Morvan's Disease Associated with Syringomyelia and Arnold-Chiari Malformation

Arnold Chiari Malformasyonu ve Siringomyeli ile İlişkili Morvan Hastalığı

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DISCUSSION

The correct answer is C: Morvan's disease.

The diagnostic work-up of a patient with deformity in the hands should include a thorough history, physical and neurological examination, and radiographic plain films.

The syringomyelic nature of Morvan's disease was previously described by Morvan in 1883. Usually, the neuropathic osteoarthropathy occurs during the later phases of syringomyelia, however joint findings may occasionally be the initial or predominant manifestation of the disease.[1]

It is well-documented that, in syringomyelia, the shoulder is the most commonly affected joint among the upper limb joints, the elbow being the next common (Table 1).[2] However, only a few studies have reported involvement of the hand and the wrist in syringomyelia.[1]

However, in the upper limb, there is little or no deformity in early cases. Deformities of the more advanced disease may include extension at the metacarpophalangeal joints, fixed flexion of the digits, wasting of the intrinsic muscles and sometimes trophic changes, swelling of the fingers, scars of burns, ulcers or excessive callus formation giving the classical picture of "la main succulente". "Morvan's disease" refers to the resorption of the terminal phalanges.[3]

Syringomyelia with or without Arnold-Chiari malformation is characterized by a slow progression over many years; the most frequently involved joints are the shoulders and the elbows.[4] In the present case, Morvan's disease of the hands was unrecognized for 10 years, and painless ulceration of the fingers with resorption of the terminal phalanges was the initial manifestation.

Dysesthetic pain is a common complaint of the patients with syringomyelia.[5]

The pathophysiology of syringomyelia involves the disruption of the adjacent gray and white matter. The initial fibers damaged are the pain and temperature sensory fibers as they cross the midline. Their loss in the upper extremity, with intact position sense and motor function, is often the first clinical sign. It is the loss of these sensory fibers that is thought to result in the etiology of neuropathic arthropathy of the shoulder. As the syrinx grows or propagates, the dorsal column fibers or anterior horn cells may be affected. Areflexia, muscle weakness, and atrophy results, commonly involving the hand intrinsics. Additionally, disruption of the sympathetic pathways may result in a Horner's disease or vasomotor and trophic changes, again most common in the hand.

In our case, the syrinx propagated proximally over time resulting in Morvan's disease, and the patient presented to our neurosurgery clinic with an unusual-appearing deformity in her hands. Morvan's disease requires evaluation of the spinal cord to assess occult causative lesions. Therefore, radiological examination of spinal cord is necessary in these cases.

Morvan's disease is a rare disease with clinical features of vasomotor problems, digital ulcers, thickening of the fingers, and absorption of the terminal phalanges.[4]

Leprosy was excluded because the patient had spent all his life in the neighbourhood.[6] We also excluded Raynaud's disease and sclerodactylia, which were suggested as possibilities since cold-simulation test and Wassermann's test were negative. She was not a diabetic patient, and gave no history of exposure to sexually transmitted diseases. Therefore, diabetes and tabes dorsalis were excluded. In addition to these, there was no spina bifida in neurological and radiological examinations. The differrential diagnosis is summarized in Table 1.

We report a rare case of absorption of the terminal phalanges, also known as Morvan's disease, secondary to syringomyelia. The pathophysiology, clinical findings, and diagnostic work-up options are discussed.

TABLE 1: Common sites of involvement in neuropathic osteoarthropathy.

Disease	Site of involvement
Tabes dorsalis	Knee, hip, ankle, spine
Syringomyelia	Glenohumeral joint, elbow, wrist, spine
Diabetes mellitus	Metatarsophalangeal, tarsometatarsal, intertarsal joints
Alcoholism	Metatarsophalangeal, interphalangeal joints
Amyloidosis	Knee, ankle
Meningomyelocele	Ankle, intertarsal joints
Congenital sensory neuropathy, hereditary sensory radicular neuropathy	Knee, ankle, intertarsal, metatarsophalangeal, interphalangeal joints
Idiopathic	Elbow, shoulder

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