Case Report

Idiopathic Fibrosis of the Quadriceps Muscle: A Case Report with Magnetic Resonance Imaging and Pathological Findings

Rachawan Suksathien MD*

* *Department of Rehabilitation Medicine, Maharat Nakhon Ratchasima Hospital, Nakhon Ratchasima*

Background: *Idiopathic fibrosis of the quadriceps muscle can be defined as progressive loss of knee flexion due to fibrosis within the muscle developed in early childhood. A few cases have been reported, but these did not detail the MRI and pathological findings.*
Material and Method: *A 28-year-old woman presented with difficulty in sitting and inability to bend both knees completely. The clinical evaluation, radiological examination, MRI, treatment and muscle pathology were demonstrated.*
Results: *The knee flexions were limited in both knee joints. Magnetic resonance imaging of both thighs showed marked quadriceps muscle atrophy with fat infiltration and fibrous band. Intraoperative findings demonstrated fibrous band of vastus intermedius and vastus lateralis muscles which correlated with the MRI. The histological findings also revealed loose connective tissue separating muscle fibers and large fibrous band. Z-plasty and release of the fibrotic muscles were performed. The result of the surgery was good.*
Conclusion: *This is the first report to correlate MRI and pathological findings of idiopathic fibrosis of the quadriceps muscle. Late diagnosis and treatment may produce irreversible changes of femoral condyles and patella. Clinical awareness with history, physical examination and radiographic findings which included MRI help in confirming the diagnosis and demonstrating the extent of fibrosis before surgery.*

Keywords: *Idiopathic fibrosis, Quadriceps contracture, Magnetic resonance imaging, Pathology*

J Med Assoc Thai 2008; 91 (4): 568-73
Full text. e-Journal: *http://www.medassocthai.org/journal*

Idiopathic fibrosis of the quadriceps muscle is not a common entity. A few cases with clinical and pathological findings have been reported. Recently, magnetic resonance imaging findings have been documented. The author conducted a physical evaluation, radiological examination, MRI finding, pathological feature and operative finding as well as in a woman with idiopathic fibrosis of the quadriceps muscle.

Case Report

A 28-year-old woman presented with a history of inability to flex both her knees fully. She was born prematurely by normal delivery and had been admitted in the hospital for 3 months. Her subsequent development was normal. The age of onset of her disability was obscure. She noticed that she had difficulty in squatting with full flexion of knee joints when she was 8 years old. The symptom slowly progressed. The knees were never swollen, warm or painful. The patient had no history of local injections or trauma to the thighs. She had no family history with a similar abnormality.

Physical examination revealed limitation of flexion of both knee joints. In supine position, the flexion of left and right knee was 0-40 degrees and 0-80 degrees respectively as well as in prone position (Fig. 1a, b, c, d). The hips were not forced to flex when both knees were forced beyond the restricted point (Fig. 1b, d). Movement was not limited by pain and even forcible attempts of flexion were not painful. Left patella was higher than the right but there was no evidence of patellar dislocation. Point of tender or

Correspondence to: Suksathien R, Department of Rehabilitation Medicine, Maharat Nakhon Ratchasima Hospital, Chang Phueak Rd, Mueang District, Nakhon Ratchasima Province 30000, Thailand. Phone: 044-235-135, Fax: 044-246-389, E-mail: rachawan.rm@mnrh.in.th

