medium, and microscopic features.[2] Occasionally, these cysts may become symptomatic and cause compressive myeloradiculopathy. We present a young man who had progressive wasting of the right hand caused by neuritic leprosy which, in the absence of any hypoesthetic skin lesion, was attributed to compression of the cervical roots by the perineurial cysts in their early course. Prolonged F-wave latency attributable to motor radicle involvement and nerve conduction studies correlated well with the clinico-radiological diagnosis of entrapment radiculopathy by the perineurial cysts. However, antileprosy drugs were started on the basis of mild ulnar nerve thickening across the medial epicondyle, and surgery was postponed. The clinical course and nerve conduction studies 6 months later confirmed neuritic leprosy as the underlying disease.

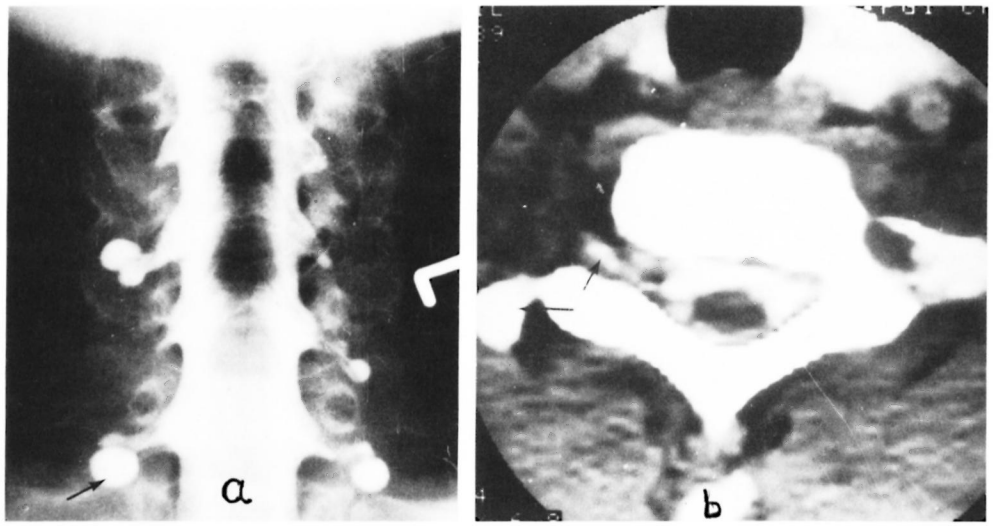
CASE REPORT

A 29-year-old, nondiabetic male (a tailor by profession) presented with a history of progressive wasting of the right hand for 1 year. He denied fasciculations, sensory loss, any febrile episodes, and any history of trauma. General and systemic examination was normal. There was a partial clawing deformity of the right hand. Hypothenar and the interossei muscles were more wasted compared to the thenar group of muscles. The power in the right dorsal and palmar interossei, lumbricals, abductor digiti minimi, and adductor pollicis brevis muscles was reduced by 30% to 50%. A subjective sensory impairment of pain and temperature was observed over the dorsal and palmar surfaces of the medial two fingers and the right forearm. However, there was no objective sensory loss or hypoesthetic hypopigmented skin patch. All deep and superficial reflexes were normal. The right ulnar nerve across the medial epicondyle was minimally thickened compared to the left side. Hemogram, blood biochemistry, plain x-ray of the cervical spine, and cerebrospinal fluid were normal. In view of progressive asymmetric wasting of the right hand, an iohexol lumbar myelogram was done which showed free flow of dye across the cervical region. There was filling of multiple bilateral cysts placed across the 5th, 6th, 7th, and 8th cervical roots (The Figure). The contrast medium could be dislodged partially by positioning the patient. The largest

[1] Holt, S. and Yates, P. O. Cervical nerve root "cysts." Brain **87** (1964) 481–490.

[2] Tarlov, I. M. Spinal perineurial and meningeal cysts. J. Neurol. Neurosurg. Psychiat. **33** (1970) 833–843.

[3] Wilkins, R. H. Intraspinal cysts. In: *Text Book of Neurosurgery.* Wilkins, R. H. and Rengachary, S. S., eds. New York: McGraw-Hill Book Company, 1985, pp. 2061–2063.



