MYOFIBROSIS

Case Report

History

A 6-year-old boy was referred to our clinic from the pediatric service for evaluation of stiff knees bilaterally. He was born after a normal pregnancy, developed normally, and walked well until the age of three years. At that time he was hospitalized for abdominal surgery to repair a congenital muscular defect in his abdominal wall. Since the surgery, he has never regained completely his preoperative ambulatory status.

Physical Examination

The patient walks fairly well, but has a stiff-legged gait. As he sits on the edge of the examining table there are extension contractures in both knees—the right more extreme than the left (Figs. 1A, 1B). There is 30° of flexion in the right knee and 60° in the left; this has been a slowly progressing deficit. Range of hip motion is normal and his feet appear normal. There is no evidence of internal derangement of the knee. The knee area has no warmth, redness, or apparent effusion. Knee flexion is smooth through its entire arc, but halts suddenly at its extreme. There are no clicks or crepitus, and no locking. Collateral ligaments are stable bilaterally, and the cruciate ligaments are intact.

Roentgenographic Interpretation

On anteroposterior and lateral views the knees are unremarkable visually (Figs. 2A, 2B). Nothing appears unusual except that the patella rides a little high.

Discussion

This patient has a fibrosis in at least one component of the quadriceps. During his hospitalization at age three he received multiple intramuscular injections into his thighs. For children, the usual site of injection accepted now in the U.S.A. is in the middle third of the thigh, in the anterolateral portion. Until January of this year, 12 cases of myofibrosis as a complication of intramuscular injection had been reported in the U.S.A. Since then Alvarez reported an additional 17 cases. Muscular fibrosis is a syndrome that has been described as occurring in the quadriceps, most commonly in children; it has also been described as occurring in the deltoids and in the gluteals. More than 2000 cases involving the quadriceps have been reported in the Japanese literature.

Many of the Japanese cases were reported to involve a rectus femoris contracture, in which a hip flexion contracture as well as an extension contracture of the knee can be seen frequently.

Muscular myofibrosis was described first in the literature in 1961 by Hnevkovsky. The condition occurred in eight girls and four boys, and he suggested
that its cause was some sort of congenital myodysplasia. Fairbanks described this condition in twin girls; he suggested that possibly the lesion might be similar to congenital torticollis. The next significant reports of this condition were two articles that appeared in a 1964 British journal, one by Lloyd-Roberts, another by Dr. Gunn of Singapore. Both of these articles tended to implicate intramuscular injections. Lloyd-Roberts reviewed the cases reported by Fairbanks and Hnevkovsky, and the others that had been published in the interim, and found that in about two thirds of those cases the patients had a definite history of frequent intramuscular injections into the muscles that were contracted. Since that time, there have been multiple reports of the condition, mostly in children, although Groves and Goldner described several cases of deltoid contracture in adults. In early reports, the disease was believed to affect females much more frequently than males. Since then, particularly in the report by Alvarez, it appears that the disease probably has no sexual predilection.

The differential diagnosis includes internal derangements of the knee and arthrogryposis, which would be an incomplete type of syndrome. Muscular myofibrosis is sometimes believed to be somewhat similar to torticollis, in which the sternocleidomastoid contracts. Some believe the disease may be similar to camptodactyly, which involves the same sort of contraction in the fingers, although that condition usually appears later in life. Another possibility in the differential diagnosis is some sort of fibromatosis of malignant origin.

Myofibrosis presents in two ways. The more common one, as described in this case report, is an extension contracture of the quadriceps. The second presentation is a dislocation of the patella laterally. A child with such dislocation usually has no real deficit in knee flexion; as the knee reaches a certain point the patella dislocates laterally and allows the flexion arc to continue. In a child with patella dislocation, however, if the patella is kept in the trochlear groove by hand pressure, an extension contracture also will be evident. Generally the patellar dislocation is associated with primary involvement of the vastus lateralis, and usually is a later finding. The patellar manifestation brings to mind patella alta and habitual dislocation of the patella, but these conditions are not associated with extension contractures of the quadriceps.

The histologic findings are those of active noninflammatory fibrosis, in and around atrophic muscle fibers, and any of the components of the quadriceps group may be involved. Vastus intermedius and vastus lateralis are reported most commonly in the U.S.A. Japanese reports implicate a frequent rectus femoris involvement, and all reports cite vastus medialis involvement as unusual. If a previous intramuscular injection cannot be implicated, the condition may be due to contractures similar to those seen in torticollis or in club foot, or may represent some localized form of arthrogryposis. This condition would be expected to appear at, or shortly after, birth. Other conditions that should not be confused with this syndrome are those in which extremities are spastic, such as cerebral palsy or myelomeningocele.
Treatment

Except in the mildest cases, efforts to halt or reverse this process with vigorous physical therapy have been unsuccessful; Alvarez reports one complication of a supracondylar femur fracture suffered during physical therapy. Surgical release of the affected muscles and/or tendons is the accepted therapy, but release must be extensive; eventual flexion will never exceed that obtained at the time of surgery, and in some cases will be less. In some instances, the fascia lata may need to be released from the lateral border of the patella, where it is frequently found to be adherent, especially in those patients with patellar dislocation. Once the fascia lata and fibrous portions of the quadriceps have been released and excised, the medial patellar retinaculum usually does not need reinforcement to prevent recurrent dislocation. After surgical release, patients must be immobilized in long-leg casts with 90-120° of knee flexion for 3-6 weeks. Active physical therapy is then instituted. Of the reported cases, most children have had initial extensor lags of 15-30°, but the lag usually resolves gradually over 12-18 months.

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