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Bilateral Claw Hands With Trophic Changes: Syringomyelia Or Leprosy- A Diagnostic Dilemma

KEYWORDS: Syringomyelia, Sensorimotor deficit, Leprosy.

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ABSTRACT *Syringomyelia is a disorder in which a cyst or cavity forms within the spinal cord. The damage may result in pain, paralysis, weakness, and stiffness in the back, shoulders, extremities and characteristic sensory deficit. In view of the insidious and widespread dissemination of leprosy, its discrimination from the diseases with close resemblance is a matter of considerable importance. It is important to suspect and recognize syringomyelia in a country like India where Hansen's disease is highly endemic to execute proper treatment. We are presenting a case of syringomyelia which is one of the rare differential diagnoses for leprosy.*

Introduction:
Syringomyelia which literally means 'tube within marrow' was first coined by Ollivier d'Angers in 1827.[1] It is a disorder in which a cyst or cavity forms within the spinal cord. This cyst called a syrinx, can expand and elongate over time, destroying the spinal cord. The damage may result in pain, paralysis, weakness[2] and stiffness in the back, shoulders, extremities and characteristic sensory deficit. These symptoms typically vary depending on the extent and often more critically to the location of the syrinx within the spinal cord.

Leprosy which is caused by Mycobacterium leprae is also characterized by sensorimotor deficit in the form of trophic changes & muscle paralysis involving upper & lower extremity. Syringomyelia is one of the rare and close differential diagnoses for leprosy due to presence of sensorimotor deficit. It is important to suspect and recognize syringomyelia in a country like India where Hansen's disease is highly endemic to execute proper treatment.

Case report:
A 42 year old male patient presented with bilateral claw hands with depigmented skin over fingers since 3 years (fig 1).

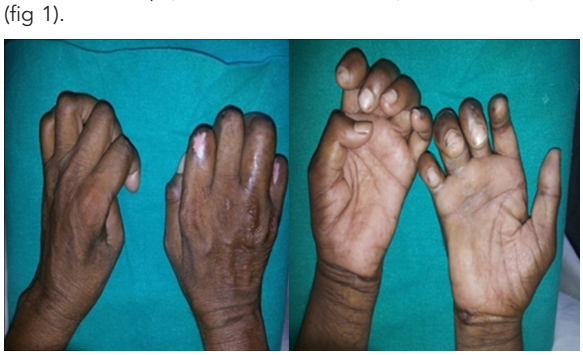

Figure 1: Bilateral claw hands with trophic changes over fingers.

There was no history of trauma over cervical region or upper extremity. On examination no skin lesions were seen and peripheral nerve examination findings were within normal limits. Pain & temperature sensations over upper extremity were impaired. Light touch was preserved. Deep tendon reflexes in upper extremity were lost. There was loss of muscle bulk of left forearm. Patient had bilateral drooping of shoulders (fig 2). Metacarpo-phalyngeal joints revealed hyperextension with flexion of inter-phalyngeal joint involving both the hands. Post inflammatory depigmentation due to healed ulcers was present over fingers of both hands.

Figure 2: Bilateral drooping of shoulders and loss of muscle bulk of left forearm.

Routine blood investigations were within normal limits. X-ray chest PA view was normal. Nerve conduction test findings of ulnar and median nerve were within normal limits. X-ray cervical spine was suggestive of degenerative

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changes in cervical spine. MRI cervical and thoracic spine revealed cystic swelling extending from C-3 to T-12 which was suggestive of syringomyelia (fig 3).

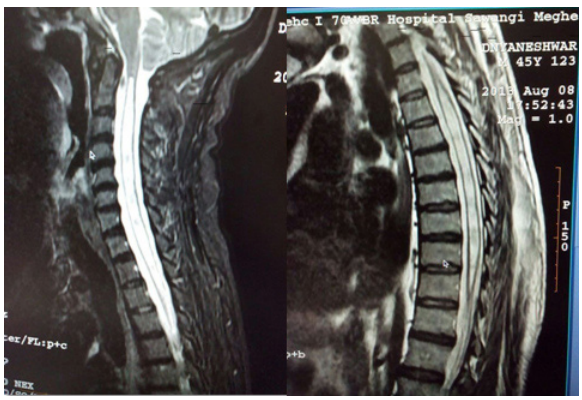

Figure 3: MRI cervical and thoracic spine revealed syrinx extending from C-3 to T-12 suggestive of syringomyelia.

Discussion:

Differential diagnosis of patient presenting with distal sensory loss and trophic ulcer are Hansen's disease, syringomyelia, hereditary sensory neuropathy - type I and III, tangier's disease, amyloidosis and diabetic neuropathy. Peripheral neuropathy produces symptoms and signs in symmetrical fashion involving upper and lower extremities. In case of peripheral neuropathy affecting lower limb, cauda equina is one of the differential diagnosis which is characterized by dermatomal sensory loss with affection of bladder and loss of sensation over perianal area.

In present case, patient had signs and symptoms suggestive of sensory and motor deficit involving upper limbs. Hansen's disease and syringomyelia were the important differential diagnosis for these presenting signs and symptoms. In Hansen's disease there is presence of skin lesion, thickened peripheral nerve, loss of pain, touch & temperature sensation and deep tendon reflexes are either normal or may show exacerbated response.

Our patient had bilateral claw hand with trophic changes involving fingers without any skin lesions. So, initially we thought of pure neural type of leprosy which was ruled out by absence of nerve thickening. Since upper extremity was grossly involved with sensorimotor deficit without nerve involvement, we thought of syringomyelia. In our patient touch sensation was well preserved over upper extremities with loss of pain & temperature and bilateral biceps reflex showed diminished response.

Clinically these findings were suggestive of syringomyelia which was confirmed by presence of cystic lesion on MRI cervical and thoracic spine. Magnetic resonance imaging (MRI) is the gold standard in the diagnosis of syringomyelia.[3,4] Then patient was referred to neurosurgery department for further management.

In view of the insidious and widespread dissemination of leprosy, its discrimination from the diseases with close resemblance is a matter of considerable importance for proper management. We are presenting a case of syringomyelia which is one of the rare differential diagnoses for leprosy.

REFERENCE

1. Ollivier CP. Traite des maladies de la Moelle Epinie, contenant l'histoire atomique, physiologique et pathologique de ce centre nerveux chez l'homme Second Edition. Paris Mequignon-Marvis pere et fi ls. 1827; 178–83. | 2. "Neurosurgical considerations in posttraumatic syringomyelia-Home Study Program". AORN Journal. January 2003. Retrieved 2009-02-12. | 3. Jack CR, Kokmen E, Onofrio BM. Spontaneous decompression of syringomyelia: Magnetic resonance imaging finding. J Neurosurg. 1991; 74:283. | 4. Kastrup A, Nagele T, Topka H. Spontaneous resolution of idiopathic syringomyelia. Neurology. 2001; 57:1519-20.