THE FIGURE. Multiple cervical perineurial cysts seen on **a)** iohexol myelogram and **b)** iohexol spinal CT scan with largest cyst over C8 radicle (large arrow).

cyst (about 15 mm in diameter) was seen in relation to the right 7th cervical foramen transversarium. Its placement outside the intervertebral foramen was confirmed in an iohexol spinal CT x-ray (The Figure).

Right ulnar motor nerve conduction studies revealed a mild increase in distal latency of 5 milliseconds (ms), normal nerve conduction velocity of 50 m/sec, and prolonged F-wave latency of 44.4 ms, suggesting a proximal lesion in the nerve trunk or radicle. Ulnar sensory nerve conduction showed a mild abnormality in the form of reduced amplitude of the sensory action potential (14 μv) as compared to 21 μv in the opposite ulnar nerve. Skin biopsies from the forehead, right ear lobule, and medial dorsum of the right hand did not reveal bacteriological or morphological evidence of leprosy. He was started on a daily dose of 100 mg of dapsone and 600 mg of rifampin. Six months later he was observed to have a well-defined, 10-cm diameter anesthetic patch over the dorsum of the right hand. However, the wasting was static although subjectively he felt improvement in hand grip. A follow-up right ulnar motor conduction revealed a distal latency of 4.8 ms; nerve conduction velocities (NCV) of 38 m/sec in the forearm, 35 m/sec in the arm, and 28.5 m/sec across the elbow. Results of a right ulnar F-wave wrist latency and an EMG study of the right abductor digiti minimi were the same as before.

DISCUSSION

This patient presented with a wasted hand. A myelogram and a spinal CT revealed multiple intraspinal cystic swellings in the cervical region. Their communication with the spinal subarachnoid space and extradural location was confirmed. The wasting of the intrinsic muscles of the right hand could have been attributed to compression of a mixed trunk of C8 motosensory radicles by the largest cyst placed near the 7th right foramen transversarium. This was substantiated by ulnar motor nerve conduction studies which showed gross asymmetry of F-wave latency and a marginal increase of distal latency suggestive of motor radicle involvement.[4] Since perineurial cysts are rarely symptomatic and the ulnar nerve was slightly thickened, the patient was given the benefit of the doubt of leprous mononeuropathy. Follow-up observations of the anesthetic patch over the right dorsum of the hand, the fall of motor nerve conduction velocities, and the minimal improvement

[4] Brown, W. F., Ferguson, G. G., Jones, M. W. and Yates, S. K. The location of conduction abnormalities in human entrapment neuropathies. Can. J. Neurol. Sci. 3 (1976) 111–122.

in the motor deficit leaves little doubt regarding the diagnosis of leprosy. Repeat skin biopsies from the forehead, ear lobule, and right hand anesthetic patch were negative for leprosy bacilli. Our patient probably had paucibacillary (tuberculoid) leprosy, a form of leprosy in which skin biopsies are usually negative for leprosy bacilli. Retrospectively, the increase in the F-wave latency during the first examination can be explained on the basis of ulnar nerve involvement in the right arm above the medial epicondyle where it lies superficially and is prone to leprosy affection.

SUMMARY

Leprous mononeuropathy may present without demonstrable maculoanesthetical patches. A case is described in whom wasting of the right hand was initially attributed to C8 T1 entrapment radiculopathy caused by cervical perineurial cysts. On the basis of the benign nature of the perineurial cysts and questionable right ulnar nerve thickening, the patient was given a trial of antileprosy drugs. A follow-up clinico-electrophysiological evaluation favored the diagnosis of leprotic ulnar mononeuropathy.

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Granuloma Multiforme in India

Leiker[1] in 1964 coined the term granuloma multiforma, "a disease resembling leprosy," to describe an entity with "a granulomatous structure and variation in clinical aspect." He described the profile of 148 such patients found in a survey in northern Nigeria. Soon after, Browne[2] reported cases from eastern Nigeria and Meyers, *et al.*,[3] from the Congo. Most patients were found in leprosy settlements, where they were mistakenly undergoing treatment for leprosy.

The etiology[1, 2] of this disease is yet to be elucidated. Airborne agents from roofing and headloads, as well as parasites such as microfilariae, have been seriously considered.

We report on two patients with granuloma multiforme who presented at a small leprosy center in South India. The clinicohistological picture seen was identical to that described by Leiker.