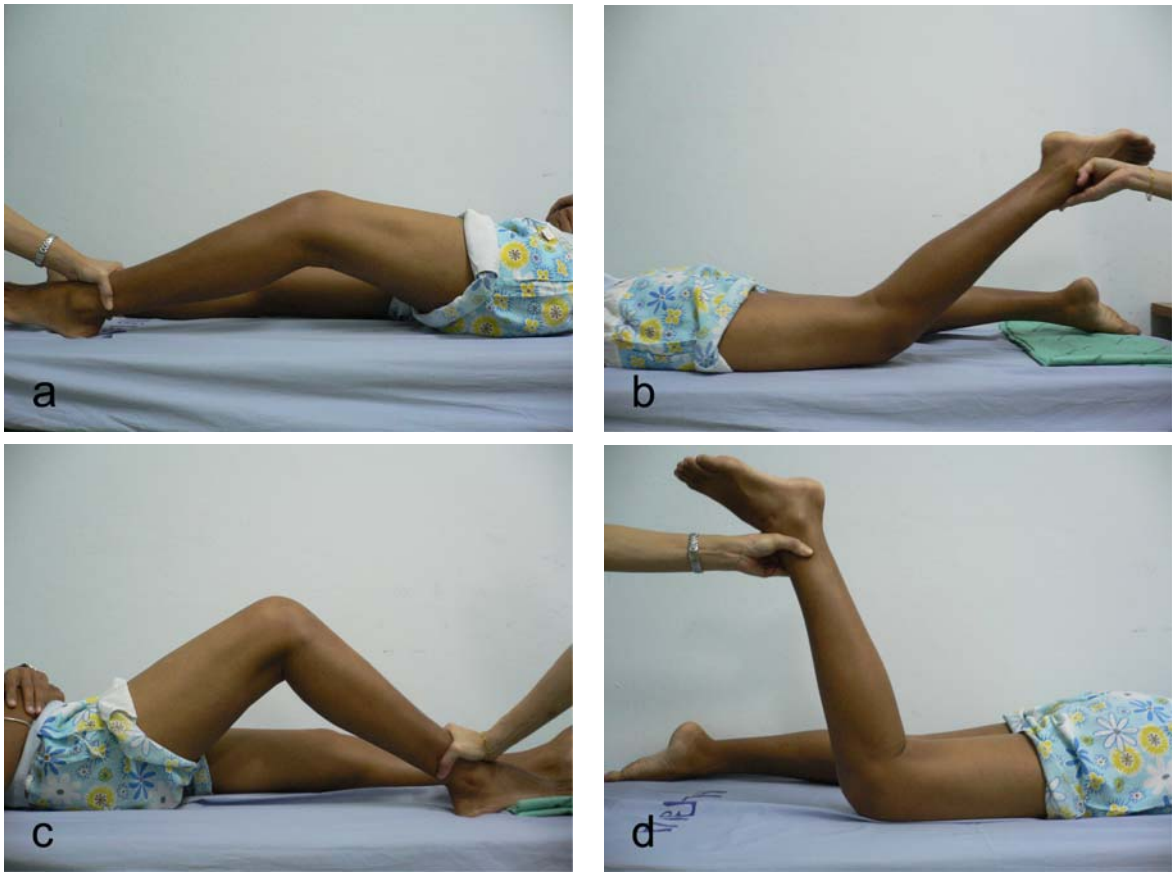

Fig. 1 Passive range of motion of both knees in supine and prone position showed flexion were limited to 40 degrees of left knee (a, b) and 80 degrees of right knee (c, d). The hips were not forced to flex when both knees were forced beyond the restricted point

palpable corded induration was not evident in both thighs. There was no scar over the skin and any neurological abnormality.

Radiographs of the knee joints were normal except the high position of the left patella and flattening of the lateral femoral condyles (Fig. 2a, b, c). Magnetic resonance imaging of the thighs demonstrated fatty atrophy of the vastus lateralis with a thick fibrous structure anterolaterally located within the muscles. These findings were more prominent in the left thigh (Fig. 3a, b).

After the diagnosis of idiopathic fibrosis of the quadriceps muscle was established the patient was treated with intensive physical therapy and static progressive knee splint but there was no improvement. The operation of the left quadriceps was performed after the failure of 4 months conservative treatment and functional limitation in daily activities persisted.

