Review Article

Indian Journal of Diabetes and Endocrinology
Volume 3 Number 1, January - June 2021

Foot Macrodactyly: A Review

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How to cite this article:
Jacob Antony Chakiath, Ravi Kumar Chittoria/Guidelines in Management of Necrotising Fasciitis/Indian Journal of Diabetes and Endocrinology, 2021;3(1):17–20.

Abstract

Macrodactyly affects the foot less frequently than the hand. Macrodactyly has yet to be identified as a cause. A gain-of-function mutation in the PIK3CA pathway causes this form of overgrowth. The two categories of clinical manifestations are static and progressive clinical symptoms. Anteroposterior (AP) measurements are taken on both feet. Young people should have soft tissue reductions, epiphysiodesis, epiphysectomies, osteotomies, and other treatments. Adults are more likely to use arthrodesis and shortening surgeries.The purpose of surgery is to reduce the size of the foot so that standard shoes can be worn and the cosmetic look can be improved.

Keywords: Macrodactyly; Foot.

INTRODUCTION

Macrodactyly of the foot is a rare congenital abnormality that causes pain, calluses[1], ulcers, difficulties wearing shoes, impairment in ambulatory capacity and gait development, aesthetic issues, and psychological issues.[2]

BACKGROUND

Macrodactyly affects the foot less frequently than the hand. One in every 18000 people is affected by primary macrodactyly, with a modest male predominance of.[3,4] Macrodactyly is also known as megalodactyly, macrodystrophia lipomatosa, macrodactyly fibrolipomatosis, lipomatous overgrowth or hamartoma, gigantomegaly, local gigantism, and digital gigantism. The second ray is usually the first, followed by the third, then the first, fourth, and fifth.[5]

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Received on: 28.04.2022
Accepted on: 26.05.2022

ETIOLOGY

The cause of macrodactyly has yet to be discovered. Improper finger irrigation, abnormal humoral mechanism, and abnormal innervation are all possible reasons (the last two causes are not well demonstrated, the first one because the nerves exert great control over the growth).

PATHOGENESIS

An overgrowth condition is caused by a gain of function mutation in the PIK3CA pathway (Phosphatidylinositol - 4, 5 - Bisphosphate 3-Kinase).[6,7] For normal cell growth, metabolism, and survival, the PI3K/AKT/mTO Rsignalling pathway is essential. Cancer and the PIK3CA-

Related Overgrowth Spectrum, a collection of overgrowth illnesses, can be caused by somatic mutations in this system (PROS). PIK 3 CA SNPs in PROS generate asymmetric overgrowth by promoting physiologically inappropriate activation of AKT and mTOR. This includes disorders like macrodactyly, CLOVES (Congenital Lipomatous Overgrowth, Vascular Malformation, Epidermal nevi, Spinal/skeletal Anomalies), hemimegalencephaly, and others. Because these mutations are postzygotic, they are only found in some cells and not others.

PATHOLOGY

Subcutaneous fat in adults looks like proliferative adipose tissue. (Image 1) Fat lobules are large, dark, and difficult to remove. The digital nerve has expanded in size, with an increase in fat and fibrous tissue seen in a cross section. The digital artery wall thickness is normal. Blood vessels, tendons, and the sheaths that surround them are normally untouched. As a result, the blood supply fails to keep up with the toe's growth, resulting in vascular insufficiency. The phalanges' medullary canals are always larger, and fatty marrow is apparent.

Patients with diagnoses of other known overgrowth syndromes or otherwise uncharacterized syndromic presentations of lower extremity enlargement were excluded, as were patients with Klippel Trenaunay syndrome, Proteus syndrome, CLOVES syndrome, Ollier's disease, Maffucci syndrome, Milroy's disease, neurofibromatosis, and vasculaanomalies.

TYPES OF CLINICAL MANIFESTATION

According to Barsky, there are two types of clinical manifestations: the static form, in which the finger is larger from birth but grows proportionally with the rest of the fingers, and the progressive form, in which the finger is normal at birth but begins to grow faster than the rest of the fingers, causing angular deviation.[8] For examples described in the hand[9], however, De Laurenzi identified the less common progressive type.

