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A Review of Macrodystrophia Lipomatosa: Revisitation

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Macrodystrophia lipomatosa (MDL) is a rare congenital non-hereditary disorder that has significant impact on patient morbidity. This study provides a comprehensive review of the natural history, diagnosis, management, and outcomes of the disorder. A literature search in PubMed was conducted to identify cases of MDL from January 1950 to 14 February 2014. After ruling out articles without information related to the management of the disorder, a summary of 32 studies was performed. An additional three cases from the authors are also presented. Based on 57 journal articles and three additional cases from the authors, around 108 cases of MDL were reviewed. Most patients were males who were admitted to a treatment clinic in the first four years of life. The lower extremities were more frequently affected, with unilateral presentation being most common. They commonly underwent a single-staged surgical procedure with follow-up periods ranging from more than one year up to 21 years. Out of 43 cases that underwent surgical procedures, 13 reported no complications, and there were seven cases of esthetic satisfaction and 15 cases of significant functional improvement. Depending on the severity of a patient's condition, the use of non-invasive diagnostic tools should be carefully considered. Surgery might be a better choice of management than observation, taking into account possible future complications in the absence of surgery and the beneficial outcomes of surgical procedures.

Keywords Congenital limb deformities / Fingers / Lower extremity / Upper extremity

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INTRODUCTION

Macrodystrophia lipomatosa (MDL) is a rare congenital non-hereditary disorder in which a disproportionate overgrowth of fibroadipose tissue manifests as gigantism of a small part or the whole of an extremity(-ies). It causes significant functional and esthetic problems for patients and families. Although its impact on the well-being of patients is significant, not much is known about this disorder. A systematic review of more than 100 studies by Tahiri et al. [1] discussed the diagnostic approaches and management of 180 cases of MDL. However, the summary could be considered inadequate, because the cases reported were only those involving the median nerve.

In addition to the invasive gold standard of histopathological examination, other non-invasive examinations could help diagnose MDL. Both surgical and non-surgical approaches can be used in the management of MDL, with varying complications and outcomes in different cases [2,3].

To our knowledge, there is currently no study that provides comprehensive guidance on how to diagnose and manage pa-

tients with MDL. The present study provides a comprehensive summary of the disorder in a review format. Furthermore, discussion of diagnosis and management choices of the given cases is included to provide better understanding of the results of the review.

METHODS

A literature search in PubMed was conducted to identify cases of MDL from January 1950 to 14 February, 2014. MDL and macrodactyly were used as the search terms. Studies that fulfilled the inclusion and exclusion criteria were included. The inclusion criterion was all case series and case reports with the diagnosis of MDL. Exclusion criteria were articles with systematic review and literature review, as well as letters to editors. The abstract and full text of each article were obtained and screened by two independent reviewers (E.H. and W.A.). Duplication of cases was prevented by repeated matching by the authors.

An initial literature search revealed a total of 73 articles. Articles without information related to the management of the disorder were excluded, after which a total of 32 studies was obtained that was further appraised by using forms adapted from the Center for Evidence-Based Management [4].

Additionally, a case series of three patients who were enrolled at our hospital unit between January 2013 and December 2013 is also presented in this study. These cases are included in the summary.

Details, such as age, gender, anatomical location and size of affected areas, diagnostic tools used, management, length of follow-up, post-surgery complications, and outcomes were collected as basic data. The data was then simplified into several tables.

Age was defined as the age of the first presentation to the hospital due to MDL. Anatomical location was categorized as unilateral or bilateral and which part(s) of the anatomical area was affected. Management of MDL was divided into surgical (multi-staged, single-staged, or unspecified) and non-surgical. Unspecified surgery was categorized as unspecified when the article did not provide enough information for us to decide, and observation was not a part of this category. Non-surgical management consisted of on-demand and intentional observation. Length of follow-up was defined as the period of time after the first management was implemented until the last time the patient presented for assessment. Post-surgical complication(s) were defined as any mild or severe event related to the affected area after surgery. Finally, outcomes were divided into esthetic and functional. While esthetic outcome is defined subjectively by the patient or parent, the functional outcome is decided objectively by doctors or authors. Significant and fair functional improvements differed in how the surgery affected the patient's daily function. If the surgery had a strong positive impact on the patient's daily activity, improvement was classified as significant. If the changes only had a mild impact, improvement was defined as fair. Improvement was defined as unspecified when there was no information on the function of the affected extremity prior to the surgery.