Case 1. A 75-year-old female presented with skin lesions associated with a mild burning sensation of 1-year duration.

Examination revealed 8 to 10 lesions distributed over the arms, back and forearms. They ranged in size from ½ cm to 10 cm. All of her lesions were rimmed by an erythematous or flesh-colored papular border. The larger lesions showed mild atrophy and hypopigmentation of the central areas; whereas this feature was not marked in the smaller lesions. There was no sensory loss or peripheral nerve thickening. Over the next 4 months, she was observed to develop new lesions on the flanks, forearms and dorsum of the hand. The patient was then lost to follow up.

Further investigation gave a negative result for skin smear for acid-fast bacilli (AFB). Her ESR was 40 mm in the first hour, and she had a two-hour postprandial blood sugar of 100 mg%.

A skin biopsy from the edge of a large lesion showed dense perivascular and peri-

[1] Leiker, D. L., Kok, S. M. and Spaas, J. A. J. Granuloma multiforme, a new skin disease resembling leprosy. Int. J. Lepr. **32** (1964) 368–376.

[2] Browne, S. G. Granuloma multiforme in eastern Nigeria. Int. J. Lepr. **34** (1966) 27–29.

[3] Meyers, W. M., Connor, D. H. and Shannon, R. Histological characteristics of granuloma multiforme. Int. J. Lepr. **38** (1970) 241–249.

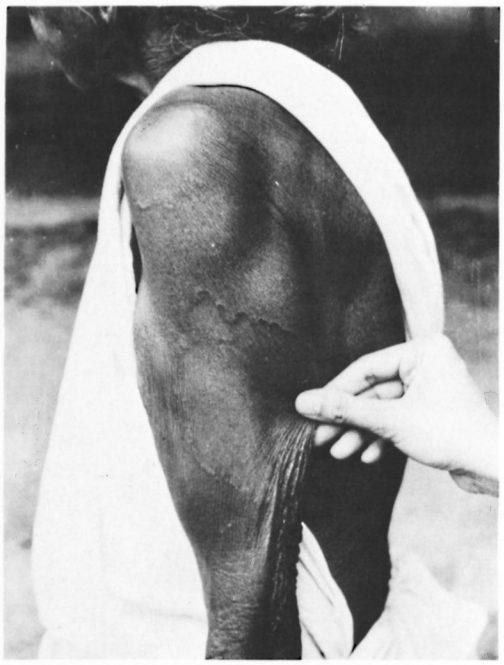

FIG. 1. Large, well-defined lesions with raised borders on the arm and shoulder; central area shows hypopigmentation.

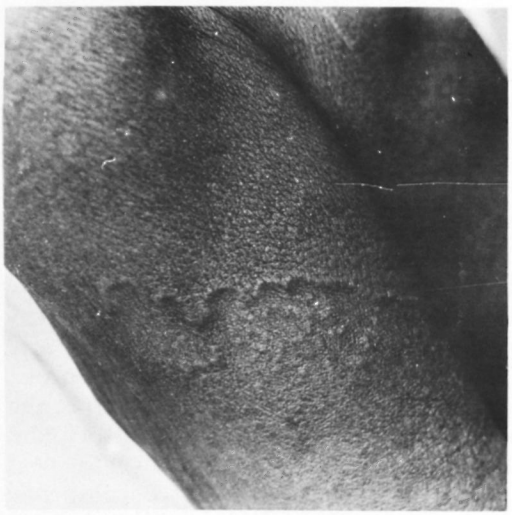

FIG. 2. Close up of raised border of Fig. 1 lesion showing closely set papules.

adnexal aggregates of lymphocytes. There were foci of collagen degeneration surrounded with an inflammatory reaction composed of many histiocytes and multinucleated giant cells. A dense perineural collection of lymphocytes was seen in the mid-dermis and in one prominent nerve in the deeper dermis, with one or two lymphocytes within the nerve. No AFB were present.