The condition manifests itself unilaterally in 95% of cases. It appears in a decreasing order from the great toe to the fifth toe in males and is slightly more common in females. In the progressive form, the toe stops growing when the epiphyses close, the sensitivity is usually normal, mobility declines over time, and bud ulcers are common.

Macrodactyly is associated with syndactyly in 10% of people[10], and polydactyly and cryptorchidism in a lower percentage. Klipper Syndrome is a condition that occurs when a person Maffuci Syndrome (multiple hemangiomatosis), Proteus Syndrome (hamartomatous dysplasia, pigmented nevi, and subcutaneous tumours), hemangiomas, arteriovenous malformations, congenital lymphedema, lipomas, osteoid osteoma, and melorrestos[11] Hernia may be linked to Macrodactyl.

The innervations of the medial plantar nerve were more pronounced in the affected toes and forefoot than those of the lateral plantar nerve. The clinical and anatomic aspects of foot macrodactyly were comparable to the affected digits primarily located in the area of the median nerve in hand macrodactyly, indicating that it was "nerve territory-oriented."[12,13] Enlargement, fatty infiltration, tortuosity, or a combination of these pathologic alterations are all examples of pathologic changes.[14] The remarkably consistent distribution of the nerve abnormalities and tissue hypertrophy suggested that macrodactyly of the foot was caused by nerve-mediated expansion.[15]

INVESTIGATION

We took standing anteroposterior (AP) and oblique radiographs of both feet and evaluated the range of motion of the metatarsophalangeal joint on the first visit (MTPJ). In standing anteroposterior radiographs, evaluate the intermetatarsal breadth and forefoot area on AP radiographs (Fig. 1 and 2).

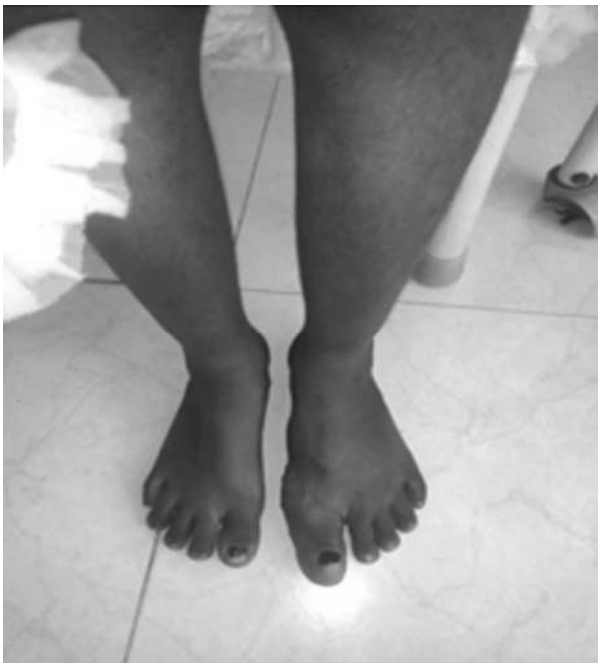

Fig. 1: Enlarged left big toe.

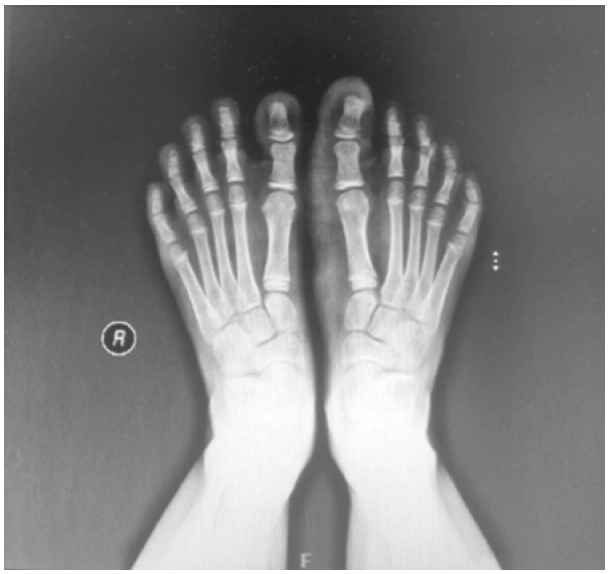

Fig. 2: Standing Antero posterior view of both foot.

