

A LARGER THAN LIFE TOE TALE: A CASE REPORT ON MACRODACTYLY

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INTRODUCTION

Macroductyly is a rare congenital deformity characterized by hypertrophy of the bones and surrounding soft tissues in one or more digits. The terms “macroductyly”, “megaloductyly”, and “digital” or “localized gigantism” are often used synonymously^[1,2]. The condition seems to affect slightly more boys than girls, and it may occur only on one foot or on both feet, and it may be symmetrical or asymmetrical. Macroductyly may occur as an isolated defect in either the hand or the foot, or it may present as an element of conditions, such as Proteus syndrome, Klippel-Trenaunay syndrome, Bannayan-Riley-Ruvalcaba syndrome, Maffucci, Ollier’s disease and Milroy’s disease. The enlargement includes skin, subcutaneous tissue, nerve, joint, and bone. The bony involvement is usually limited to phalanges^[3]. There are two types of macroductyly: static and progressive. The progressive variant is more frequently seen in the foot than in the hand. Radiologically, X-ray, computed tomography (CT) and magnetic resonance imaging (MRI) are used to determine the size of lesions, involvement of soft tissue and bone, which are important for surgical treatment planning. In macroductyly, reconstruction is aimed at decreasing the size of the foot in normal size and shape as much as possible. Many surgical options are available for the treatment of macroductyly of the foot: amputation, ray amputation, epiphyseal ablation, transverse and longitudinal osteotomies, nerve stripping, and extensive defatting. None of the available methods, however, give ideal functional and cosmetic results^[4]. Most patients are diagnosed and treated from childhood; however, macroductyly might become more severe if diagnosis and treatment are delayed for a long time. In this case, we report a rare case of long-term (23 years) macroductyly of a toe resulting from delayed treatment.

Case Report

A 45-year-old woman was admitted in our hospital with a giant deformed great toe of the left foot which had been present since birth. The walking and shoe wear were difficult for her and caused the size of the shoes of both feet to differ from each other to such a large extent that the right one is six sizes larger than the left one.

Figure 1. On physical examination, the right great toe and forefoot showed extreme enlargement and incredible abnormality.

Figure 2. A plain radiograph of the right foot showed remarkable overgrowth of the First, second, third phalanx bone with significant deformity.

Figure 3. Computed tomography of the right foot.

Figure 1



Figure 1



Figure 2.



Figure 3.



There were no positive family history and other congenital malformations. Moreover, we couldn't find any sign of a systemic disease, e.g., neurofibromatosis. On examination, the left great toe, second, third toes and forefoot showed extreme enlargement and incredible abnormality. Macroducty is combined with syndactyly in second and third toes. The deformed toe was 12 cm in length and 7 cm in horizontal diameter. The plantar skin of the sole became remarkably thick. A plain radiograph of the left foot showed remarkable overgrowth of the 1st, 2nd and 3rd phalanx bone with significant deformity (Figure 2). The major aim in the treatment of foot macroducty is the reconstruction of a pain-free, functional foot with a good cosmetic appearance. A debulking procedure and toes reconstruction with distal phalanx amputation were planned after consideration of the treatment options.

DISCUSSION

Macroducty of the foot is a rare congenital anomaly characterized by an enlargement of the soft tissue and osseous elements. The pathogenesis of macroducty is not known and defined. Heredity does not appear to play a role, and our patient did not have a family history^[5].

Hardwicke et al. described four types of macroducty

Type I: macroducty with lipofibromatosis of a nerve, either of a static or progressive subtype.

Type II: associated with neurofibromatosis;

Type III: associated with hyperostosis;

Type IV: associated with hemihypertrophy.

Macroducty is not necessarily associated with any other type of deformity, although it can be found in people with some types of syndromes, including neurofibromatosis, hemangiomas, arteriovenous malformations, congenital lymphedema, and Klippel-Trenaunay-Weber and Proteus syndromes^[6-8].

However, not every enlargement of the foot, or any element of the foot, is macroducty. Enlargement of a toe can result from the presence of a haemangioma, or some other neoplasm, in which only the overlying skin and adjacent soft tissues are involved, with no radiographic evidence of an increased size of the osseous elements^[9,10]. Other diseases in which "false macroducty" can occur are Ollier's disease, Maffucci's syndrome, vascular malformation, neurofibromatosis, and Milroy's disease^[10]. Syed et al. reported that in macroducty of the foot, excessive proliferation and accumulation of adipose tissue was the basic lesion. But in the hand, macroducty hypertrophy and tortuosity of the digital nerve were the striking features which were absent in cases affecting the foot^[11]. Barsky AJ reported that macroducty is more commonly observed in males and in the foot and he defined two types of macroducty. Static macroducty, the most common, is a congenital growth which progresses in proportion with normal growth. Patients with static macroducty have larger toes (or toe) when they are born. It then continues to grow proportionally to the other fingers. Progressive macroducty, in which the digit involved has a higher rate of growth, is less common. In macroducty, the blood vessels are frequently either of normal calibre or only slightly enlarged; the digit as a whole is thus relatively poorly vascularized. Healing potential is thereby reduced and the incidence of wound breakdown and infection is high. These complications are more likely to occur when extensive dissection and defatting have been performed^[12]. The major goal in surgical reduction of foot macroducty is to obtain a pain-free and satisfactory aesthetic foot^[13,14]. When determining the treatment, it is important to consider the severity of the deformity. Therefore, radiologically, X-ray, computed tomography (CT) and magnetic resonance imaging (MRI) are important for surgical treatment planning. Treatment varies according to which toe is affected. If surgery needs to be done, the surgeon could destroy the growth part of the bone, along with removing as much of the excess tissue as possible. The surgical options include debulking of soft tissues, amputation of the distal phalanx, middle phalanx, or the whole ray, and epiphysodesis by stapling or destroying the epiphysis to prevent additional longitudinal growth without controlling the circumferential growth. Debulking procedures can be performed with epiphysodesis to reduce the size of the toe, but it must sometimes be repeated because of progressive recurrence of the macroducty^[15-17]. In macroducty of the toes, the Tsuge method, which involves excision of the distal phalanx with soft tissue reconstruction using a dorsal flap, along with nail preservation, is generally recommended^[18]. In our case, a 45-year-old patient did not accept amputation of the distal phalanx at the interphalangeal joint combined with the removal of the Fibro-fatty tissue. Because of the rarity of the condition, few reports pertaining to the results of the surgical

treatment of this condition have been published. Bulut et al. reported 3 cases of ray amputation with good cosmetic and functional results. They 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^[19]. 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. Ray amputation is not suggested in macrodactyly of the great toe because of the important role of the great toe in normal stepping and walking, as well as the unacceptable cosmetic result. Hendrix et al. applied amputation of the finger phalanx and painfullump excision of the 23-year-old male patient who was operated on before from the 2nd and 3rd fingers, and the resulting defect closed with a rectus abdominis Flap.No consensus has been reached regarding the treatment of foot macrodactyly. Many surgical options are available for the treatment of macrodactyly of the foot. Thus, the most appropriate technique should be chosen for the patient individually. For severe macrodactyly of the great toe, amputation of the first phalanx could be an alternative treatment.

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