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CASE REPORT

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Ray amputation for the treatment of macrodactyly in the foot: report of three cases

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Foot macrodactyly is a rare congenital anomaly which is characterized by an overgrowth of the soft tissue and bone of the toes. The aim of treatment is to obtain a cosmetic and functional foot. We present three cases of lesser toe macrodactyly on which we performed ray amputation. Postoperative cosmetic and functional results were good in three cases. Ray amputation is a possible surgical treatment that provides good cosmetic and functional results in severe lesser toe macrodactyly.

Key words: Foot; macrodactyly; ray amputation.

Macrodactyly is a rare congenital deformity characterized by hypertrophy of the bones and surrounding soft tissues in one or more digits.[1-4] It may be both isolated or accompanied by different abnormalities including neurofibromatosis, hemangiomatosis, arteriovenous malformations, congenital lymphedema, and Klippel-Trenaunay-Weber and Proteus syndromes.[3,5-8] There are two types of macrodactyly: static and progressive.[8,9] Etiology remains controversial although changes in peripheral nerves are suspected.[1,3] The aim of operative treatment is to obtain a pain-free and functional foot. Suggested procedures include digit amputation, epiphysiodesis, and debulking of soft tissues accompanied by total or segmental resection of distal phalanx and ray amputation.[1,2,4,5,10-12] However, in procedures other than ray amputation, no viable results are obtained, necessitating repetitive operations.[4,5,8,11,13]

In this study, we present the cosmetic and functional results of three patients who underwent ray amputation due to macrodactyly in the foot.

Case reports

Three patients suffering from pain and the inability to wear footwear due to congenital overgrowth of toes, were admitted to our department between 2004 and 2009. The patients were 3, 6, and 10 years old, including two males and one female. Anteroposterior and lateral radiographs were obtained for both feet of each patient. Macrodactyly was confined to the right foot in all patients. The involved digits were the second toe in the first case, the third and fourth toes in the second case, and the third toe in the third case. Macrodactyly was progressive in the three-year-old patient, and static in the other two. The third patient had hereditary multiple osteochondromatosis. In addition, the second patient underwent a surgical operation to debulk the third and fourth toes, yet, recurrence developed as a result of the progressive enlargement of the third toe.

All operations were performed by the same surgeon (LK) under general anesthesia. A complete excision of the skin and subcutaneous tissues was performed on the dorsal and plantar surface of the foot, through a triangular incision, whose top was crested at the tarsometatarsal joint. Disarticulation was achieved through complete removal of the metatarsals beginning from the tarsometatarsal joint. Segmental excision was performed for the hypertrophic fibroadipose tissue among the metatarsals. Primary closure was

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performed. A short leg cast was applied with medial and lateral compression. Patients were provided a subcalcaneal pad and allowed weight bearing as tolerated two weeks following surgery. Casts were removed at the end of the sixth week.

Case 1

A ten-year-old girl presented to our department with complaints of pain and inability to wear footwear due to abnormal growth of the third and fourth digits in the right foot. Segmental resection of two phalanxes and debulking of soft tissues were achieved. As the third digit continued to enlarge and the complaints persisted, ray amputation was performed on the third digit in the third year. A short leg cast was applied with medial and lateral compression. The patient had acceptable cosmetic and functional result in the final examination at the first postoperative year. Radiology detected a small space among the metatarsals that underwent the ray amputation (Fig. 1). The patient's only complaint remains a mild pain triggered with long-distance walks.

Case 2

A six-year-old boy was admitted to our department with complaints of pain and inability to wear footwear due to congenital hypertrophy of the third digit. Radiological examinations revealed accompanying multiple hereditary exostoses. Ray amputation was performed on the third digit. Excision was performed for the calcaneal exostosis. In the fourth year of follow-up, the patient revealed good cosmetic and functional results. Radiology confirmed the intermetatarsal space to be completely closed. A recurrent exostosis was detected in the plantar surface of the first metatarsal and in the calcaneus. A second surgery was not performed as the patient had no complaints (Fig. 2).

