

A Literature Review of Pedal Macroductyly

by Ellise Natividad, DPM Candidate¹ ✉, Kinna Patel, DPM Candidate² ✉

The Foot and Ankle Online Journal 3 (5): 2

Macroductyly is a congenital disease where the digits affected increase in size faster than can be attributed to normal growth of unaffected digits. Its etiology is ambiguous and hereditary patterns do not play a role. The abnormality develops in one or more toes and involves thickening of soft tissues, bone and accumulation of fat. The accepted treatment is reduction of the fibro-fatty bulk via dissection and ablation; the major aim being reconstruction of a pain-free functioning foot. Complications with surgery include delayed wound healing and inadequate initial de-fatting which could require a more proximal amputation.

Keywords: Macroductyly, congenital deformity, thickened digit.

Accepted: April, 2010

Published: May, 2010

This is an Open Access article distributed under the terms of the Creative Commons Attribution License. It permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited. ©The Foot and Ankle Online Journal (www.faoj.org)

Macroductyly was first loosely coined by Feriz when he described lymphomatous degenerative lesions and called them “*macroductyly lymphomatosa progresiva*.”¹ In the foot, the most common presentation of this pathology consists of excessive fibro-fatty tissue on the plantar aspect which causes dorsal curling of the toes.² However, due to the rarity of this condition, there is scarce literature which discusses its incidence, prevalence and treatments.³ The purpose of this article is to discuss the etiology of macroductyly and treatment options by the review of previous case reports.

Macroductyly is a rare congenital disease that can be divided into two types – static and progressive. Static macroductyly is present at birth and the digit appears to have increased in size and grows proportionately with the rest of the body. Progressive macroductyly is characterized by disproportionate growth – the digit affected increases in size faster than can be attributed to normal growth patterns of unaffected digits.⁴ Pedal macroductyly tends to be of the progressive type. Most reported cases indicate a slight male preponderance^{5,6} and the condition can occur unilaterally, bilaterally, symmetrically, or asymmetrically.⁷ One study showed that macroductyly may be associated with syndactyly of the adjacent digit in as much as 10% of cases reported.⁷

Address correspondence to: Temple University School of Podiatric Medicine

¹ Ellise Natividad 10104 Rodeo Drive, Waco, TX 76712. (254) 495-9891.

² Kinna Patel 8654 Marlamoor Lane, West Palm Beach, FL 33412. (646) 279-8657.



Figure 1 Macrodactyly affecting the second digit of a child.

The etiology of macrodactyly is ambiguous. Heredity does not seem to play a factor and no other congenital abnormalities occur in combination with the disease since chromosomal studies performed were negative.⁴ Streeter hypothesizes that the condition might be a *“consequence of germ plasm abnormality or pathology; by accident during cell cleavage or organogenesis, certain areas become supersensitive to growth stimulus.”*⁸ During fetal development, some disturbance of growth limiting factor occurs in areas affected and because of this lack of inhibitor the part affected continues to increase in size and accounts for progressive overgrowth in later years. Barsky also states that the *“condition might be caused by interaction of genetic and environmental factors and may be the result of interaction of two or more extrinsic agents each incapable of producing teratological effects by themselves.”*⁴

Enlargements may also be due to hemangiomas where soft tissues, skin and x-rays do not show any increased size of the digits affected.⁴ Some propose that the development of neurofibromatosis in the periosteal nerves due to bony destruction and regeneration may be the cause of rapid localized overgrowth.⁵

Macrodactyly seems to occur in isolated findings without other evidence of systemic involvement though it may constitute one of the many features of a number of syndromes such as Proteus syndrome, Banayan-Riley-Ruvalcaba, Maffuccin, Ollier’s disease, and Milroy’s disease.⁹

The abnormality develops in one or more toes and involves thickening of both soft tissue and bone of the affected digit. (Fig. 1) It is further characterized by increased size of all elements – tendons, nerves, vessels, subcutaneous fat, nails, skin and phalanges – all but the metatarsals. The increase of nerve size is noticeable and the digit appears to be infiltrated with fatty tissue.⁴ In pedal macrodactyly, accumulation of fat appears to be the most striking feature^{5,15} and histopathological findings suggest that excessive proliferation of adipose tissue is the basis of etiology for this abnormality whereas macrodactyly of the hand involves hypertrophied and tortuous digital nerves.⁹

