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Treatment of Achilles tendon calcinosis in juvenile dermatomyositis with external ilizarov fixator

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ABSTRACT

Calcinosis is a devastating complication of juvenile dermatomyositis and a challenging therapeutic problem. We report the use of an external Ilizarov fixator for the treatment of Achilles tendon calcinosis causing severe disability in a young girl with juvenile dermatomyositis.

Introduction

Calcinosis of the skin or muscle occurs in up to 40% of children with dermatomyositis and poses a challenging therapeutic problem. It may be prevented by aggressive early treatment of the myositis, but once established, there is no effective therapeutic solution (1). We describe a young girl with juvenile dermatomyositis (JDM) complicated by calcinosis along both Achilles tendons, causing severe contracture and functional disability, especially on the left side. Application of the external Ilizarov fixator successfully alleviated the contracture of the left ankle. This is a report of the use of this orthopedic procedure for calcinosis-induced ankle contracture in JDM.

Case report

A 4-year-old girl presented with a rash and progressive, painful muscle weakness in November 1994. Creatinine kinase level was 12,000 U/L. Findings on electromyography and muscle biopsy confirmed the diagnosis of JDM.

The patient was treated with corticosteroids, methotrexate and hydroxychloroquine, with remarkable improvement of the myositis. Because of persistent rashes, methotrexate was replaced by cyclosporin A.

Calcifications were first noted in March 1996. The calcium deposits increased rapidly and extensively, causing intermittent fever, recurrent cellulitis, venous congestion of the upper body, and impaired gait.

In January 1998, treatment with probenecid was begun, and 5 months later, the first improvement in the calcifications was documented. At the follow-up visit in June 1999, the skin and muscle disease was found to be in remission, and the calcifications were markedly reduced, except in the legs, along the Achilles tendons. As a result, the ankle joint ranges of motion were impaired in both legs, but more severely in the left.

In October 1999, the left ankle was fixed at a plantar flexion of -30°. Nine months later, it was fixed at a plantar flexion of -40°, and the girl was practically walking on tiptoe. In August 2000, lithotherapy was applied to both ankles but failed to yield any improvement. In additional, neither the calcinosis nor the contracture worsened as a result of the lithotherapy. By June 2001, the left heel was fixed at a plantar flexion of -70°, with no movement. Soon thereafter the patient's condition worsened: the left ankle joint was completely stiff with an 80° equino-deformity (Fig. 1). At this point the patient had x-ray evidence of moderate osteoporosis, however, a DXA scan was not done.

At that point, we decided to use the Ilizarov circular frame to the left ankle. At the time that the surgical treatment was used, the JDM was not active ant the patient received probenecid, plaquenil and caltrate+D. The first stage of surgical treatment was performed in August 2002 and consisted of Achilles tendon elongation with removal of the calcium deposits and posterior capsulotomy. No calcium deposits remained following the surgical debridement. A circular frame with two rings was then assembled on the patient's leg and connected to a foot frame. The hinges were placed in the center of rotation of the ankle joint, and a wire was inserted to the talus in order to control and avoid subluxation of the joint during correction. The correction was completed after one month, and the device was kept in position for another 4 months (Fig. 2). On removal of the frame, a plaster of Paris cast was placed to secure the correction. Examination revealed a stable, fixed, plantigrade ankle. At her last visit (May 2006), after almost 4 years of follow up neither the calcinosis nor the ankle contracture recurred (Fig. 3).

Discussion

The occurrence of extensive calcinosis in JDM is usually associated with a severe and unremitting disease course. In these cases, the calcinosis may be responsible for more long-term disability

than the residual effects of the acute myositis (2).

None of the approaches suggested for the treatment of calcinosis have proved consistently effective. This is true for the many medications attempted, such as colchicine, aluminum hydroxide, probenecid, bisphosphonate, diltiazem, intravenous ethylenediamine tetra-acetic acid (EDTA) and warfarin (2). Surgical intervention is usually contraindicated because of the risk for secondary calcinosis deposition. In our patient, the extensive calcinosis was treated successfully with probenecid, except for the deposits along the Achilles tendons (3). The deposits also failed to respond to repeated courses of lithotherapy, resulting in severe ankle contracture and significant function impairment, necessiting surgical interference.