Case 3

A three-year-old boy applied to our department with complaints of overgrowth in the second digit of the right foot and inability to wear footwear. The parents stated that the second digit was enlarging much

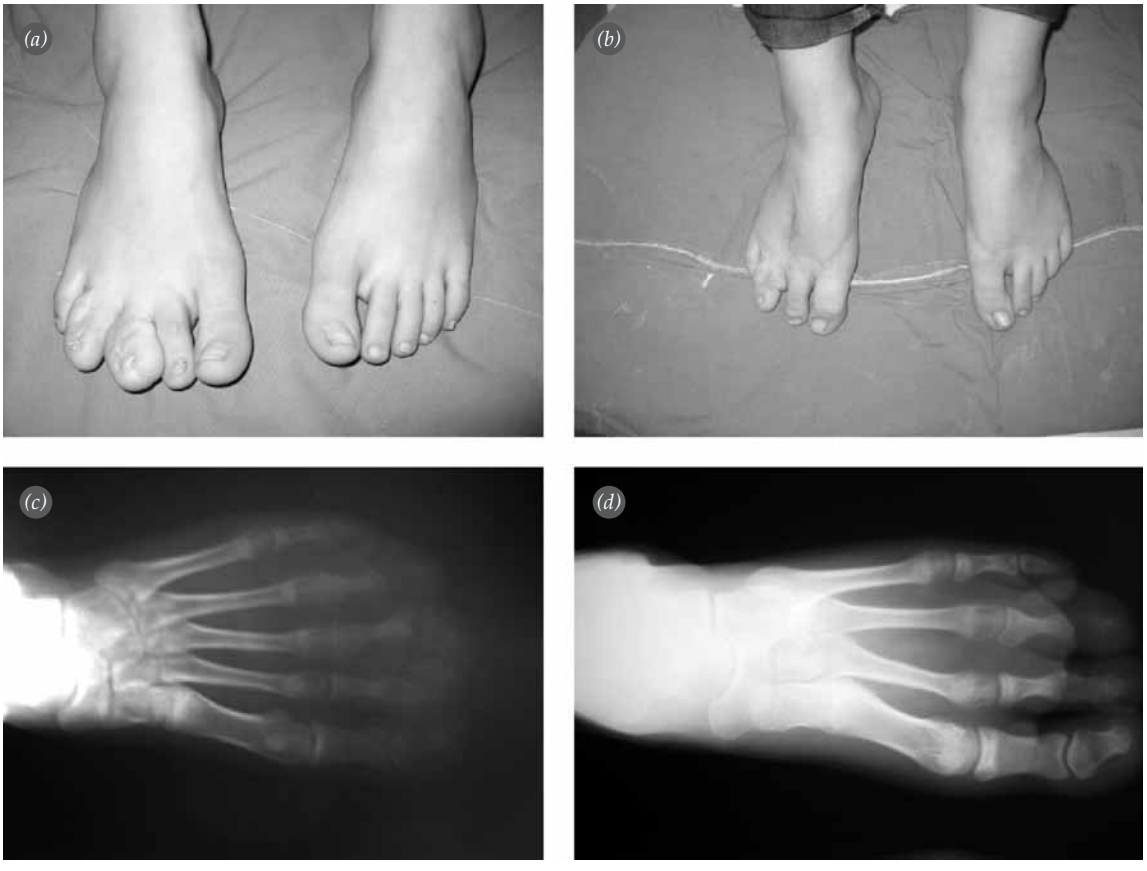

Fig. 1. Case 1. **(a)** Preoperative view of the foot, **(b)** postoperative view of the foot in the first year, **(c)** preoperative roentgenogram, and **(d)** postoperative roentgenogram in the first year.

