Congenital contracture of the quadriceps muscle: confirming the diagnosis with magnetic resonance imaging

Oya Özdemir a,*, Ayçe Atalaya a, Reyhan Çelikera, Ülkü Kerimoğlu b, Özhan Özdemirc

a Department of Physical Medicine and Rehabilitation, Hacettepe University Medical School, Ankara, Turkey
b Department of Radiology, Hacettepe University Medical School, Ankara, Turkey
c Second Orthopedic and Traumatology Clinic, Ankara Numune Training and Research Hospital, Ankara, Turkey

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Abstract

Congenital contracture of the quadriceps muscle can be defined as progressive loss of knee flexion due to fibrosis within the muscle without a history of trauma or intramuscular injection into the thighs. In the course of time, secondary changes might develop and vitiate the end result so this rare childhood disease needs particular attention for early diagnosis and treatment. Herein, we report a 14-year-old girl presented with inability to bend her knees completely. The clinical and radiological assessment was detailed with magnetic resonance imaging findings.

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1. Introduction

Unilateral or bilateral contracture of the quadriceps muscle, resulting in limitation of flexion of the knee joints in children, is not an uncommon entity. It is classified into three types according to the part of the muscle which is affected—namely rectus femoris, vastus and mixed type [1]. Among the etiological factors mentioned in the literature, the most common is administration of intramuscular injections into the thighs. In case of significant limitation of motion interfering with activities of daily living, surgery is inevitable choice of treatment with satisfactory results. In this report, we render a case of congenital quadriceps contracture which is seen extremely rare in daily practice, including magnetic resonance images.

2. Case report

A 14-year-old girl was seen for the complaint of inability to bend both knees completely. She declared that she had never experienced knee pain. The age of onset was obscure. It was first noticed by her parents when she was 5 years old and became more obvious as she grew. She had been born in full term by spontaneous delivery and her subsequent development was normal. There was no history of local injections or trauma to the thighs and also no family history of a similar condition.

Physical examination revealed that in supine position the range of both knee joints with the hip in flexion were from 0 to 100°. When the hips were extended by taking the patient in prone position, right knee flexion was restricted to 80° and the left one to 90°. Furthermore, it was disclosed that in prone position when the knee was forced to flexion beyond 80°, the unilateral hip was also forced to flexion simultaneously (Fig. 1a, b). However, no extension lag of the knee joints was detected. Bilateral painless tenseness were palpated in both quadriceps. The power of the muscles within the range of motion was normal. No evidence of any vascular or neurological abnormality was observed.

Complete blood count, erythrocyte sedimentation rate and serum biochemistry including creatine phosphokinase were all within normal ranges. Radiographs of the knee joints were considered as normal except contour irregularity and fragmentation of the right tibial tuberosity (Fig. 2). Transverse T1-weighted spin-echo and T2-weighted gradient-echo magnetic resonance images demonstrated that bilateral vastus lateralis muscles were diminished in volume, associated with a thick...
fibrous structure anteromedially located within the muscle, and fatty atrophy was detected on both sides (Fig. 3a, b).

After the diagnosis of congenital contracture of the quadriceps muscle was established with clinical and radiological findings, the patient was referred to the Department of Orthopedic Surgery. Since the patient had no functional loss in her daily activities apart from slight difficulty in sitting with full flexion of the knees, no surgical treatment was recommended and she was called for controls.

3. Discussion

Contracture of the quadriceps muscle, which is characterized by progressive loss of knee flexion due to fibrosis within the muscle, is not an uncommon entity. It was first described by Hnevovsky [2] as a muscular dysplasia of congenital origin of the vastus and rectus femoris muscles. Fairbank and Barrett [3] reported identical twins with similar findings, thus they suggested a genetic origin to explain the etiology of this condition. Additionally, six cases were noted who had limitation of full flexion of the knee joints with an unknown origin. Two of these patients had concurrent other congenital anomalies like Hnevovsky’s three cases and this fact led Karlen to postulate this condition as a congenital disease [4]. Histological examinations of affected muscle have thrown little light on the subject. The biopsy materials, obtained from the patient who

Fig. 1. In prone position the right (a) and the left knee joint (b) flexion were restricted to 80 and 90 degrees, respectively. When the knee was forced to flexion beyond this limitation, the unilateral hip was also forced to flexion simultaneously (b).