At the operation, the range of flexion was not increased under anesthesia. The rectus femoris and vastus medialis muscles were normal. There were fibrous bands at the vastus intermedius and vastus lateralis muscles that limited full flexion of the left knee (Fig. 4). Z-plasty of the vastus intermedius muscle was done together with release of vastus lateralis muscle and lateral parapatellar ligament. Flexion of left knee was 0 to 110 degrees after the operation. Cylindrical slab was placed to maintain knee in maximum flexion. The physical therapy program and continuous passive motion machine were started on the 3rd day postoperative. Five months after the operation, flexion of left knee was 110 degrees with no extension lag. This knee motion was sufficient for functional movement and the patient reported that her activity was improved.

Histological findings showed the diameter of muscle fibers varied in size. The individual muscle

Fig. 2 Plain film of both knees AP (a) and lateral (b, c) show left patella placed higher than the right and flattening of lateral femoral condyles. Other findings are normal

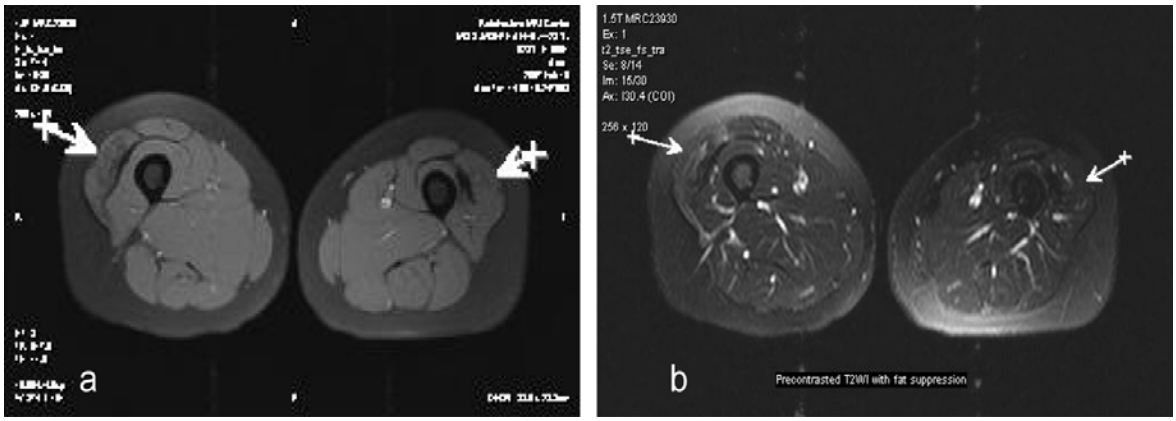

Fig. 3 Axial MRI of the thighs showing the T1-weighted (a) and the T2-weighted (b) image with fatty atrophy of the vastus lateralis muscles associated with fibrous structure anterolaterally

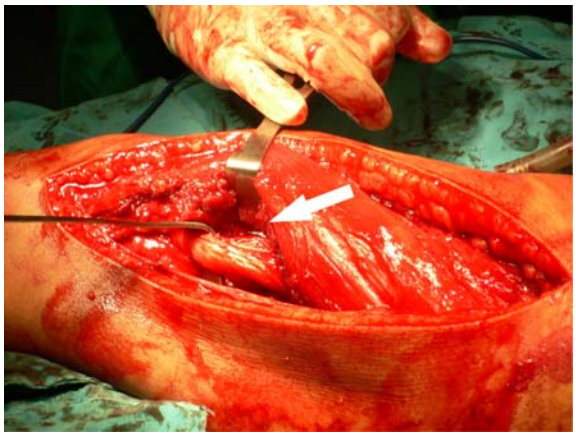

Fig. 4 Operative findings showed fibrous band at vastus intermedius muscle

fibers were separated by loose connective tissue and fat. Large fibrous band was presented (Fig. 5a, b).