RESULTS

There were a total of 108 cases of MDL identified from both the

Table 1. Characteristics of patients in the authors' unit

No.	Age (yr)	Gender	Length of follow-up (mo)	Anatomical Location	Size	Diagnostic tools	Management	Compli-cations	Outcome	
									Esthetic	Functional
1	18	Male	14	Calcaneal and Achilles area of the right foot	Right foot shoe size was three sizes higher than the left	MRI	Childhood: amputation of 4th and 5th toes. At 18 yr: mass reduction	N/A	Satisfactory	Significant improvement
2	6	Female	3	Second, third, and fourth toes of the left foot	Metatarsal head-to-head size was double that of the normal right foot; third toe was six times the normal length and circumference	MRI	Central ray amputation and soft tissue and bone reduction of the second and fourth rays	Delayed wound healing; healed by skin graft	Satisfactory	Fair improvement
3	14 mo	Male	2	Thumb, index, and thenar area of the right hand	Circumferences were three and four times higher than normal	Plain radiography	Staged mass reduction and joint reconstruction separated by six-month interval	N/A	Satisfactory	Fair improvement

MRI, magnetic resonance imaging; N/A, not available.

57 journal articles obtained from the literature search combined with the three additional cases from the authors. Among these cases, only 43 journal articles contained information about clinical management of MDL. With the addition of three cases from the authors, there were 46 cases that could be reviewed. A summary of these all-inclusive studies is provided in the Appendices (Appendices 1, 2). The details of the three additional MDL cases can be seen in Table 1.

Table 2 [5] describes the clinical characteristics of all the cases, including three cases from the authors. Twenty-three percent of patients admitted were children, particularly in the first four years of life; more than half of the patients were male. Out of 46 patients, 28 underwent a single-staged surgical procedure, 11 had multi-staged surgery, and four patients went through unspecified surgery. Only three patients with two unilateral cases and one bilateral case were under observation alone. Among the 46 cases, only one indicated bilateral involvement, specifically in the upper extremities. The treatment was different for each side in this case; the left side underwent surgery, while the right side was only observed.

The length of follow-up was not provided by most articles. From the available information on follow-up, only nine patients were monitored up to one year, while 13 patients were followed-up for periods ranging from more than one year up to 21 years. A length of follow-up for more than 21 years occurred in only two patients. After surgery, most patients reported no significant complications and good esthetic and functional outcomes. The unavailability of such data, however, is around 50% among all patients who had surgery. In the patient with bilateral involvement, the operated extremity was performing better functionally compared to the non-operated extremity.

The review also shows that almost 60% of the cases involved the lower extremity (Table 3). The hand and foot were found to be common sites of MDL, with percentages of 54.1% and 64.4%, respectively. MDL affects upper limbs in 31.7% of cases and 5.1% in lower limbs only.

There are several characteristics that describe patients who undergo surgical and non-surgical treatment. A patient could have an amputation if there are severe functional and esthetic disturbances due to MDL. Debulking or reduction is usually performed if nerve and tendon preservation are planned in addition to esthetic preservation. Other types of surgery are performed depending on the condition of the patient. On the other hand, non-surgical cases are divided into observation on demand and intentional observation. Observation on demand occurs in patients who refused surgical treatment, while intentional observation is chosen in the presence of other urgent medical conditions or as decided by the physicians. Local single-digit involvement is said to be an indication for observation due to the principle of digit preservation in reconstruction.

