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Toe Walking as a Presenting Sign of Systemic Lupus Erythematosus

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Keywords

Lupus; Musculoskeletal; tendon; contracture; toe walking

Introduction

Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disorder that can affect any organ system. Musculoskeletal involvement is one of the most prevalent organs involved with a wide range of manifestations.[1, 2] Arthralgia and arthritis represent the largest proportion of musculoskeletal involvement, followed by myalgia, subcutaneous nodules, avascular necrosis, myopathy and tendon rupture.[1, 3–5] We report a case of toe walking and multiple flexion contractures without arthritis as a presenting symptom.

Case

A previously healthy 14 year old Vietnamese female had a transient facial rash for several months and then, one month prior to presentation, developed toe walking and generalized tight tendons. She then developed thinning of her hair and a rash on her hands and ears and, two days before presentation, developed left sided chest pain. The morning of admission she woke up with mild confusion. She could not recall her phone number and “did not seem right” to her father. She had a dry mouth, generalized weakness, and left upper quadrant pain and was taken to an emergency department where she was found to be pancytopenic with cardiomegaly. She had a generalized tonic clonic seizure upon arrival to our hospital that was aborted with lorazepam. Further evaluation revealed vasculitic rash with digital ulcers, white blood cell count 2.1, hemoglobin 10.2 g/dL, platelets 61 thousand/uLiter, Absolute Neutrophil Count (ANC)1040 uL, Absolute Lymphocyte Count (ALC)800 uL, Direct Coombs Negative, Alanine Aminotransferase 76 U/L, Amylase 111 U/L, Lipase 529 U/L, Aspartate Aminotransferase 343 U/L, Creatinine Kinase 183 U/L, Triglyceride 429 mg/dL, Ferritin 1450 ng/mL, D-Dimer 3.9 ug/mLFEU, Fibrinogen 140 mg/dL, ESR 68 mm/hr, C-Reactive Protein <0.5 mg/dL, urine protein/creatinine ratio 2.1, C3 11 mg/dL, C4

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Conflict of Interest Statement

The authors report no conflicts of interest.

1.8 mg/dL and CH50 196 mg/dL. She had the following elevated antibodies: ANA 1:1280 in a speckled pattern, DNA Binding 246 IU/mL (0-99.9), Anti-Smith AB 135.7 units (0-19), Anti-RNP 132.3 Units (0-19), Anti-SSA 106.6 Units (0-19) and Ribosomal P 150 AU/mL (0-40). She had the following antibodies in the normal range: Anti-SSB, NMO (Cerebral Spinal Fluid and Serum) Anti-Cardiolipin IgG and IgM, Anti-Beta-2-Glycoprotein IgG and IgM, Rheumatoid Factor and Scl-70. Her Dilute Russell Viper Venom Time was normal. She had features consistent with Macrophage Activation Syndrome (MAS) given her pancytopenia (white blood cell count 3.5, hemoglobin 7.9 g/dL, platelet count 29 thousand/uL, ANC 3825 uL, ALC 234 uL), hypertriglyceridemia 429 mg/dL, hypofibrinogenemia 121 mg/dL, hyperferritinemia 1450 ng/mL and an elevated d-dimer 4.3 ug/mLFEU. She was treated with 5 days of methylprednisolone 30 mg/kg/day, apheresis, cyclophosphamide, hydroxychloroquine and rituximab.

One of the most prominent findings on her examination included profound limitations in range of motion of joints. She was unable to stand with her heels to the floor due to ligamentous, not joint, tightness (figure 1). With her wrist flexed she had full finger range but when the wrist was put in neutral to extension she had progressive finger contractures at the proximal and distal interphalangeal joints (figure 2). Additionally, her shoulder abduction was limited to 100 degrees bilaterally and she had hip flexion contractions. Her memory, knowledge and attention were normal as were cranial nerves II-XII, extremity strength, sensation, and coordination. Her reflexes were brisk throughout with a crossed adductor on the right and slightly more brisk overall on the left. A computed tomography scan of her head demonstrated enlargement of the lateral and third ventricles and cerebral sulci with mild rounding of the third ventricle with mild prominence of the right temporal horn. A magnetic resonance (MR) angiogram of the brain with and without gadolinium showed abnormal T2/Flair hyperintensity within the optic chiasm, hypothalamus and intracranial portion of the optic nerve without evidence of vasculitis or venous sinus abnormality. An MRI of her entire spinal column with and without gadolinium was normal. A neuro-ophthalmology evaluation was normal. MR images of her lower extremities with and without gadolinium at 3 months after presentation were normal. She has been in physical and occupational therapy over the past 9 months including serial casting for 8 weeks and had only slight improvement in her range of motion.

Discussion

Toe walking is considered part of the normal immature gait, but is expected to resolve prior to school age years. [6] Our patient had a meticulous evaluation with her underlying lupus ultimately being the etiology for her flexion contractures and resultant toe walking. She participated in intensive physical and occupational therapy including serial casting. Despite this and the fact that her lupus is otherwise under control, at nine months since onset she continues to have contractions. Of interest, we care for another teenage female with long standing lupus, who, over time, developed similar tight finger flexion tendons without arthritis. Although SLE can present with a wide range of musculoskeletal manifestations, to our knowledge, toe walking has not been reported as a presenting symptom. There is a case report of spontaneous contractures of abductor digiti minimi in a patient with lupus thought to be related to myositis and was managed surgically. [7] There are no reports of widespread

tendonopathy and in particular significant Achilles tendonopathy. The differential diagnosis for Achilles tendonopathy includes inherited disorders, metabolic processes, rheumatoid arthritis, gout and spondyloarthropathies, none of which were present in our patient. [8] While our patient had an MRI performed, there is a potential role for ultrasound to detect subclinical tendon thickening or effusions in patients with persistent toe walking. Lupus should be considered in the appropriate clinical context in children who have new onset contractures due to tight tendons and it may be very resistant to physical and occupational therapy.

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Figure 1.
Posterior (A) and lateral (B) view of her resting standing position of her ankles

Figure 2.
Resting finger position with wrists in neutral (A) and with wrists extended (B). Her fingers could not be extended with the wrists in extension