Congenital Longitudinal Deficiency of the Tibia
A Case Report

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ABSTRACT

A 3-month old male with congenital longitudinal deficiency of the right tibia had been treated in our department. There was no family history. There was also no history of taking drugs or traditional herbal medicine during pregnancy. Based on clinical and radiographic evidence, the case is congenital deficiency of the tibial type II. The patient was operated with tibio-fibular synostosis combined with osteotomy correction of the fibula, fusion of the distal fibular to the calcaneus and bone grafting at the gap of the distal tibia.

Keywords: congenital deficiency of the tibia, varus foot, ankle reconstruction

INTRODUCTION

Congenital aplasia or deficiency of the tibia is a rare condition characterized by various degrees of tibial deficiency. According to the literature, in two-third of cases it is usually combined with other anomalies. The purpose of this paper is to present a case with congenital longitudinal of the tibia anomaly.

CASE REPORT

The parents came to our department with the complaint of deformity of the right lower extremity in their male child of 3 months old. There is no maternal disease and no history of taking drugs or traditional herbs during pregnancy. The delivery had been full term and normal. There was no family history of congenital anomaly.

On physical examination, the condition of the child is good. ROMs of the hips were normal but the affected knee is in 5 degrees of flexion contracture. The quadriceps powers of the patient were normal. The discrepancy of the right extremity was 0.5 cm short according to Choi et al method.[1] The medial side of the foot was curved inwards and it can manipulate in plantigrade position. The number of rays in the foot are normal but the first web space is widened compared with the normal foot. The second digit is laterally deviated to the dorsum of the third digit (Figure 1).

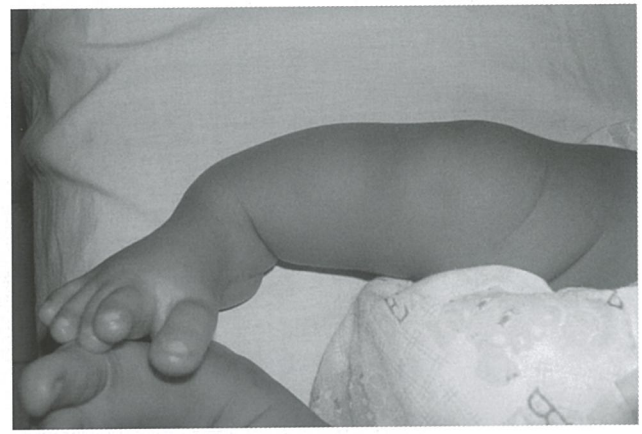

Figure 1. Clinical appearance at the time consultation (3 months old)

Our diagnosis during consultation was varus deformity of the right foot. Our management was serial plaster casting. After serial plaster casting on two occasions, the result of treatment was not successful. Radiographic films taken showed the diameter of the distal femur to be widened but the proximal portion of the fibular head is slightly displaced proximally. The x-rays showed aplasia of the distal tibia and medial bowing of the fibula at the end level of the tibia. The talus is in normal position (Figure 2).

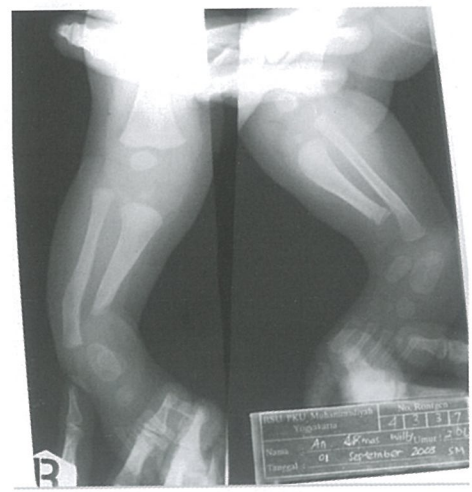

Figure 2. Radiographic data at 3.5 months old (AP and lateral views)

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The problems encountered by this patient included medial angulation of the fibula with aplasia of the distal part of the tibia and instability of the ankle with incurving of the foot medially.

The goals of the treatment are to preserve the length and alignment of the affected right lower leg, restore stability of the ankle and to maintain a plantigrade position of the right foot. The plan was to use serial long leg plaster cast in plantigrade position of the foot until the child was six months old. Correction of the medial angulation of the fibula was achieved by osteotomy at the level of the tibial end to achieve a normal lower leg alignment combined with synostosis of the proximal tibio-fibular through a lateral incision of the lower leg. Fibulo-calcaneal fusion was achieved through a small incision behind the lateral malleolus without damage to the distal physis of the fibula and iliac crest bone grafting in the gap of the distal tibia was performed with grafts from the apophysis of the iliac through a small anteromedial incision of the distal lower leg and fixed by double K-wires through the calcaneo-talus to the distal tibia. After the operation, the foot was held plantigrade in an above knee plaster cast. The K-wires will be removed after six weeks and plaster cast immobilization will be continued for three months.

DISCUSSION

Congenital longitudinal deficiency of the tibia is an extremely rare condition with the incidence of about one per million live birth. According to Kalamchi and Dawe publication, in two-thirds of the affected cases there are anomalies in the hand (43%), the male gonads (43%), other foot (39%), in the ipsilateral femur (29%), hernia (19%), congenital heart diseases (14%), learning disabilities (14%) and scoliosis (9.5%). This deformity is of autosomal dominant inheritance. Based on clinical and radiographic findings, this anomaly is divided into three types.[2, 3, 4, 5, 6]

There are two classifications of tibial deficiency. The first is Jones Classification (1978) which is divided into types I (a & b), type II, type III and type IV. The last classification has been made by Kalamchi and Dawe (1985)[2]. The latter is used routinely because this classification is very simple to implement. It included type I, the foot clinically is in marked inversion and adduction and the medial rays are absence infrequently, the knee joint in flexion contracture with palpation of the fibular head is in proximal displacement. The quadriceps power is very weak or absent. On x-rays, there is total absence of the tibia with proximal migration or dislocation of the head of the fibula and marked hypoplasia of the distal femur. This hypoplasia of the femur includes reduction in the width of the distal metaphysis and marked retardation of the ossification of the distal epiphysis. According to Jones Classification, it is type Ia. There is less marked flexion contracture on the knee. The radiographic films showed that the distal end of the tibia is absent. Sometimes the proximal tibia is not visible because it is cartilagenous bone but the normal presence of the distal femur and ossification of the epiphysis means there is a part of proximal tibia. According to Jones Classification, it is type Ib and II. Tuli and Varma (1972) reported that the condition is caused by localized distal tibial epiphyseal growth disturbance but Bose (1976) wrote that deformity is caused secondary to congenital hypoplasia of the distal tibia, talus and hypoplasia of the medial ray of the foot.[2, 6, 7, 8, 9]

Management of the longitudinal tibial anomaly depends on the condition of the femur, quadriceps muscle power, knee joint, tibia and fibula, ankle and foot. There are many procedures that had been published in the literature, for example fibulo-femoral arthrodesis or arthroplasty (Brown procedure with centralization of fibula), fibulo-tibial proximal fusion, central fibulo-talus fusion, above knee, lower leg, and Syme's or Boyd's amputation. Frequently, one or more additional procedure was needed to achieve ambulation.[2, 4, 9, 10, 11, 12]

According to the clinical and radiographic data, our case is type II of congenital longitudinal tibia anomaly. Therefore, our management for this case is to preserve the normal length of the lower leg by fusion of the proximal tibio-fibular joint and fibulo-calcaneal fusion without damage to the center of fibula development and stabilization of the ankle with the iliac crest grafting at the distal tibia.

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