Different radiographic tools are utilized in MDL. Plain radiography (X-ray) can detect any abnormalities in bone, soft tissues, and joints. Magnetic resonance imaging (MRI) can show fat tissue predominance and the condition of nerves and their sheaths. Computed tomography (CT) scans are used to detect proliferation of fat with bone overgrowth. Ultrasonography can be performed to detect any calcification and abnormal blood flow. There are also other additional examinations, such as nerve conduction tests and electromyography that are performed as necessary.

Table 2. Clinical characteristics of all patients

Characteristic	Reported with management (n = 46)	Reported without management (n = 62)	Total (n = 108)
Age on admission (yr)			
0–4	19 (41.3)	6 (9.7)	25 (23.1)
5–9	5 (10.9)	8 (12.9)	13 (12.0)
10–19	6 (13.0)	4 (6.5)	10 (9.3)
20–29	5 (10.9)	0	5 (4.6)
30–39	2 (4.3)	3 (4.8)	5 (4.6)
40–49	5 (10.9)	4 (6.5)	9 (8.3)
≥ 50	3 (6.5)	0	3 (2.8)
No data	4 (8.7)	34 (54.8)	38 (3.5)
Gender			
Male	31 (67.4)	27 (43.5)	58 (53.7)
Female	15 (32.6)	24 (38.7)	39 (36.1)
No data	0	11 (10.2)	11 (10.2)
Management			
Surgical			
Single	28		
Multiple	11		
Unspecified	4		
Non-surgical[a]	4		
Length of follow-up (yr)			
0–1	9		
> 1–20	12		
≥ 21[a]	3[a]		
No data	22		

Values are presented as number of cases (%).
[a]Bilateral case patient [5].

Table 3. Affected areas in patients with macrodystrophia lipomatosa

Value	No. of cases (%)
Upper extremities (n = 41)	
Upper extremities	13 (31.7)
Hand only	23 (56.1)
Unspecified	5 (12.2)
Lower extremities (n = 59)	
Lower extremities	3 (5.1)
Foot only	38 (64.4)
Unspecified	18 (30.5)

DISCUSSION

About 89 years after its first report [6], MDL has become widely known as a disorder of proliferation of mesenchymal and adipose tissue. The affected area grows in a normal pattern or much faster than the unaffected area, causing a gigantic disproportional appearance [7]. In recent years, there have been few articles describing cases diagnosed as MDL.

In this study, we found that the admission with MDL occurs primarily before the age of four. Problems start to surface with the growth of the child. Toddlers are reported to have difficulty in wearing shoes and to sustain repeated injuries, which may affect their daily activities, especially their learning development, social interaction, and self-confidence [2]. Secondary functional problems, such as secondary osteoarthritis and compression of neurovascular elements, usually arise when the patients are older [8]. This is why some patients first present at a later age. Esthetic problems, on the other hand, can begin at any age [2,8-11]. Patients or their parents seek treatment typically due to the cosmetic appearance of the affected area, although functional reasons also play an important role.

MDL can be categorized into static and progressive types. In static MDL, the affected area grows proportionately, while in progressive MDL, disproportionate growth occurs [12]. Some studies have postulated that the growth of the affected area may slow down or cease if patients have finished their growth spurts [8,10,13]. This idea, however, seems more likely to apply to the static type of MDL. As the rest of the body ends its growth, the growth of the affected area may stop as well.

MDL has no gender predominance [14-16]. The difference in the number of males and females with MDL in this study are too insignificant to be considered as predominance. Our study also found that unilateral distribution is common, because bilateral distribution only occurred in three cases. Hand or foot involvement is more common than whole extremities. Specifically, there is a high possibility for the first, second and third digits of the hand or foot to be involved singularly or adjacently. The combination of the second and third digits has the highest frequency (Table 4) [17-20].

The findings may be related to the theory that MDL manifests according to the distribution of the sclerotome [21]; this is a group of mesenchymal cells that gives rise to the skeletal tissue of the body and develops into the vertebrae and ribs [22]. As the little finger is rarely affected (Appendices 3, 4), it is also possible that the distribution is based on the median or plantar nerve innervations.

