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CONGENITAL MACRODACTYLY: A CASE REPORT

Orthopedics

Rahul Anshuman* Senior Resident, Department of Orthopaedics, UCMS and GTB Hospital *Corresponding Author

Aditya Nath Aggarwal Professor, Department of Orthopaedics, UCMS and GTB Hospital

Rohit Pandey Senior Resident, Department of Orthopaedics, UCMS and GTB Hospital

Himanshu Bhayana Senior Resident, Department of Orthopaedics, UCMS and GTB Hospital

Saurabh Kumar Rai Ex Resident, Department of Orthopaedics, UCMS and GTB Hospital

Karan Raj Jaggi Senior Resident, Department of Orthopaedics, UCMS and GTB Hospital

ABSTRACT

Macrodactyly is an increase in the size of one or several fingers or toes. It is characterized by an increase in all mesenchymal elements particularly fibro-adipose tissue. This should be distinguished from secondary digital/limb enlargement that occurs in vascular anomalies or as a result of other syndromes. The accepted treatment is reduction of the fibro-fatty bulk via dissection and ablation to obtain a cosmetic and functional foot.

KEYWORDS

Congenital, Syndactyly, Macrodactyly ; Distal limb

Introduction

Macrodactyly is a rare congenital deformity characterized by hypertrophy of the bones and surrounding soft tissues in one or more digits. There are two types of macrodactyly: static and progressive.

In the static type, the growth rate of the involved digit is the same as the normal digits whereas in the progressive type, growth is accelerated compared to the rest.[1]

Macrodactyly most commonly exists without other conditions but syndactyly is associated with macrodactyly in about 10% of cases.[2]

The aim of operative treatment is to obtain a pain-free and functional foot. Various operations have been described including carpal tunnel release,[3] bulk reduction procedures,[4] stripping of nerves,[5] resection of nerves,[6] epiphyseal arrest,[4] toes shortening,[5] , angulation osteotomy ,ray amputation and amputation of foot[5]

In our case we chose to do disarticulation of great toe with 2nd and 3rd ray amputation and debulking procedure.

Case Report

A two year old girl came to our department with complaints of overgrowth in the right foot and inability to wear footwear. The parents stated that the right foot was enlarging much faster than the other.(Fig1) Radiographically, 2nd metatarsal, first 3 proximal phalanx and 1st distal phalanx was bigger than other metatarsals and phalanges, but normally linked and articulated. First web space was enlarged.(Fig 2)

Disarticulation of great toe with 2ndand 3rdray amputation and debulking was performed. The foot was operated by giving racquet shaped incision over base of great toe extending to the navicular tuberosity on the medial side of foot. Another racquet shaped incision was given over syndactyly going proximally upto great toe and dorsally between 2nd and 3rd metatarsal. Over great toe, flexor and extensor tendon were identified and resected as proximal as possible. Great toe amputation done at the level of 1st metatarsal neck. Digital nerves and vessels were identified. Nerve was resected and vessels were cauterized. 2nd and 3rd syndactyly was removed from the base of 2nd and 3rd metatarsal. Debulking of plantar and dorsal surface was done. Skin flap revised and closure done.

Intraoperatively abundant adipose tissue like material was found more on plantar surface than on the dorsum of foot. Digital nerves were found to be thickened.

Discussion

Macrodactyly represents a rare congenital anomaly characterized by distal limb overgrowth of all tissue elements. This should be distinguished from secondary digital/limb enlargement that occurs in vascular anomalies or as a result of other syndromes. Macrodactyly is defined by an enlargement of all the structures of fingers or toes and characterized by hypertrophy of the bones and surrounding soft tissues in one or more digits.[7-8] Barsky, in a study of 64 cases, reported that macrodactyly is more commonly observed in males and in the foot.[1]

Surgical options mentioned earlier focuses on decreasing the size of the affected foot and allowing the patient to fit in normal shoes. Debulking is the most commonly attempted procedure and as the name suggests it involves bulk reduction of the involved soft tissue. Although a great procedure, used alone it is not sufficient for size reduction and only provides short-term success. Rechnagel reported 6 cases that required a second operation of ray amputation after undergoing soft tissue debulking.[9]

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.[10-12]Many studies hold that the intermetatarsal space resulting from the implementation of ray amputation at younger ages is easier to close.[10]

In our case, the final result is good in terms of outcome (function, form). Function: patient is able to ambulate in normal footwear and carry on all activities of daily living. Form: all wounds healed well and patient's attendants were satisfied with the appearance of foot.

Postoperative complications have been noted in the literature according to the procedures they accompany. Variable rates of infection 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.[8,10,13-14] None of these complications was observed in our case Only complication associated was skin necrosis which was seen on the 5th day.(fig 3)

A B

Fig 1: A and B are Clinical Preoperative Photograph of the affected foot

Fig 2: Preoperative Radiograph

A.

B.

Fig 3: A and B are Post operative clinical photographs at 8th day after operation of the affected foot

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