PROGRESSIVE FIBROSIS OF THE QUADRICEPS MUSCLE

Isik Akgün, MD
Önder Aydingöz, MD
Hayrettin Kesmezacar, MD
Tuncay Centel, MD

Limitation of active and passive flexion of the knee without pain developing secondary to fibrosis of the quadriceps muscle has been the focus of many studies. In 1961, Henevovsky10 coined the term "progressive fibrosis of the vastus intermedius muscle in children" and reported that this could be a kind of muscle dysplasia. In the same year, Fairbank and Barrett8 observed this type of contracture in a pair of premature twins.

Lloyd-Roberts and Thomas6 reported that the common factor in these cases was that patients all had intramuscular injections in the thigh, and this factor played a role in these cases. Mukherjee and Das7 presented mid-term results on 50 cases of quadriceps contracture in 37 children. No definite conclusion on the etiology of these type of cases has been proven or reported.

This article describes progressive fibrosis of the quadriceps muscle in a 10-year-old girl.

CASE REPORT

A 10-year-old girl was referred to our clinic with limited flexion of her right knee of 6 months' duration. She had undergone physical therapy for 1 month without any improve-

Fig 1: Preoperative flexion in the right knee was limited to 90°.

Fig 2: MRI confirmed patella alta in the right knee (A). The transverse MRI of the right thigh shows the tendinous structure of the vastus intermedius muscle (B).

ment. She had been born after a full-term pregnancy and an uncomplicated delivery with a birth weight of 3400 g. The postnatal period was normal except for an umbilical cord infection. The patient had remained in the hospital for treatment of this infection for 1 week and was given many injections in both thighs. Her medical history had been normal until she began experiencing increasing disability in her right knee.

On physical examination, she had full active and passive extension, but only 90° of flexion (Fig 1). A bandlike depressed area was observed on the lateral side of the thigh when the patient flexed her knee. No stretched band was palpated. Both legs were equal in length. The right patella was situated 1.5 cm more proximal than the contralateral patella, and her right thigh was atrophied compared with the left.

Radiographs were normal except for patella alta in the right knee. Magnetic resonance imaging (MRI) demonstrated patella alta in the right knee and hypoplasia of the vastus intermedius muscle from the mid-femur extending distally to form a tendinous structure. Signal intensities in accordance with the fibrosis process and fatty tissue also were observed in the lodges of the thigh (Fig 2).

Her neurologic examination and laboratory studies were normal. The diagnosis was progressive fibrosis of the quadriceps muscle, and the patient underwent surgery. The muscle was exposed through an anterolateral incision. The rectus femoris, vastus medialis, and vastus lateralis muscles appeared normal, but the vastus intermedius was a stretched fibrous band attached to the periosteum of the femur. Subcutaneous adhesions were released by blunt dissection, and the fibrous vastus intermedius muscle was divided and partially excised. The quadriceps femoris muscle was lengthened using a V-Y plasty until 140° of flexion was achieved. Biopsy material was taken from the fibrotic bands, the skin was closed, and an anterior ankle-to-thigh cast was applied with the knee flexed at 110°.

Postoperatively, the cast was removed after 2 days, and the patient began using a flexion brace. Passive knee motion during daytime also was begun. At night, the knee was kept in 120° of flexion. By the end of the second postoperative week, the flexion degree of the brace was increased to 140° and quadriceps exercises were started. A small detachment at the incision site healed without any problem. The patient was started on a physical therapy program after the quadriceps muscle
strength failed to improved satisfactorily.

Two months after surgery, the patient had full knee extension, 120° of active and 135° of passive flexion, and continuity of quadriceps atrophy (Fig 3). She was able to walk without any physiologic problems and still used the brace at night. The patella was in its correct position (Fig 4).