The diagnosis of MDL is made through clinical presentation and radiographic examination. Many modalities have been reported to act as adjuncts in confirming the diagnosis, providing histopathological examination could not be performed [11,13,15]. Basic radiographic modalities that have widely been used are X-ray [23-26], MRI [10,16,27-31], CT scan [11,14,32,33], and ultrasonography [2,14,33,34]. Additional examinations, such as nerve conduction tests and electromyography [27,35], may also be performed. In fact, X-ray alone might be sufficient to diagnose MDL. This idea is supported by the presence of cases that used radiography as the single diagnostic tool, including one case from the author. If the extent of the disorder is unclear, or the composition of the macrodactyly tissue is doubtful, an MRI may be performed. Nonetheless, choices of diagnostic modality are made according to the clinical features, the availability of the modality and the goal of the examination.

As discussed previously, the management of MDL depends on age, clinical manifestation, and the extent of the disorder [2,8,36]. It can be divided into non-surgery and surgery. The decision regarding frequency of follow-up observation generally lies with the patients or parents, and with the physicians. Two types of observation are stated: observation on demand and intentional observation. The first type is determined by the patients or parents, while the latter might be led by the physicians. The benefits of observation are still unknown. However, there might be a lower recurrence rate once the patients have had surgery. In the bilaterally affected case report, there was an example of a worse outcome that was related to the decision of observation [5]. The follow-up duration of 42 years resulted in an unspecified functional outcome for the operated limb and a reduced functional outcome for the non-operated limb. The reason behind these

Table 4. Digital combinations in macrodystrophia lipomatosa

Value	Hand	Foot	Unspecified extremity	Total
One digit only	10	16	2	28
1st and 2nd digit	5	1	3	9
2nd and 3rd digit	3	6	7	16
3rd and 4th digit	1[a]	0	2	3
4th and 5th digit	2[a]	1	0	3
1st, 2nd, and 3rd digit	2	4	1	7
2nd, 3rd, and 4th digit	1	5	2	8
3rd, 4th, and 5th digit	0	0	1	1
1st, 2nd, 3rd, and 4th digit	0	1	1	2
2nd, 3rd, 4th, and 5th digit	0	0	3	3
1st, 2nd, 3rd, 4th, and 5th digit	0	0	1	1
Unspecified digit(s)	12[b]	7[c]	0	19
Total	36	41	23	100

Values are presented as number of cases.
Digital combination is adapted from Gupta et al. [17].
[a]One bilateral case with information on each side [5]; [b]One bilateral case [18]; [c]Two bilateral cases [19,20].

observations was not discussed in the study. Considering the occurrence of such an outcome, it might have been better if the patient had had surgery on both extremities. A suggestion by the first author is that the right non-operated limb should undergo amputations of the third and fourth rays with an additional approach to the little finger, so that the patient could maintain his right hand grasp function. Overall, there is not enough strong evidence to support non-surgical management of MDL.

Surgery is more preferable than non-surgery to manage MDL [5,36]. The type of surgery often performed is amputation, followed by debulking or reduction. These are mostly executed in single-staged surgery. Multi-staged surgery usually occurs in more complex cases with a longer duration of follow-up. This could involve epiphysiodesis, osteotomy, and carpal tunnel release. Epiphysiodesis and osteotomy are performed with an objective to halt the growth of the abnormal limb and correct the deformities caused by disproportionate growth [34,37]. Carpal tunnel release is only performed when the patient complains of pain of the affected area.