Discussion

Idiopathic fibrosis or progressive contracture of the quadriceps muscle is a condition in which extension contracture of the knee develops in early childhood due to fibrosis of one or more components of the quadriceps muscle. The exact cause of fibrosis is not known. Hnevkovsky[1] first described this condition as a muscle dysplasia of congenital origin. Fairbank and Barrett[2] reported identical twins with quadriceps contractures. They believed these contractures were congenital and genetic in origin. Karlen and Chiu[3,4] also postulated this condition as a congenital disease. On the other hand Lloyd-Robert and Gunn[5,6] reported

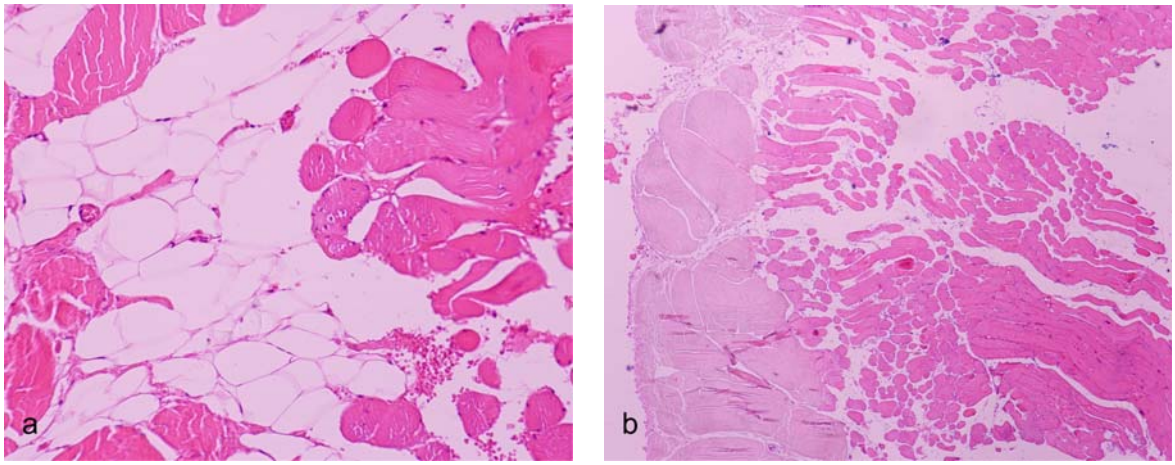

Fig 5 The histological findings of left vastus lateralis and intermedius muscles fixed in 10% formalin and stained by Hematoxylin and Eosin. Sections show the diameter of muscle fibers varies in sizes, separating individual muscle fibers by loose connective tissue and fat (a). Large fibrous band is present (b)

quadriceps contractures in infants resulting from multiple injections or infusions into the thigh soon after birth. Subsequently, quadriceps contracture as a complication of multiple intramuscular injections has been reported with increasing frequency[7-9]. Nearly all affected children had a history of serious illness during early infancy. This patient had no history of injection into thighs but she had been born prematurely with a history of admission in the hospital for 3 months. Most patients in the previous reports were Eastern European[1,2,13], Chinese[3,7], Japanese[4,14] and Thai[16]. This suggested that the incidence of the disease is high in mongoloid races[14].

The pathophysiology of progressive fibrosis is unclear, but suggested causes include compression of the muscle bundles and capillaries by the volume of medication injected and the toxicity of the drug[9-11].

Magnetic resonance imaging (MRI) demonstrated quadriceps muscles were diminished in volume, fatty atrophy and muscle had been replaced by fibrosis. There were fibrous bands at the vastus intermedius and vastus lateralis muscles in gross pathology and loose connective tissue separating the muscle fibers with fibrous band in histology correlated with the MRI. The MRI in the present study revealed a nearly the same result as the previous studies[14,15]. However, the previous studies did not include operative and pathological findings. Therefore, this is the first report to include MRI and pathological findings of idiopathic fibrosis of the quadriceps muscle.

Quadriceps contracture was classified into 3 types by the Ad Hoc Committee of the Japanese Orthopaedics Association for muscular contracture: rectus femoris type, vastus type, and mixed type[12]. According to this classification the patient's physical examination and operative findings were consistent with the vastus type. The vastus type was quite rare from Ad Hoc Committee[12] while many studies reported the vastus intermedius was most commonly involved[8,11].