Histopathologic examination of the tissue samples showed hyalinized connecting tissues. Mature fatty tissues also were present in this connecting tissue. In the sample taken from the medial side of the vastus intermedius muscle, focal proliferation was present in synovial cells covering the fascial connecting tissue. Rare inflammatory mononuclear cells and vascular sections also were observed under the proliferated synovia. In some areas of the material, intensive collagen fibers, wavy and partially lined, were noted.

**DISCUSSION**

Although quadriceps contracture in children was unheard of until 30 years ago, its incidence, particularly in Asian countries, has been on the rise in recent years. For this reason, its uncertain etiology has been debated. The pathology has been observed not only in the quadriceps muscle but also in the deltoid, glue
tal, and triceps muscles. The incidence is higher in girls, and the average age is 4 years. In some cases, the contracture starts at birth.

and generally progresses, slowing in later childhood years. Our patient was 10 years old when the lesion was diagnosed.

Hnevkovsky, who first defined the pathology, believed it was a muscle dysplasia of genetic origins. Fairbank and Barrett and Gammie and Taylor concluded that the findings of their cases were similar to localized arthrogryposis and congenital torticollis, and thus proposed genetic factors in the etiology. Others supported this view. In some cases, the contracture was bilateral.

Fibrosis in other muscles together with the quadriceps muscle also has been reported. The fact that fibrosis is observed in more than one site in many cases supports a genetic etiology. Furthermore, it has been reported that scar tissue at the incision site demonstrated hypertrophic growth in patients who underwent surgery.

Lloyd-Roberts and Thomas reported that multiple thigh injections in the postnatal period played a major role in the etiology of progressive fibrosis of the quadriceps muscle. Most of the drugs used in the injections were antibiotics and vitamin K6. In a study of 71 cases, Fernandez and Esteve attributed fibrosis of the gluteus maximus muscle to postnatal injections. They believed the histology was similar to muscle dystrophies.

Others have noted that intramuscular injections of antibiotics, vitamins, and antitetanic serums were administered in cases of progressive quadriceps fibrosis. In general, as with the patient described here, patients were given injections in the postnatal period for pathology. The lower the age at injection, the more serious the pathology. In a rat model, Jarvinen et al demonstrated that hematoma, scar formation, and inflammatory reaction of the muscles were distinct at early ages. We believe the injections our patient received in the postnatal period were the determining factor.

There are other factors to consider in chronic compartment syndrome. Among the quadriceps muscle group, the muscle that is most affected is the vastus intermedius, whereas the vastus medialis is almost never affected. The fact that the injection site is usually on the lateral side of the thigh and the vastus intermedius muscle is less vascular may explain why this muscle is at more risk. Matsuse et al reported post-traumatic fibrosis of the gastrocnemius muscle. They connected this incident with the limited vascularization and anastomoses in this muscle. It also has been proposed that in addition to the increase in the compartment pressure due to the amount of the injected materials, the toxic effects of the injected materials also cause fibrosis and secondary contracture.

We believe injections play a major role in the etiology of muscle fibrosis through both volume and toxicity. However, considering the fact that not all babies who are subjected to multiple injections develop such a pathology, personal factors including genetic features should not be completely ruled out. Therefore, such a tendency that is already present genetically may combine with another factor to start the fibrosis process in areas of poor vascularity. This factor is usually intramuscular injections, although in rare cases, it may be trauma.
Although the etiology of quadriiceps contracture in children is controversial, the treatment of choice is surgery. Contracted sections should be excised, and a Z or V-Y plasty performed if necessary. In late cases, capsulotomies are also necessary. In progressive cases, there may be recurrent patella dislocation, changes in the ligaments and joint capsule, and secondary disorders in the patellar cartilage. For these reasons, early surgical treatment is recommended.

In our case, patella alta was noted and despite excision of the fibrosis, only 120° of flexion was achieved intraoperatively because of shortening of the quadriiceps muscle and patella alta. Therefore, a quadriiceps plasty also was performed. In cases with lateral patellar luxation, treatment should be combined with a lateral release.

REFERENCES