Many patients underwent single-staged surgery in this study. It is important to note that these cases might have no follow-up data available, because many of them were presented as case reports or case series. The patients might or might not have presented for subsequent surgery in the future. Despite this, the length of follow-up in this study can be regarded as acceptable, as it covers more than a year. This time may be adequate for complications, both early and late, to surface. Some complications are loss of sensation, under-correction, and skin blackening [2, 9,27]. Loss of sensation can result from the damage from extensive lesions, which need extensive surgery. The risk of complication after extensive surgery is about 30%–50% for nerve injury, and 33%–60% for recurrence of MDL [8,11]. Our study could not actually give the rate of recurrence of MDL due to the variety of patients' baseline characteristics and different rate of follow-up. Under-correction usually happens when there is an underestimation of the lesion dimension; this can be corrected readily. Skin blackening is caused by injury to the digital arteries [2]. A cautious approach during surgery and proper wound treatment are essential to prevent this. In order to ensure fewer complications, it is reported that ray removal might be a better choice [8]. If in the planning stage it is predicted that the function of the affected limb could not be preserved or that the digit may not function well despite its preservation, then ray amputation could be performed to prevent further complication, such as nerve entrapment, in the future. A suitable example can be observed in the second case given (Fig. 1).

Esthetic outcomes, as mentioned earlier, play an important part in the evaluation of surgery outcomes [10,11]. The satisfaction from the patients or parents may be higher than the physicians' expectation. Drastic changes in the size and shape of the affected area may give an impression that any reduced appearance of the area is good enough. In contrast, significant improvement in functional outcome can be hard to achieve. Retention of the function of the extremity, as well as procurement of appropriate function, depends solely on the surgery process and the severity of the disorder prior to surgery. An algorithm is proposed to guide physicians, who have more limited resources in dealing with MDL in particular (Fig. 2).

In this review, three additional cases are also given (Table 1). The cases consist of two males and one female patient aged 18 years, 14 months, and 6 years, respectively. The age distribution is older than in the other studies; however, the first case had had

Fig. 1. Presentation of patient no. 2 in the unit

A 6-year-old female presented with a metatarsal head-to-head size that was double that of the normal right foot; her third toe was six times the normal length and circumference (A, B). X-ray indicated an enlargement of the second, third, and fourth digits (C, D).