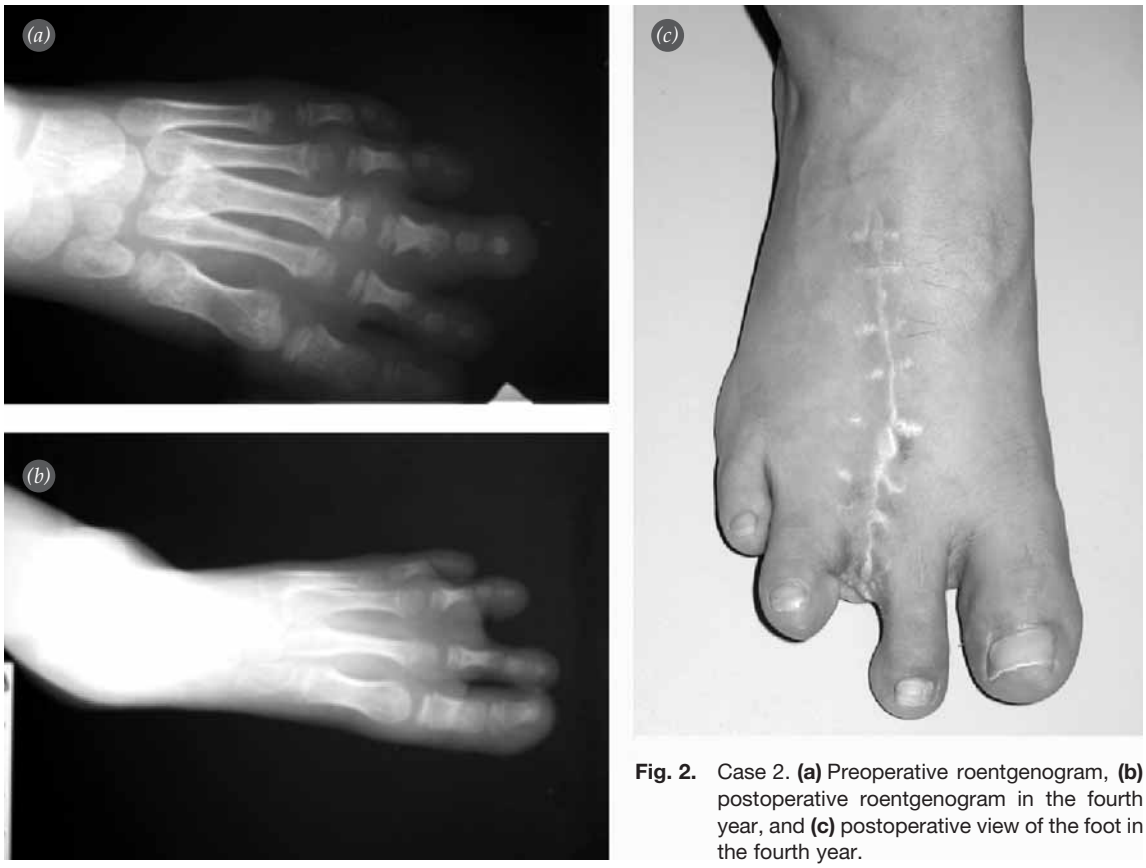

Fig. 2. Case 2. **(a)** Preoperative roentgenogram, **(b)** postoperative roentgenogram in the fourth year, and **(c)** postoperative view of the foot in the fourth year.

faster than the others. Ray amputation was performed as no accompanying pathology was detected. The patient had acceptable cosmetic appearance and satisfactory functional and radiological results in the second year of follow up (Fig. 3).

Postoperatively, no infection or necrosis was observed during wound healing. In all three cases the affected feet were in size to the unaffected feet, and the patients were able to use the same-size footwear for both feet.

Discussion

Macrodactyly is a rare congenital anomaly characterized by the congenital overgrowth of the bones and soft tissues in the fingers and toes. Metacarpals, metatarsals, tendons and veins in the digits are not

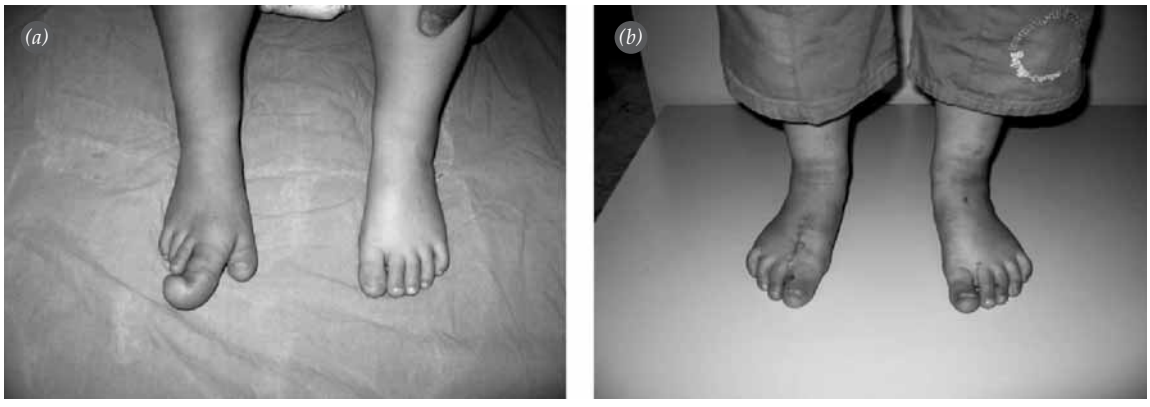

Fig. 3. Case 3. **(a)** Preoperative and **(b)** postoperative views of the foot in the second year.

affected.[4] However, the involvement of metacarpals and metatarsals have also been reported.[1,14,15]

Barsky defined two types of macrodactyly.[9] Static macrodactyly, the most common, is a congenital growth which progresses in proportion with normal growth. Progressive macrodactyly, in which the digit involved has a higher rate of growth, is less common. Wu et al. studied 73 cases (12 static and 61 progressive) in which the most common involvement was in the second toe.[16] Etiological reasons remain controversial although several theories have been stated; the role of heredity has yet to be confirmed.[1,3,14,17,18] Two of our patients had static and one progressive macrodactyly. The role of heredity was not evident in our patients.

