Cranial neuropathy in patients with leprosy

Leprosy is a common cause of peripheral neuropathy in tropical countries including India (1) and neuropathy is observed across the spectrum of leprosy from polar tuberculoid (TT), borderline (BB, BB, BL) to polar lepromatous (LL) types. Diagnosis is not difficult when cutaneous lesions are present, but in the ‘pure or primary neuritic’ form where skin lesions are absent, a high index of suspicion is required to look for thickening of nerves and proceed with confirmatory investigations of nerve conduction and biopsy. The delineation of this pure polyneuritic form as a distinct clinical entity and inclusion in the classification of leprosy is a contribution of Indian leprologists (2). In some of these patients, skin lesions may appear while on treatment (3). As a result of implementation of the intensive National Leprosy Eradication Program in India, with early case detection and treatment, neurologists may be confronted with partially treated cases with resolution of skin lesions but persisting neuropathy (history of treatment is often not revealed by the patient due to the underlying stigma) or pure neuritic leprosy.

Mononeuritis, mononeuritis multiplex or symmetrical polyneuropathy forms are seen in leprosy. Cranial nerve paralysis generally occurs along with any of the above three types of peripheral neuropathy and rarely may also be an isolated feature. The reported frequency varies from 10 to 22% (4). Facial, trigeminal and olfactory nerves are the commonest nerves to be affected. In this issue, Kumar and colleagues (5) in the setting of neurology department in a tertiary health care center, observed that 9 of 51 (18%) patients admitted during a period of 8 years (1995 to 2003) with leprous neuropathy had involvement of cranial nerves. They had looked for involvement of only 3rd to 12th cranial nerves. Facial (5 patients) and trigeminal (4 patients) nerves were the commonest to be involved, and oculomotor, auditory, glossopharyngeal, vagus, spinal accessory and hypoglossal nerves were affected in one patient each. There were 4 patients with multiple cranial nerve paralysis. Lepromatous leprosy was the common type (5 patients), and there was a single patient with pure neuritic leprosy.

Thickening of peripheral nerves and cutaneous branches, particularly the greater auricular nerve (GAN), is a diagnostic marker of leprosy. Dharmendra (6) cautioned that in some normal individuals, GAN is thickened and therefore in leprosy endemic areas, if there is isolated thickened GAN, it would be necessary to confirm its involvement by nerve conduction study of this nerve (7). The elegant description by Mourad-Krohn in 1923 of the unique patchy bilateral facial paralysis in leprosy due to affection of small
Leprosy: Face beyond the cranial nerves

Leprosy is endemic in certain regions of the world, particularly the tropics. The clinical spectrum of leprosy ranges between two principal microscopic types: tuberculoid and lepromatous leprosy. In the tuberculoid type, cell-mediated immunity is good; although the skin exhibits limited clinical features, peripheral nerve involvement may be pronounced. Few, if any, demonstrable bacilli are present in these lesions. On the other hand, the lepromatous type represents poor cell-immunity, resulting in extensive intracellular bacilli in the skin and nerves. Borderline leprosy is an entity with features belonging to these two polar types.

Physicians and surgeons who frequently manage problems in the head and neck should be familiar with manifestations of leprosy in these parts of the body. Besides the usual features of possible cranial nerve deficits, unrelated structures in the face may be involved. In the nose, leprosy may result in obstruction, epistaxis, hyposmia, septal perforation and deformities. In the ear, cosmetic deformities of the pinna and external ear canal stenosis may result. In the mouth, there may be palatal destruction. These lesions are a result of lepromatous leprosy and are usually clinically obvious. An uncommon facial manifestation is hemifacial erythematous swelling, which is a result of reversal reaction. Although reversal reactions can occur in tuberculoid and lepromatous types of leprosy, nearly 90% occur in the borderline type.

Spontaneous remission is frequent in leprosy, whereas 'reversal' is lesser known. Reversal reactions are episodes of acute inflammation affecting skin and nerves, occurring when a patient develops increased cell-mediated immunity towards M. leprae and moves towards the tuberculoid end of the leprosy spectrum. The clinical features may be confused with those of facial cellulitis and sinusitis. It is important to accurately diagnose and promptly treat this condition, as the rapid and severe nerve destruction may result in permanent damage.

References