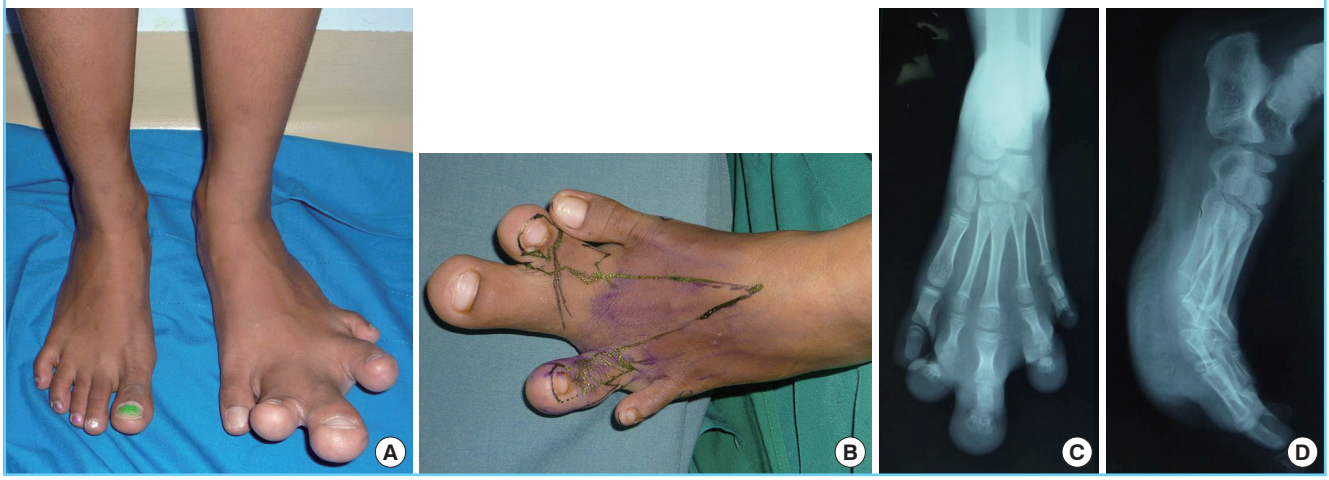
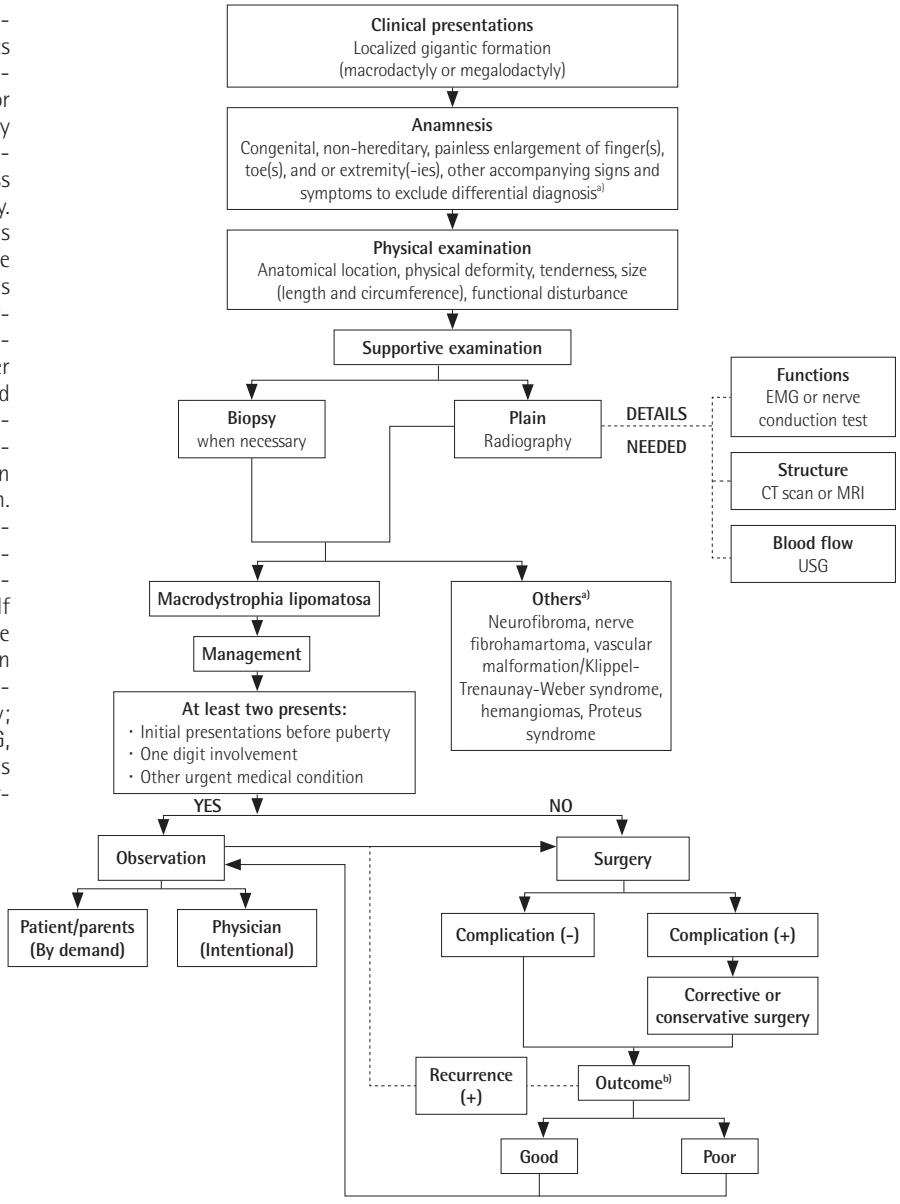


Fig. 2. Algorithm for management of macrodystrophia lipomatosa

This algorithm shows how to manage macrodystrophia lipomatosa (MDL) patients from early consultation to definitive management. Presentations of macrodactyly or megalodactyly should be accompanied by precise anamnesis. It should be asked whether the condition is painful or painless and if it is congenital and/or hereditary. Detailed physical examination should focus on the anatomical location, nature of the physical deformity, tenderness, dimensions of the affected area, and functional disturbances. At the least, X-ray should be performed as a diagnostic test, with further imaging tests performed if necessary and available. When the diagnosis of macrodystrophia lipomatosa is confirmed, management strategies comprise observation only or surgery followed by observation. Presentation of macrodystrophia lipomatosa before puberty, or with single digit involvement, or with concurrent urgent medical conditions calls for observation only. If these conditions are absent, surgery can be performed with subsequent observation for outcome and recurrence. EMG, electromyography; CT, computed tomography; MRI, magnetic resonance imaging; USG, ultrasonography. [a]They usually manifest as macrodactyly without adipose tissue overgrowth; [b]Both esthetic and functional.