The Ilizarov method is based on the biological concept of distraction osteogenesis (4). New local bone formation is induced mechanically by corticotomy wherein the two bone edges are pulled apart and stabilized with an externally fixated ring (4). The joint distraction transfers the forces from the bone to the soft tissues; the tendons, ligaments and capsular structures, which are composed of collagen slowly accommodate to the new position (5).

This method is applicable for bone elongation, bone transport to replace bone loss, correction of nonunion fractures, treatment of osteomyelitis, and correction of long bone and joint deformities (4). As in the present case, the circular foot frame permitted stable fixation of the tibia segment near the ankle joint. By correct hinge placement, distraction and angulation adjustments, it is possible to realign the abnormal joint by controlled gradual correction (5).

In 1997, Correll and Trunckenbrodt (6) published the first and only report to date of the use of the Ilizarov method to treat rheumatic conditions in children. Their series included 2 children with scleroderma, one child with rheumatoid arthritis, and 3 children with dermatomyositis. All presented with severe stiff clubfoot, leg-length discrepancy, and flexion contracture or deformity of the hips and knees. Conservative management had failed, and 4 of the 6 children were wheelchair-bound. The 3 children with dermatomyositis also had severe soft tissue calcifications and were operated by a one-step procedure: tenotomy of the medial hamstrings and the biceps femoris muscle, followed by placement of the circular external fixator to the femur and lower leg. No cor-

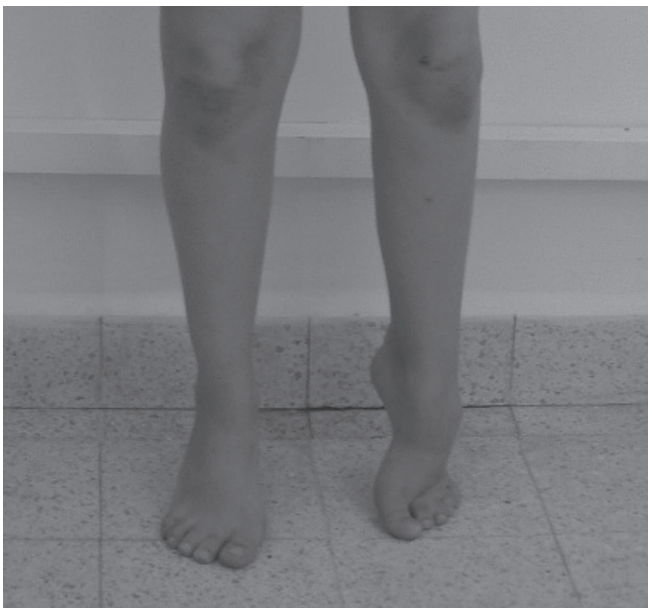

Fig. 1. June 2001: left ankle at severe equino deformity.

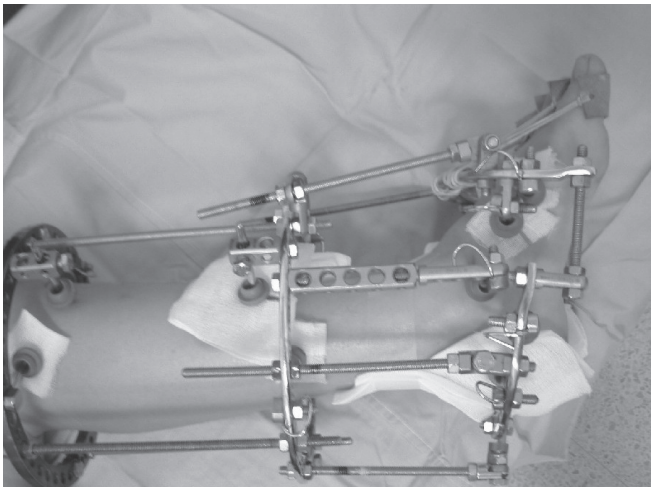

Fig. 2. August 2002: push & pull construct with hinges in the center of rotation of the ankle.

