

Macrodystrophia Lipomatosa of Right Hand and Right Foot: Case Report

Leela Sai Parnam¹, E Ruchitha¹, Makkapati Pravalika¹, Boyanaga Mahesh¹,
B Victor Kumar¹, Tilak Ram¹

¹Department of General Medicine,
Mediciti Institute of Medical Sciences, Ghanpur Medchal, Telangana-501401 (India)

Corresponding Author: Tilak Ram

DOI: <https://doi.org/10.52403/gijhsr.20240208>

ABSTRACT

Macrodystrophia Lipomatosa (MDL) is an uncommon congenital disorder characterized by excessive growth of mesenchymal tissue. Etiology of this disorder is obscure. It results in disproportionate increase in size of affected limbs or digits manifesting as localized gigantism. Diagnosis of MDL depends on clinical features and imaging studies. We present a case of MDL causing gigantism in the right upper and right lower limb.

Keywords: Macrodystrophia Lipomatosa MDL, macrodactyly, congenital localised gigantism.

INTRODUCTION

Macrodystrophia Lipomatosa (MDL) is a rare congenital, nonhereditary disorder characterized by progressive overgrowth of mesenchymal tissues with a disproportionate increase in fibroadipose tissues which results in development of localized gigantism involving digits and extremities. The affected part is usually the lateral aspect of hand and the medial aspect of foot in the distribution of median nerve and medial plantar respectively. We report a case of Macrodystrophia Lipomatosa affecting right hand and right foot in a young male patient.

CLINICAL PRESENTATION

A 23 years-old right-hand-dominant male presented with a progressive disproportionate enlargement of digits of right foot and right hand since early childhood. He did not have any neurological or vascular symptoms. There was no history of any trauma. On examination he was found to have enlarged right hand and right foot (Figure 1). The enlargement mostly involved medial aspect of foot and lateral aspect of hand. The involvement mainly affected the right 2nd and 3rd toes along with syndactyly. The involvement of the other side of limb was minimal. There were no overlying skin changes or edema. There was no associated neurological abnormality.

Plain radiograph of the right foot showed enlargement of right 2nd and 3rd digits along with soft tissue excessive growth. Plain radiograph of the right hand showed enlargement of right thumb, index and middle fingers along with soft tissue excessive growth (Figure 2).

Other investigations for associated abnormalities were non-contributory. As he did not have any disability and there were no significant symptoms, and he did not have much cosmetic concern, he was treated conservatively with advice for regular follow up and consideration for surgical intervention as and when needed.

Figur:1. Enlarged Right and Right Foot-Localized Gigantism (A)



Figure 2. Plain Radiograph Showing Enlarged Right Hand and Right Foot Digits (B)



DISCUSSION

MDL is a rare congenital nonhereditary anomaly characterized by excessive growth of mesenchymal tissues.^[1] It usually affects the medial side of foot and lateral side of the hand.^[2] Mostly it is unilateral but it may be bilateral. There is a slight male preponderance. Overgrowth tends to involve the limbs in specific sclerotome region of the body i.e. along the distribution of median nerve and medial plantar nerve. Although both upper and lower limbs are affected, the lower limb is usually affected more. Often 2nd and 3rd digits are more commonly involved.^[3]

Various names have been used to describe the excessive growth of mesenchymal tissue like macrosomia, partial acromegaly, localized gigantism, club finger, dactomegaly, macrodystrophia lipomatosa. The term macrodystrophia lipomatosa was first coined by Feriz in 1925 to describe

localized gigantism in the lower limb. Barsky described the disorder in greater details. MDL is divided in two forms: static form and progressive form of macrodystrophia lipomatosa. The static form is characterized by increase in size of the affected digits proportionally to the rest of the body while in the progressive form there is disproportionate growth of the enlarged digits compared to the rest of the body.^[4] Exact Etiology of the disorder is obscure; probable mechanisms include: abnormal nerve supply, disturbed foetal circulation, lipomatous degeneration, disturbances of growth inhibiting factors, an error in segmentation, and trophic influence of tumefied nerve^[5] The rate of accelerated growth varies among patients and also among affected digits.^[6] The excessive growth of affected bones normally ceases by puberty. Beside the involvement of subcutaneous tissue, there is involvement of