In a study of 64 cases, Barsky reported that macrodactyly is more commonly observed in males and in the foot –especially the second digit–.[9] Conversely, Kelikian, in his study of 300 cases, reported that the involvement of the hand is more common than the foot.[14] Another study by Kotwal and Farooque, in which they inspected 23 cases, reported 61% of cases were in females and that the foot was more involved.[2] Our series comprised of two males and one female. Macrodactyly was present in the third and fourth digits in the first case, in the third digit in the second case, and the second digit in the third case.

A number of treatment methods have been suggested for macrodactyly. However, many do not provide satisfactory results, requiring additional procedures. Rechnagel reported 6 cases that required a second operation of ray amputation after undergoing soft tissue debulking.[13] Dedrick and Kling performed soft tissue resection in 9 cases, 4 of which required additional surgeries.[11] Kotwal and Farooque, performed a two-step surgery including the complete removal of phalanxes and defatting of the affected digits of both the hand and foot.[2] Results were good in 12 (57.2%), satisfactory in 7 (33.3%), and poor in 2 (9.5%) cases. The two cases with poor results underwent amputation due to unacceptable cosmetic results. Chang et al. reported a series of 17 patients with macrodactyly of the lesser toes (n=13) and the great toe (n=4).[5] Amputation (n=6), shortening (n=2) and primary or secondary ray amputation (n=5) was performed on the lesser toes, while an epiphysiodesis or local excision was performed for the great toe (n=4). Moreover, patients received soft tissue debulking as a complementary procedure. Results were graded for intermetatarsal angle, pain, and the ability to wear ordinary footwear. Acceptable results were obtained in only three of the 8 patients who underwent amputation and shortening. However, in all 5 cases that received ray amputation –either as a primary or secondary procedure– acceptable results were obtained. Results for the great toes (n=4) were fair and required additional operations.

Ray amputation remains the ideal method for the macrodactyly cases with accompanying metatarsal involvement. Primary advantages are the acceptable cosmetic results and the reduction in the size of the involved digit, which can only be achieved by this procedure.[5,11,19] In all three of our cases, the patient was able to wear the same-size footwear on both feet postoperatively. Therefore, it can be said that ray amputation equalizes –if not completely– the width and length of the affected and unaffected foot. This equalization provides a proper foot with acceptable cosmetic and functional results.

Ray amputation is not suggested initially in macrodactyly of the great toe. A debulking procedure accompanied with/without phalangeal and/or metatarsal shortening and/or epiphysiodesis is preferred, likely because of the important role of the great toe in normal stepping and walking, as well as the unacceptable cosmetic result.[5] In a study on the cosmetic and functional importance of nail preservation, Dautel et al. performed a number of procedures for cases with macrodactyly of the great toe, including resection of the distal phalanx, debulking, and vascularized nail bed transfer.[7] Acceptable outcomes were obtained in all cases with the exception of stiffness in the proximal interphalangeal joint.

Dedrick and Kling recommended complete ray amputation in young patients whose affected foot is two standard deviations away from the normal foot.[11] Similarly, Chang et al. noted that ray amputation is particularly indicated when the intermetatarsal angle is 10 or more degrees greater than normal.[5]

In our first case, we applied resection of two phalanxes and debulking of soft tissues to the third and fourth toes. The third toe was reoperated with ray amputation as the enlargement continued. During the first postoperative examination, the only complaint was a mild pain resulting from excessive walking. This compliant was considered to be largely dependent on the older age of the patient and the shortness of the follow-up period which prevented the complete

closure of the intermetatarsal space. A number of studies hold that the intermetatarsal space resulting from the implementation of ray amputation at younger ages is easier to close.[5] Accordingly, we think that the better results obtained in the other two cases –aged 3 and 6– were primarily dependent on age.

Postoperative complications have been noted in the literature according to the procedures they accompany. Variable rates of infection, necrosis, and stiffness in the proximal interphalangeal joint have been seen, following procedures of epiphysiodesis, segmental or complete resection of the phalanx, digit amputation and debulking. Complications, including hallux valgus and similar angular deformities, scar contraction, and recurrence have been reported with amputation of the second toe.[1,2,5,7] None of these complications were observed in our series.

The relation of the length of the metatarsals must be considered when determining the surgical method. In a case with accompanying metatarsal involvement, an intervention that only involves the phalanx may prevent desirable results. An excessively short metatarsal resulting from epiphysiodesis can also become a serious problem. Our results suggest that ray amputation is an effective single-stage treatment method for cases of advanced or recurrent macrodactyly of the lesser toes that provides acceptable cosmetic and functional results.

Conflicts of Interest: No conflicts declared.

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