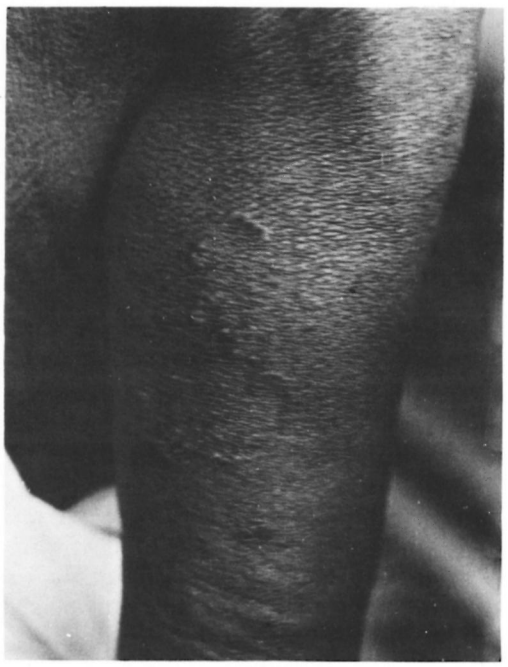

FIG. 3. Circinate lesions on back of the arm.

Case 2. This 60-year-old female was seen 3 months later. She presented with lesions of 2 years' duration which were itchy initially and extended slowly. She gave no history of contact with leprosy. An examination showed about eight plaques situated mainly on the trunk and upper limbs. The lesions were large, from 5 cm to 15 cm in diameter, irregular in shape, and rimmed by a papular border. The bigger lesions showed central clearing; the others were slightly raised. Again, there was no sensory loss or peripheral nerve thickening.

During subsequent visits, this patient was seen to develop new lesions. A single new lesion on the flank was carefully observed. It began as a papule, and then extended into a plaque with a finely wrinkled surface. A month later it showed central clearing. A few weeks later, the lesions were flat except for the papular margin; the central area then showed mild hypopigmentation. A smear for AFB from the patches was negative. Her ESR was 30 mm/hr and her two-hour postprandial blood sugar was 100 mg%.

Histopathology of the skin from the edge of a plaque showed localized areas of degeneration of collagen in the superficial dermis, some fairly well defined, others relatively ill defined. These areas were associated with an infiltrate of lymphocytes and histiocytes, some of which were epithelioid. One discrete epithelioid granuloma was present, and several multinucleate macrophages were seen. There was focal necrosis of the dermis with considerable disarray of collagen in these areas. A fairly dense perivascular infiltrate was seen, but no vessel-wall thickening. One of the two dermal nerves seen contained a few lymphocytes. AFB and detectable mucin were absent.

Discussion. The clinical features and histology of the lesions described in these two patients are classical for granuloma multiforme. The well-defined nature of the lesions with raised borders and central hypopigmentation closely simulates borderline tuberculoid leprosy. The itching and burning sensation associated with the onset of the lesions mimics the paresthesia sometimes associated with leprosy lesions. Differentiation is based on the absence of objective sensory loss, normal size of peripheral nerves, negative skin smears, and a histological picture of necrobiosis.

Granuloma annulare ties in with leprosy in the differential diagnosis of granuloma multiforme. Unlike granuloma annulare which affects a younger age group, granuloma multiforme is more commonly seen after 40 years of age and is thought to have a female predilection.[2] Again, the large lesions distributed on the back and arms in granuloma multiforme are in contrast to the smaller rings of granuloma annulare seen mainly over the distal parts of the limbs.

Both entities are classified as necrobiotic disorders. Granuloma multiforme differs in the dense perivascular infiltrate seen, the absence of mucin, the absence of palisading of cells and, particularly, the prominent multinucleate giant cells in the infiltrate. Of special interest are the intraneural lymphocytes seen in both of these cases. This feature has not been reported earlier.

Necrobiosis lipoidica shares many histological features with granuloma multiforme, but does not cause confusion on clinical grounds. The distinguishing features are the presence of fibrosis intermingled with the necrosis, vessel-wall changes, and fat deposition. The clinical differentiation is by the yellowish color of the plaques, telangiectasia, and its predilection for the lower limbs.

Tinea corporis is excluded by the absence of scaling and a negative scraping for fungus.

In summary, these two case reports from South India are of patients who presented with granuloma multiforme, an entity thought to be confined to Africa and Indonesia.[4] Interest lies in its possible misdiagnosis as leprosy.

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[4] Leiker, D. L. Differential diagnosis. In: *Leprosy.* Hastings, R. C., ed. London: Churchill Livingstone, 1985, pp. 185–186.