The fatty tissues in pedal macrodactyly found in children resemble adult subcutaneous fat – dark lobules fixed by many fine vessels which traverse the tissue - rather than the fat of the normal child’s age.^{4,6} Most of the abnormal bulk is due to excessive fibro-fatty tissues and is abundant on the sides and plantar aspects of the toe. The fibro-fatty tissue extends from the toe in the forefoot and causes the bulk to expand laterally. The skin is markedly thickened and the digits involved have a soft and rubbery consistency.⁵ The bone age, as denoted by epiphyseal centers in the phalanges of affected digits, is increased when compared to unaffected bone.¹⁰ There is also an increased length and breadth of phalanges⁵ with proliferation of fibroblastic tissue between the cortex and periosteum and accounted for cortical thickening and gigantism of phalanges in affected digits.¹⁰

As previously stated, macrodactyly may be related to several neurogenic disorders. Abnormalities of peripheral nerves are uncommon in the pedal form though there are nerves more prominent with much proliferation of epineural and perineural tissues.⁵

Macroductyly may also result in reduced function owing to the secondary degenerative joint disorders and may cause compressions of adjacent nerves and vessels as well as disfigurement.⁹

Treatment for macroductyly is on a case by case basis and the major aim is the reconstruction of a pain-free functioning foot.¹¹ The growth of affected digit in children may be retarded by destroying, stapling, or wiring the epiphyseal plate.¹² The accepted treatment for pedal macroductyly is reduction of the fibro-fatty bulk via dissection and ablation combined with removal of as much fat as possible.⁵ De-fatting occurs in a two step procedure where the first step involves only the convex side of the digit and reducing its thickness by 10% to 20% while taking care to preserve its vascularity. The second procedure involves de-fatting the remaining side and performing a phalangectomy – shortening of the bone and resectioning and suturing of the excess skin. However there are new one-step procedures which greatly reduce compromising vascularity. Amputation is also an option, though it is reserved as a last option in most cases.¹¹ Complications with surgery include delayed wound healing and inadequate initial de-fatting could cause subsequent re-growth of the fibro-fatty tissue and may require a second and more proximal amputation.⁵

Case Review One

Since the treatment for macroductyly is on a case by case basis, there will be a review of two different treatment options-one at an earlier stage in life and one at the adult stage. In a case study by Lagoutaris, early treatment of macroductyly is discussed and could be viewed as a better option. Most patients that came in for treatment for macroductyly were found to be in their late teens or adults;¹⁵ although the condition is rare, an earlier treatment would benefit the patient in the future. A pediatric patient will not only benefit from an aesthetically pleasing foot, which will allow him or her to avoid any emotional distress, but will also be privileged to be fitted with normal footwear.

It is probable that the pediatric patient would have to return in the future for additional treatment as hypertrophy cannot be halted; however, any adult patient would require the same post-operative treatments.¹³

In this particular case study, a three year old girl presented with a hypertrophic fourth toe with flexion contracture; all neurovascular findings were normal.¹³ In effort to make the foot more accommodating of footwear, more aesthetically pleasing, and more functional, removal of a large piece of full-thickness skin and subcutaneous tissue via an elliptical incision over the midline of the fourth toe and osteotomy of two-thirds of the proximal phalanx was performed.¹³ A K-wire was placed to help stabilize the osteotomy and the patient was placed in a short-leg walking cast for 6 weeks.¹³ The pediatric patient had no post surgery complications but will most likely require a simple epiphysiodesis as age progresses.¹³ Amputations, of course, were suggested only after surgical options were exhausted. It has been noted that when pathology is in the lesser toes, ray resections produce a more cosmetic and functional outcome. However, when the hallux is involved, ray amputation is coupled with repeated soft-tissue debulking.¹⁴

Case Review Two

Another case study performed by Uemura discussed a twenty-four year old patient's yearn for a cosmetically pleasing second toe. The patient's parents had noticed a large degree of growth difference in the toe when she was about three years old but had not sought any treatment.¹¹ The patient's chief complaint was that of cosmesis, as she was able to run and walk without experiencing any difficulties and had a normal range of motion and sensation.¹¹ Furthermore, she complained about the need to purchase two differently sized shoes for her right and left foot in order to be well fitted and her inability to wear sandals during warmer months for aesthetic reasons.¹¹ Upon evaluation, it was found that the affected toe was 15mm longer and 15mm thicker around the nail than the unaffected toe on the opposite foot and only the metatarsal and phalanges were enlarged.¹¹