We drew a line on the anatomical axis of the first metatarsal and labelled the point of intersection with the distal end of the first metatarsal as M1 to calculate the inter metatarsal breadth. A similarly generated point on the fifth ray was given the designation M5. The distance between M1 and M5 was used to compute the intermetatarsal breadth.The area of the soft tissue shadow distal to the tarsometatarsal joint was calculated using Image J software (NIH, Bethesda, Maryland) and recorded as the forefoot area. We measured these two radiographic parameters in the contralateral, normal foot and calculated the intermetatarsal width ratio as the intermetatarsal width of the macrodactylic foot divided by that of the normal foot, and performed a similar calculation with the forefoot areas of both feet to derive the forefoot area ratio.

Although the forefoot area, as a two dimensional measurement, is superior to a one dimensional measurement, such as foot length or foot breadth, a three dimensional CT or MRI could be used to quantify the volume of the foot prior to surgery.

To determine the nerve involvement, a nerve conduction examination is performed.

TREATMENT AND MANAGEMENT

The goal of surgery is to reduce the size of the foot so that it may be used in regular shoes and the appearance can be enhanced. Young people should have soft tissue reductions, epiphysiodesis, epiphysectomies, osteotomies, and other treatments. Adults[16-21] are more prone to undergo shortening operations and arthrodesis. Surgical indications for ray amputation, as previously stated by Bulut et al., were metatarsal involvement, joint immobility, or involvement of several digits.[22] Because the immobile macrodactylic toe impairs foot function, this is the case.

Hop and van der Biezen[23] described three cases of foot macrodactyly involving several digits and recommended amputation of the most swollen ray, shortening the adjacent ray, ray transposition, and soft-tissue debulking.

Kotwal operated on 21 cases and after 9 years of follow-up, the aesthetic aspect of the finger was good only in 57% of patients. The techniques used were two-stage soft tissue reduction and phalangectomy.[24]

When the metatarsal spread angle is raised by 10° relative to the normal side, or toe amputation would create a broad, cosmetically unattractive interphalangeal area, ray resection of the lesser toes should be performed. Because removal of the first ray is not suggested, treating macrodactyly of the great toe may necessitate many surgical procedures. After a comprehensive examination of the malformation, the surgeon can perform certain procedures on the child while he or she is still young.

After macrodactyly reconstruction techniques, complications are fairly common. Cutaneous necrosis is the most common, and it can be severe enough to jeopardise the toe's viability. After soft tissue reduction, 25 percent of patients developed cutaneous necrosis, according to Kostakoglu18. Dell also found this complication frequently after reconstruction techniques.[25]

Despite years of experience treating this difficult congenital defect, most individuals require many surgical operations during childhood, and a large percentage of them have an ugly and useless finger as a result.[26,27]

CONCLUSION

We may conclude that macrodactyly is a rare and complicated illness with a wide range of treatment options and frequent complications.

REFERENCES

1. Barsky AJ. Macrodactyly. J Bone Joint Surg Am. 1967;49:1255–66.
2. Natarajan M, Dhua S, Garg C. Macrodactyly of lower limbs-an update. J Evol Med Dent Sci. 2016;5:3806–10.