Fig. 3. At follow up: left knee after correction.

rection was performed intraoperatively (as in our case). Gradual correction in all cases was begun after wound healing. One of the patients with dermatomyositis fell at home while wearing the external fixator and broke her femur. She was treated by extending the external fixator around the fracture, and it healed without any other problems. No other major complications were reported in the children with dermatomyositis (6).

Ours is the second report of the application of the Ilizarov device in a child with rheumatic disease and the first to describe its use specifically for the treatment of Achilles tendon calcinosis. Our patient had a completely stiff ankle joint with an 80° equino-deformity.

The Ilizarov method has several advantages, including minimal soft-tissue injury and gradual correction without the use of anesthetics (7). However, it also has some disadvantages: a long healing time, sometimes with chronic pain, resulting in significant psychological stress for the child and parents; risk of nerve or major vessel injury; and risk of pin-site infection. The complication rate varies from 14% to 134% (more than one complication per patient), depending mostly on the type of pathology corrected (7). There are also special concerns regarding the use of the Ilizarov method in children with JDM. First, the procedure should be used with caution in JDM patients with active disease or progressive calcinosis or in which there is an underlying osteoporosis. However, the risk for increased calcinosis following surgical procedures exists, yet reduced using the Ilizarov method due to minimal soft-tissue dissection and gradual postoperative correction adjustment. Second, careful patient (and surgeon) selection is very important to ensure success (8). Third, the risk of anesthesia in some rheumatic diseases, including JDM, is high (9, 10).

In conclusion, we describe, for the first time, the use of an external Ilizarov fixator to treat disabling Achilles tendon calcinosis in a young patient with JDM. The treatment proved to be safe and long-lasting. We believe the procedure might also be applicable to severe flexion contractures associated with JDM that are unrelated to calcinosis. Successful application of the Ilizarov method in such cases requires close cooperation of the patient and family as well as an intimate familiarity of the surgeon with the device, the technique, and the postoperative management protocol.

References

1. CALLEN JP: Dermatomyositis. *Lancet* 2000; 355; 53-7.
2. CASSIDY JT, PETTY RE: Juvenile dermatomyositis. *In* CASSIDY JE, PETTY RE (Eds.): *Textbook of Pediatric Rheumatology*. 4th edition. WB Saunders, Philadelphia, 2001: 465-504.
3. HAREL L, HAREL G, KORENREICH L, STRAUSSBERG R, AMIR J: Treatment of calcinosis in juvenile dermatomyositis with probenecid: the role of phosphorus metabolism in the development of calcifications. *J Rheumatol* 2001; 28: 1129-32.
4. PALEY D: Principles of foot deformity correction: Ilizarov technique. *In* GOULD JS (Ed.): *Operative Foot Surgery*. WB Saunders, Philadelphia, 1994, p. 476-514.
5. ARONSON J, GOOD B, STEWART C, HARRISON B, HARP J: Preliminary studies of mineralization during distraction osteogenesis. *Clin Orthop Relat Res* 1990; 250: 43-9.
6. CORRELL J, TRUCKENBRODT H: Correction of severe joint contractures and leg length discrepancies by external fixation (Ilizarov method). First results in children with rheumatic diseases. *Rev Rhum Engl Ed* 1997; 64 (10 Suppl.): 163S-166S.
7. BIRCH JG, SAMCHUKOV ML: Use of the Ilizarov method to correct lower limb deformities in children and adolescents. *J Am Acad Orthop Surg* 2004; 12: 144-54.
8. SLOMKA R: Complications of ring fixators in the foot and ankle. *Clin Orthop Relat Res* 2001; 391: 115-22.
9. KHAN FA, ANJUM I, KAMAL RS: Anaesthetic hazards in dermatomyositis. *J Pak Med Assoc 1991*; 43: 69-71.
10. YAZIGI A, YAZBECK P, ANTAKLY MC: Problems posed by a patient with dermatomyositis undergoing general anesthesia. *Cah Anesthesiol* 1991; 39: 557-8.