Since the patient's primary concern was to aesthetically preserve the toe, a nail graft was used in an osteotomy as well as debulking of subcutaneous tissue, instead of a simple amputation.¹¹ Pho, et al., stated that although shortening of the toe via a simple amputation could have improved the overall morphology of the toe, without a nail, the toe would have looked abnormal.¹⁶ In this case, the need for a grafting procedure outweighed the simplicity of an amputation due to the patient's needs and concerns for an aesthetically beautiful toe.

Both cases were performed with minimal post-operative complications. Consequently, it is difficult to decipher whether treatment at an earlier age would be any more beneficial than treatment as an adult. While treatment at a younger age would save years of emotional distress and provide earlier comfort and relief of symptoms, a child may need to return for additional surgeries later in life. Furthermore, Uemura, et al.,¹¹ stated that it would be difficult to apply that type of procedure to progressive macrodactyly in children. If aesthetics is not of primary concern and if there are no related complications, then electing surgery at the adult stage when the patient is capable of making his or her own decision would seem ideal. Regardless, each case should be treated individually, based on the patient's symptoms, needs and concerns.

Conclusions

Macrodactyly is a congenital disease in which the soft tissue, bone, or fat of the digits affected is enlarged. Due to the rarity of the condition, the prevalence is unknown but predominantly associated with males more than females. Although there has been research conducted on macrodactyly, there is not much known about this deformity, including the etiology. Surgical options vary, and include but are not limited to debulking of the soft tissue and fat, partial osteotomies, and amputations. However, each deformity must be treated on a case by case basis.

References

1. Feriz H. Makrodystrophia lipomatosa progresia. *Virchow Arch Pathol Anat Klin Med* 1925 260: 308-368.
2. Kumar K, Kumar D, Gedegone W, Kapahtia N. Macrodactyly of the hand and foot. *International Orthopedica* 1985 9: 259-264.
3. Cooke S, Davies H, Harris N. Chronic digital infection presenting with gross enlargement of the toes: two case reports and review of the literature. *Cases Journal* 2009 2: 102.
4. Barsky AJ. Macrodactyly. *JBJS* 1967 49A: 1255-1266.
5. Dennyson WG, Bear JN, Bhoola KD. Macrodactyly in the foot. *JBJS* 1977 59B: 355-359.
6. Syed A, Sherwani R, Quamar A, Haque F, Akhter K. Congenital macrodactyly: A clinical study. *Acta Orthop Belg* 2005 71: 399-404.
7. Kotwall PP, Farooque M. Macrodactyly. *JBJS* 1998; 80B: 651-653.
8. Streeter GL. Focal deficiencies in fetal tissues and their relations to intrauterine amputation. *Contrib. Embryol.* 1930 22: 1-44.
9. Yuksel A, Yagmur H, Kural BS. Prenatal diagnosis of isolated macrodactyly. *Ultrasound Obstet Gynecol* 2009 33: 360-362.
10. Ben-Bassat M, Casper J, Kaplan I, Laron Z. Congenital Macrodactyly. *JBJS* 48A: 359-364.
11. Uemura T, Kazuki K, Okada M, Egi T, Takaoka K. A case of toe macrodactyly treated by application of vascularized nail graft. *Br J Plast Surg* 2005 58: 1020-1024.
12. Tsuge K. Treatment of macrodactyly. *Plast Reconstr Surg* 1967 39: 590-599.
13. Lagoutaris E, DiDomenico L, Haber L. Early surgical repair of macrodactyly. *JAPMA* 2004 94: 499-501.
14. Hsieh C, Kumar J, Riddle E. Macrodactyly of the foot. *JBJS* 2002 84A: 1189-1194.
15. Fengdong Z, Rongrong Z, Peihua S, Shunwu F. Macrodactyly of the great toe for thirty-five years. *J Plas Recon Aes Surg* 2009 62: e520-e522.
16. Pho RW, Patterson M, Lee YS. Reconstruction and pathology in macrodactyly. *J Hand Surg Am* 1988 13:78-83.