3. Kowtharapu DN, Thawrani D, Kumar SJ. Macrodactyly. In: McCarthy JJ, ed. Drennen's the child's foot and ankle. 2nd ed. Baltimore: Lippincot Williams and Wilkins; 2009:443–449.
4. Kalen V, Burwell DS, Omer GE. Macrodactyly of the hands and feet. J PediatrOrthop 1988;8:311–315.
5. Temtamy SA, Rogers JG. Macrodactyly, hemihypertrophy, and connective tissue nevi: report of a new syndrome and review of the literature. J Pediatr 1976;89:924– 927.
6. Keppler-Noreuil KM, Rios JJ, Parker VE et al. PIK3CA-related overgrowth spectrum (PROS): diagnostic and testing eligibility criteria, differential diagnosis, and evaluation.Am J Med Genet A. 2015, 167A (2):287-95.
7. Labow BI, Pike CM, Upton J. Ovegrowth of the hand and upper extremity and associated syndromes. J Hand Surg Am. 2016,41:473- 82;quiz 482.
8. Barsky AJ (1967) Macrodactyly. J Bone Joint Surg 49:1255-1266.
9. The hand, part II. In: May JW, Hillter JW, editors (1990) McCarthy plastic surgery. Philadelphia: WB Saunders 8: 5362-5365.
10. Ozturk A, Baktiroglu L, Ozturk E, Yazgan P (2004) Macrodystrophialipomatosa: A case report. ActaOrthopTraumatolTurc 38: 220-223.
11. Krengal S, Morales AF, Carrcasco P, Vazquez M, Mickinster CD, et al. (2000) Macrodactyly: Report of eight cases and review of the literature. Pediatric dermatology 17: 270-276.
12. Lagoutaris ED, DiDomenico LA, Haber LL. Early surgical repair of macrodactyly. J Am Podiatr Med Assoc. 2004;94:499 –501.
13. Hart ES, Grottkau BE. Isolated macrodactyly of the foot.OrthopNurs. 2012; 31:212 –5.
14. Vivanco I, Sawyers CL. The phosphatidylinositol 3-kinase –AKT pathway in human cancer.Nat Rev Cancer. 2002;2:489 –501.
15. Turra S, Santini S, Cagnoni G, Jacopetti T. Gigantism of the foot: our experience in seven cases. J PediatrOrthop. 1998;18:337 –45.
16. Tsuge K (1967) Treatment of macrodactyly. PlastReconstrSurg 39: 590-599.
17. Kelikian H (1974) Congenitaldeformitie of the hand and forearm. Philadelphia: WB Saunders.
18. Kostakoglu N, Kayikcioglu A, Safak T, Ozcan G, Kecik A, et al. (1996) Macrodactyly: report of eight cases of a rare anomaly. Turk J Pediatr 38: 73-79.
19. Bertilli JA, Pigozzi L, Pereima M (2001) Hemidigital resection with collateral ligament transplantation in the treatment of macrodactyly: a case report. J Hand Surg 26: 623-627.
20. Tsuge K (1985) Treatment of macrodactyly. J Hand Surg 10: 968-969.
21. Chen SH, Huang SC, Wang JH, Wu CT (1997) Macrodactyly of the feet and hands. J Formos Med Assoc 96: 901-907.
22. Bulut M, Karakurt L, Belhan O, Serbest S. Ray amputation for the treatment of macrodactyly in the foot: report of three cases. ActaOrthopTraumatolTurc 2011;45:458–462.
23. Hop MJ, van der Biezen JJ. Ray reduction of the foot in the treatment of macrodactyly and review of the literature. J Foot Ankle Surg 2011;50:434–438.
24. Morris C, Doll HA, Wainwright A, Theologis T, Fitzpatrick R. The Oxford ankle foot questionnaire for children: scaling, reliability and validity. J Bone Joint Surg [Br] 2008;90-B:1451–1456.
25. Dell PC (1985) Macrodactyly. Hand Clin 1: 511-524.
26. Álvarez R, Peña L, López H, Hernández R, Hernández O, et al. (2002) Transposición digital en el tratamiento de la macrodactilia. Rev CubanaOrtopTraumatol 16: 65-69.
27. Isik A, Gursul C, Peker K, Aydın M, Fırat D, et al. (2017) Metalloproteinases and Their Inhibitors in Patients with Inguinal Hernia. World J Surg 41: 1259-1266.