surgery in his childhood, and thus the first presenting age was actually younger. The last two cases are also younger; they presented due to cosmetic and functional difficulties. Parents might have sought treatment earlier due to the need for the affected limbs to look 'normal' before the children started school. The distribution of the affected digits in these patients is similar to the findings of the review, which were that MDL occurs in multiple adjacent digits with second digit involvement. After radiographic examinations were performed, surgery planning resulted in single-staged surgery for all patients. However, due to technical reasons during the surgery, the 6-year-old female case received treatment in a multi-staged surgical manner. Joint management, rather than bony intervention, was performed by rebalancing the joint ligament. Ray amputation in the second case was performed with the consideration of removing ineffective digits to achieve better functional outcome. This decision was also in line with the findings of this review. Furthermore, from all three cases, only one patient had a complication of delayed wound healing. This can be fixed with a skin graft of the area. Impressively, all cases had positive outcomes, which might rationalize the decision of management made by the first author.

There are several limitations to this study. Level of evidence is an important aspect of this study. As it only uses case series and case reports, its evidence level is a four, the lowest level except

for that of an expert opinion. Thus the evidence we present is not as strong as a cohort or case-control study. However, it is difficult to conduct this type of studies in surgical settings due to ethical issues. Thus, this study can be considered sufficiently relevant to be used. Other weaknesses might be a bias during article selection. Incomplete information from the article title and abstract might infer different meanings of the real condition of the patients than can be clearly shown in full-text articles. Additionally, only a single database was used to find the relevant articles. Any studies that were not indexed by the database could have been missed. In addition, the inability to add more cases from the author and new cases from journal articles due to period-related inclusion criteria, might have caused many very recent cases of MDL to be neglected and excluded from this study.

In relation to the diagnosis and management of MDL, this study suggests several things. The use of non-invasive diagnostic tools should be carefully considered, according to the severity of the condition. Surgery might be a better choice of management than observation, taking into account future complications in the absence of surgery and the beneficial outcomes of surgical procedures for patients. Patients should be followed up regularly to determine the incidence of reoccurrence. Although an algorithm is proposed in this study, it has not been tested yet; its accuracy and efficiency might therefore still be questionable.

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Appendix 1. The summary of age, gender, and anatomical location of the inclusive articles