Fig. 2. Lateral plain film of the right knee, depicting contour irregularity and fragmentation of the right tibial tuberosity.

Fig. 3. Transverse T2-weighted gradient-echo MR image (a) showing bilateral vastus lateralis muscles reduction in volume, associated with a thick fibrous structure anteromedially located within the muscle. Transverse T1-weighted spin echo MR image (b) demonstrating fatty atrophy and fibrosis in the anterior aspect of bilateral vastus lateralis muscles.
underwent surgical treatment, showed considerable excess of collagen and infiltration of muscle by fatty tissue [2,3,5]. They were partly in the form of dense fibrous strands which had a few buried muscle fibers and partly in the form of loose fibrofatty tissue. Nowhere did the nuclei show degenerative changes such as pyknosis or clumping [5]. On the other hand, Gunn suggested that an important factor in the etiology is the administration of intramuscular injections into the thighs. Among the 22 cases with shortening of the quadriceps, 15 had a history of such as pyknosis or clumping [5]. On the other hand, Gunn suggested that an important factor in the etiology is the administration of intramuscular injections into the thighs. Among the 22 cases with shortening of the quadriceps, 15 had a history of injection therapy was certainly used [6]. Subsequently, quadriceps contracture as a complication of multiple intramuscular injection have been reported with increasing frequency [7–10]. Because our patient had no history of trauma or injection into thighs, we named this condition as congenital contracture of the quadriceps muscle which is seen extremely rare in daily practice.

Ad Hoc Committee on Muscular Contracture, which was formed by the Japanese Orthopedic Association in 1975, classified quadriceps contracture into three types as the rectus femoris, vastus and mixed type [1]. According to this classification our patient’s physical examination was consistent with the mixed type. Because the examination revealed not only the vastus type symptom as restricted flexion of the knee joint with hip flexion and also the rectus femoris type symptom of restricted knee flexion in the prone position.

Contracture of the quadriceps muscle may be unilateral or bilateral. The most striking clinical finding is the definite block to flexion of the knee at a variable angle. In due course secondary changes develop in the capsule and the ligaments, as well as in the articular cartilage and later in the bony parts of the joint, unfortunately those will vitiate the end result. In long standing cases there is marked flattening of the femoral condyles, particularly of the lateral condyle which may be explained by the tightness of the tensor fascia lata [4]. Likewise, fibrosis of the quadriceps muscle can drag the patella to a higher than normal position and stretching of the patellar tendon takes place during this process [6]. Indeed, radiographs of the right knee joint of our patient showed the contour irregularity and fragmentation of the tibial tuberosity, probably resulting from patellar tendon tightness across the joint and loss of flexibility.

Indisputably, because the disability itself is largely mechanical, it can be relieved by surgery [5]. Operative treatment is suggested when the knee flexion is limited to 30° or less in prone position [1]. The most satisfactory results were obtained from the simplest division of the affected muscles [1–5]. The optimum age for surgery is about 5–6 years in order to have the child’s cooperation in active physiotherapy [5]. Furthermore, this procedure will be more efficient if it is performed at an earlier age before the irreversible secondary changes occur [4]. Recently, it was stated that magnetic resonance imaging (MRI) will be helpful to demonstrate the extent of fibrosis before surgery [11]. No surgical treatment was recommended for our patient since her disability was so slight.

In the relevant literature there is only one case report of congenital quadriceps contracture with MRI findings [11]. To the best of our knowledge, we now present the second case with similar radiological views of fibrosis and replacement of muscle by adipose tissue. Thus, we want to draw attention to the role of MRI in confirming the clinical diagnosis. This rare childhood disease should be kept in mind so that early diagnosis and treatment might prevent further complications.

References