other tissues including muscles, periosteum, and nerve sheaths. Radiological investigations play an important role in making the diagnosis of MDL. In typical cases plain radiographs along with the clinical picture may be sufficient to make the diagnosis. The plain films may show enlarged soft tissue along with broad, lengthened and splayed phalanges with endosteal and periosteal deposition. [7] Severe secondary degenerative changes may affect the joints in late childhood. Some of the affected individuals may have other skeletal abnormalities affecting the digits like syndactyly, polydactyly, and clinodactyly. CT scan shows excessive fatty proliferation along the nerve territory and muscle tissues. On MR imaging there is abundant fibrofatty tissue in the affected segment of the limb with signal characteristics similar to subcutaneous fat i.e. high signal on T1W, intermediate signal on T2W, and low signal on fat suppressed sequences. This excessive fatty tissue has no capsule. [8]

Localized gigantism may be seen in various conditions. The Differential diagnosis of MDL includes fibrolipomatous hamartoma of nerve, neurofibromatosis, haemangiomas, lymphangiomatosis, Proteus syndrome, Klippel-Trenaunay-Weber syndrome, Beckwith-Wiedemann syndrome, and Maffucci's syndrome. [9] MDL can be differentiated from other disorders based on family history, radiological investigations including ultrasound and Doppler to distinguish vascular and lymphatic malformations in selected cases.

Treatment of MDL can be surgical or nonsurgical depending upon the severity of deformity, age and clinical manifestations. [10] Surgical interventions are usually carried out after puberty when growth ceases. Patients usually seek medical care for cosmetic reasons. The surgery may include microsurgical techniques including debulking and partial amputation of the affected part. Recurrent interventions may

be required for a more aesthetic and functional result.

Declaration by Authors

Acknowledgement: None

Source of Funding: None

Conflict of Interest: The authors declare no conflict of interest

REFERENCES

1. CS Prabhu, K Madhavi VN Amogh. Macrodystrophia Lipomatosa; A Single Large Radiological Study of a Rare Entity. *J Clin Imaging Sci.* 2019 Feb 27; 9: 4
2. S Sahu, A Kumar, A Sen. Macrodystrophia Lipomatosa; *Med J Armed Forces India*, 2011 April;67 (2), 162–164
3. Rizwan A Khan, Shaguta Wahab Ibne Ahamad. Macrodystrophia lipomatosa: four case reports; *Ital J Pediatr.* 2010 Oct, ;36: 69
4. Boren WL, Henry REC, Wintch K. MR diagnosis of fibrolipomatous hamartoma of nerve: macrodystrophia lipomatosa. *Skeletal Radiol* 1995 May; 24(4):296–297).
5. Maheswari S Uma, Sampath V, Ramesh A. Macrodystrophia lipomatosa: An unusual cause of localized gigantism. *Ind J Dermatology.* 61 (3): 347, May-Jun 2016
6. Deepika Upadhyay, Umesh C Parashari, Sachin K. Macrodystrophia Lipomatosa: Radiologic-Pathologic Correlation; *J Clin Imaging Sci* 2011 Mar, 1:18
7. Rakesh Mehra, Richa Bhatiya, Pallavi Agarwal. Macrodystrophia Lipomatosa: Radiologic-Pathologic Correlation. *Ind J Med Paediatr Oncol.* 2017 Oct-Dec; 38 (4): 559-562
8. Alagar Raja Durairaj1 Surya Rao Rao Venkata Mahipathy; Macrodystrophia Lipomatosa of the Toe; *Journal of Clinical and Diagnostic Research.* 2016 Apr, Vol-10(4): PD27-PD28
9. Jae Hyn Kwon, So Young Lim, Ha Seong Lim. Macrodystrophia

- Lipomatosus. Arch Plast Surg. 2013 May; 40(3): 270-272
10. Theddeus Oh Prasetyono, Eneline Hanafi, Windi Astriana. A Review of Macrodystrophia Lipomatosa. Arch Plast Surg. 2015 Jul; 42 (4): 391-406

How to cite this article: Leela Sai Parnam, E Ruchitha, Makkapati Pravalika, Boyanaga Mahesh, B Victor Kumar, Tilak Ram. Macrodystrophia lipomatosa of right hand and right foot: case report. *Gal Int J Health Sci Res.* 2024; 9(2): 55-58.
DOI: <https://doi.org/10.52403/gijhsr.20240208>