The clinical hallmark of idiopathic fibrosis of the quadriceps is progressive and painless limitation of knee flexion. This condition may be unilateral or bilateral. The skin dimple may be present over the area of fibrosis especially when the knee is flexed. In long standing cases there is flattening of the femoral condyles, genu recurvatum, anterior dislocation of the tibia, gross degeneration changes in the joint and displacement and hypoplasia of the patella[1,3,4,7,10,11]. The presented patient had delayed diagnosis and treatment so the contractures had progressed to adulthood. She had a high position of the left patella and flattening of bilateral lateral femoral condyles.

Physical therapy is often prescribed initially, but it rarely improves knee flexion significantly[8,11]. Once the scar contracture is well established, surgical treatment is indicated to prevent late changes in the femoral condyles and the patella. The optimal age for surgery is 6 years or older in order to have the child's cooperation in active physical therapy[10,13]. A previous report recommended surgical treatment if the knee flexion is limited to 45 degrees or less in the supine position for vastus type and 30 degrees or less in prone position for rectus femoris and mixed types. This

patient had the operation because of marked limitation of function and the knee flexion was less than 45 degrees. The result of the operation was good. She had smooth gait and could sit comfortably.

Conclusion

This is the first report of idiopathic fibrosis of the quadriceps muscle with MRI and pathological findings. These revealed fibrous band and replacement of muscle by adipose tissue. The MRI will be helpful in confirming the clinical diagnosis as well as demonstrating the extent of fibrosis before surgery. This rare condition of idiopathic fibrosis of the quadriceps muscle should be kept in mind. The most satisfactory results of treatment were obtained if it is performed at an earlier age before the irreversible changes of femoral condyles and patella occur. Prevention from repetitive injection to the same area of the muscle[16], early diagnosis and treatment can help to prevent and decrease disability.

Acknowledgements

The author wishes to thank Dr. Panithan Visalsawadi, Department of Anatomical Pathology, Maharat Nakhon Ratchasima Hospital, and Dr. Yingyong Suksathien, Department of Orthopaedics Surgery, Maharat Nakhon Ratchasima Hospital for their help with the pathological review and operation.

References

1. Hnevkovsky O. Progressive fibrosis of the vastus intermedius muscle in children. J Bone Joint Surg Br 1961; 43: 318-25.
2. Fairbank TJ, Barrett AM. Vastus intermedius contracture in early childhood. J Bone Joint Surg Br 1961; 43: 326-34.
3. Karlen A. Congenital fibrosis of the vastus intermedius muscle. J Bone Joint Surg Br 1964; 46: 488-91.
4. Chiu SS, Furuya K, Arai T, Nakagawa M, Tida M. Congenital contracture of the quadriceps muscle. Four case reports in identical twins. J Bone Joint Surg Am 1974; 56: 1054-8.
5. Lloyd-Robert GC, Thomas TG. The etiology of quadriceps contracture in children. J Bone Joint Surg Br 1964; 46: 498-517.
6. Gunn DR. Contracture of the quadriceps muscle. A discussion on the etiology and relationship to recurrent dislocation of the patella. J Bone Joint Surg Br 1964; 46: 492-7.
7. Euliano J. Fibrosis of the quadriceps mechanism in children. Clin Orthop Relat Res 1970; 70: 181-6.
8. Stark WA. Quadriceps contracture in children. Am J Dis Child 1970; 120: 349.
9. McCloskey JR, Chung MK. Quadriceps contracture as a result of multiple intramuscular injection. Am J Dis Child 1977; 131: 416-7.
10. Diabach JA. Non traumatic soft tissue disorder. In: Canale ST, editor. Campbell's operative orthopaedics. 10th ed. Philadelphia: Mosby; 2003: 885-8.
11. Birch JG, Carter PR, Cummings DR, Damsey ME, Ezaki M, Frobish CE, et al. Muscle disease. In: Herring JA, editor. Tachdjian's pediatric orthopaedics. 3rd ed. Philadelphia: W.B. Saunders; 2002: 1499-500.
12. Sano S, Kokubun S. Report of the diagnosis and treatment of muscular contracture - the Ad Hoc Committee of the Japanese Orthopaedics Association of Muscular Contracture. J Jpn Orthop Ass 1985; 59: 223-53.
13. Gammie WFP, Taylor JH, Urich H. Contracture of the vastus intermedius in children. A report of two cases. J Bone Joint Surg Br 1963; 45: 370-5.
14. Nozawa S, Tanaka C, Shikata J, Yamamuro T. Congenital contracture of the quadriceps muscle: a case report with magnetic resonance imaging. Arch Orthop Trauma Surg 2004; 124: 272-4.
15. Ozdemir O, Atalay A, Celiker R, Kerimoglu U, Ozdemir O. Congenital contracture of the quadriceps muscle: confirming the diagnosis with magnetic resonance imaging. Joint Bone Spine 2006; 73: 554-6.
16. Sudasna S, Pornputtkul C. Quadriceps contracture, a complication of repeated intramuscular injections in children: report of 4 cases. Chiang Mai Med Bull 1974; 13: 347-58.

