Tr. J. of Medical Sciences
29 (1999) 711-713

Short Report

Flexion Contractures in a Diabetic Young Man (Rosenbloom Syndrome)

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Received: 03.07.1998

Key Words: Diabetes Mellitus, Rosenbloom syndrome, limited joint mobility

Introduction

Limited joint mobility syndrome was initially described by Rosenbloom as painless limitation of the fingers and large joints in association with short stature, thick, tight, waxy skin, delayed sexual maturation, and early microvascular complications (1). Limited joint mobility (LJM) is thought to be a manifestation of the diffuse collagen abnormalities in diabetic patients (2). Limited joint mobility is of particular interest because it is common in young patients and associated with an increased risk for the microvascular complications of diabetes (2,3). A 21 year-old diabetic man who had multiple joint contractures and nonfamilial short stature with microvascular complications is presented.

Case Report

A 21-year-old man with insulin-dependent diabetes mellitus (IDDM) complained of stiffness and progressive limitation of the mobility of his joints for one year. He also complained of severe paresthesias and pain in his hands and feet. He had had IDDM for 17 years. He had never experienced arthritis and there was no family history of rheumatic disease. His mother had non-insulin-dependent diabetes mellitus (NIDDM).

His weight was 40 kg and his height was 146 cm. He had flexion contractures at the distal interphalangial, proximal interphalangial and metacarpophalangial joints of his hands and elbows, and the metatarsophalangial, proximal interphalangial and distal interphalangial joints of feet (Figure 1). His hands and feet were more severely affected. He was unable to use his hands properly and wear shoes. The range of motion in his wrists, knees, ankles and subtalar joints was severely limited. He was unable to oppose the palms of his hands (Prayer sign) (Figure 2). Lumbosacral movement was limited in all directions. Modified Schober test was 13 cm. There was no local tenderness over the sacroiliac joints. He had hypothenar and thenar atrophy. He was in Tanner stage III of puberty. Pubic and axillary hair showed insufficient development and he had no facial hair.

Laboratory investigations showed normal results of complete blood counts. The fasting glucose level was 220mg/dl. The HbA1c level was 10% (<7%). Urinalysis revealed glucosuria (150mgr/dl) and macroalbuminurea (450mg/24 hours). The erythrocyte sedimentation rate was 25mm/h. C-reactive protein, rheumatoid factor, antinuclear antibodies were not detected. Immunoglobulins and complements were at normal levels.

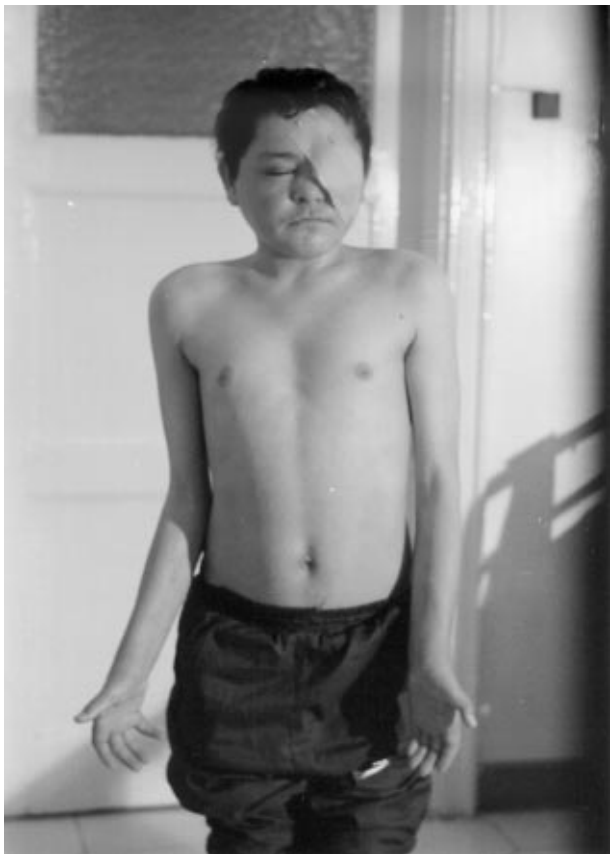

Figure 1. The patient with Rosenbloom Syndrome.

Electroneuromyography revealed severe sensorimotor polyneuropathy. Respiratory function tests were performed in order to establish the vital capacity. Forced vital capacity was 2 l (60% of the predicted value). Ocular examination showed neovascular glaucoma in the right eye and diabetic proliferative vitreoretinopathy in the left eye.

Skeletal age was 15 according to a wrist roentgenogram. A lumbosacral roentgenogram revealed a loss of lumbar lordosis. Facet joints and sacroiliac joints showed no abnormality. Feet roentgenograms revealed osteoporosis, tarsal abduction deformity, cavus deformity, severe Achilles tendon calcifications and large calcaneal spurs.

His pain was resolved with antiinflammatory medication. Range of motion exercises and stretching was performed for one month. There was no improvement in any joint range at the end of the program.

LJM has recently been recognized as part of the clinical spectrum of IDDM by Rosenbloom (1). LJM is seen not only in IDDM but also in NIDDM (4). The joints most frequently affected in LJM are the proximal interphalangial joints of the fingers, the wrists, elbows, spine, knees, ankles and the toes (5). Thick, tight, and waxy skin, most prominent over the dorsum of the hand

Figure 2. The prayer sign.

and the forearm was described in diabetic patients leading to restriction of joint movements (1, 2). The exact pathogenesis of LJM is unclear. Evidence suggests that the diabetic hyperglycemic state leads to an increase in nonenzymatic glycosylation causing increased cross-linking of collagen (2). The most commonly suggested explanation for LJM has been that impaired degradation of collagen leads to its accumulation (6). The degree of the cross-linked collagen correlates with the presence of retinopathy and nephropathy (2).

Rosenbloom et al. suggested that alterations of periarticular connective tissue are related to changes occurring in the microvasculature (3). Our patient with LJM had retinopathy, nephropathy and neuropathy as microvascular complications. Rosenbloom et al. found microvascular complications in 50% of patients with LJM (3). They suggest that after 16 years of diabetes, the risk for microvascular complications is 83% if LJM is present, but only 25% if LJM is absent. Starkman et al. found that LJM is associated with the duration of diabetes in IDDM patients of less than 40 years of age but did not find any relationship in NIDDM patients (7).

Our patient's pulmonary capacity was diminished. Shuyler et al. initially reported the diminished pulmonary capacity in diabetes (8). Barta subsequently described a single patient with limited joint mobility and limited pulmonary capacity (9). The pathologic findings in the thickened skin of subjects with LJM have been postulated to occur in the lungs as well (8).

Our patient also had diabetic hand syndrome superimposed on LJM. In diabetic hand syndrome there is pain, muscle atrophy, neuropathy and disability. None of them are seen in LJM (10). It is likely that people affected with LJM would be at higher risk of developing of other connective tissue proliferative problems and possible neuropathy (2).

Our patient had no improvement in joint range with physical therapy. However Rosenbloom reported the responsiveness of LJM to physical therapy, and Sherry et al. described an adolescent patient with LJM who had improvement in joint range with physical therapy (2,5). It was reported that for the prophylactic treatment of LJM in diabetic patients, range of motion exercises should be done routinely (11).

LJM is a risk factor for the development of microvascular complications in diabetic patients. Earlier detection of LJM may help us to identify patients with an increased risk of microvascular complications. Beginning the prophylactic range of motion exercises after the diagnosis of diabetes may help prevent future contractures and disability in diabetic patients.

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