ภาวะกล้ามเนื้อ quadriceps หดรั้งจากพังผืด: รายงานผู้ป่วยโดยมีผลเอกซ์เรย์คลื่นแม่เหล็กไฟฟ้าและผลทางพยาธิวิทยา

รัชวรรณ สุขเสถียร

***ภูมิหลัง**: กล้ามเนื้อ quadriceps หดรั้งจากพังผืดเป็นภาวะที่มีการยึดแข็งของกล้ามเนื้อ quadriceps ทำให้เกิดการจำกัดการงอเข่าซึ่งเกิดตั้งแต่วัยเด็ก ในต่างประเทศมีรายงานเพียงไม่กี่ฉบับ และยังไม่เคยมีรายงานผลตรวจเอกซ์เรย์คลื่นแม่เหล็กไฟฟ้าร่วมกับผลทางพยาธิวิทยา*

***วัสดุและวิธีการ**: ผู้ป่วยหญิงอายุ 28 ปี มีอาการงอเข่าไม่สุดและนั่งลำบาก รายงานผู้ป่วยพร้อมผลการตรวจร่างกาย การตรวจทางรังสี เอกซเรย์คลื่นแม่เหล็กไฟฟ้า การรักษาและผลทางพยาธิวิทยา*

***ผลการศึกษา**: ผู้ป่วยมีการจำกัดการงอเข่าทั้ง 2 ข้าง เอกซเรย์คลื่นแม่เหล็กไฟฟ้าบริเวณต้นขาพบกล้ามเนื้อ quadriceps ลีบ มีไขมันแทรกและพังผืดในกล้ามเนื้อ ผ่าตัดพบพังผืดที่กล้ามเนื้อ vastus intermedius และ vastus lateralis ซึ่งสัมพันธ์กับผลเอกซเรย์คลื่นแม่เหล็กไฟฟ้า ผลทางจุลพยาธิวิทยาพบเซลล์ไขมันแทรกระหว่างใยกล้ามเนื้อและพบพังผืด ผู้ป่วยได้รับการผ่าตัดยืดกล้ามเนื้อที่เป็นพังผืดซึ่งผลการรักษาเป็นที่น่าพอใจ*

***สรุป**: รายงานนี้เป็นรายงานแรกที่มีผลเอกซ์เรย์คลื่นแม่เหล็กไฟฟ้าและผลทางพยาธิวิทยาของภาวะกล้ามเนื้อ quadriceps หดรั้งจากพังผืด การวินิจฉัยและการรักษาที่ล่าช้าทำให้เกิดพยาธิสภาพที่กระดูกแบบถาวร ประวัติการตรวจร่างกาย การตรวจทางรังสีวิทยารวมถึงการตรวจเอกซ์เรย์คลื่นแม่เหล็กไฟฟ้าช่วยในการวินิจฉัย และประเมินบริเวณที่เกิดพังผืดก่อนการผ่าตัด*