No.	Author(s)	Year	No. of cases	Age (yr)	Gender	Anatomical location
Upper extremities						
1	Yesilada et al. [1]	2013	1	42	Male	Right upper extremity
2	Kwon et al. [2]	2013	1	14	Male	Thumb, index, and thenar area of left hand
3	Cöloğlu [3]	2013	1	28	Male	Fourth finger of the left hand
4	van der Meer et al. [4]	2011	1	2 mo	Male	Ring and little fingers of his left hand and the middle and ring fingers of the right hand
5	Rohilla et al. [5]	2012	1	20	Female	Thumb, index and middle fingers of the left hand, lateral aspect of the left hand, and the left shoulder
6	Gao et al. [6]	2010	1	14	Female	Ring finger of the right hand and right upper extremity
7	Chiang et al. [7]	2010	1	28	Male	Middle finger of the left hand
8	Singla et al. [8]	2008	1	2	Male	Dorsal and palmar aspects of the fourth and fifth digits of the left hand
9	Mahafza et al. [9]	2008	1	1	Female	Index, middle and ring fingers of the right hand
10	D'Costa et al. [10]	1996	1	34	Male	Thumb, index finger and radial half of the right hand
11	Ho et al. [11]	2007	1	6	Male	Index and long fingers of the left hand
12	Brodwater et al. [12]	2000	1	3	Male	Middle finger of the left hand
13	Meyer and Röricht [13]	1997	1	35	Female	Fourth finger
14	Boren et al. [14]	1995	1	42	Male	Thumb of the right hand
15	Pearn et al. [15]	1983	1	12	Male	Index finger, palm, forearm of the left side
16	Goldman and Kaye [16]	1977	2	5	Female	Second and third digits of the hand
				N/A	Male	Second digit of the hand
17	Ranawat et al. [17]	1968	1	55	Male	Thumb, index, and long fingers of the left hand; the palm and the lower one third of the left forearm
Lower extremities						
1	Ceylan and Tuzuner [18]	2013	1	23	Female	Second toe of right foot
2	Sudesh et al. [19]	2012	6	3	Male	Second toe
				3	Female	Second toe
				2	Male	Second toe
				4	Male	Third toe
				5	Male	Second toe
				4	Female	Third toe
3	Upadhyay et al. [20]	2011	1	22	Male	Second, third, and fourth toes of the left foot
4	Guzoglu et al. [21]	2012	1	1 day	Male	Second, third, and, fourth toes of the right foot and the dorsal aspect of the right foot
5	Kozanoglu et al. [22]	2008	1	48	Male	Great toe of the right foot, right ankle and cruris
6	Ho et al. [11]	2007	1	4 mo	Female	Second and third toes on the left foot
7	Tuzuner et al. [23]	2005	1	64	Male	First, second, and third toes of the right foot
8	Oztürk et al. [24]	2004	1	40	Male	First toe of the right foot
9	Watt and Chung [25]	2004	1	1 mo	Female	Great, second, and third toes and the plantar aspect of the right foot
10	Wang et al. [26]	1997	1	10	Female	Plantar surface of the second digit of the right foot
11	Soler et al. [27]	1997	1	8 mo	Male	Distal ends of the second and third digits and left foot
12	Viola et al. [28]	1991	1	3 mo	Male	Marked hypertrophy of the first and second rays of the right foot and mild hypertrophy of the entire medial portion of the right lower limb
13	Bansal and Harmit [29]	1989	1	6	Male	Left lower limb
14	Curry et al. [30]	1988	1	52	Female	Second and third toes of the left foot
15	Baruchin et al. [31]	1988	1	2	Female	Gigantism of the first, second, and third toes, with syndactyly of the second and third toes
16	Moran et al. [32]	1984	1	18	Female	Second, third and fourth toes of the left foot
17	Goldman and Kaye [16]	1977	4	N/A	Male	Second, third, and fourth digits of the foot
				N/A	Male	Second and third digits of the foot
				N/A	Male	Second digit of the foot
				44	Male	Fourth digit of the foot

Appendix 2. The summary of length of follow-up, diagnostic tools, management, complications, and outcomes of the inclusive articles

No.	Author(s)	Year	No. of cases	Age (yr)	Length of follow-up	Diagnostic tools	Intervention	Compli-cations	Outcome	
									Esthetic	Functional
Upper extremities										
1	Yesilada et al. [1]	2013	1	42	N/A	N/A	Amputation of the thumb 4 years ago. Resection of hypertrophic carpal bone and debulking of large soft tissue mass.	N/A	N/A	Unspecified improvement
2	Kwon et al. [2]	2013	1	14	N/A	X-ray and MRI	Previous debulking surgery twice Debulking operation of the thumb and amputation of the index finger at the mid-level of the mid-phalangeal bone	Some degree of sensory loss in the first web space	Unsatisfied	Significant improvement
3	Cöloğlu [3]	2013	1	28	18 mo	X-ray and MRI	Reconstruction of the fourth finger, intraneural fascicular dissection and limited excision of the ulnar nerve with extension to the digital nerve of the ring finger.	None	Satisfied	Fair improvement
4	van der Meer et al. [4]	2011	1	2 mo	42 yr	X-ray	12-years-old: amputation of the 4th and 5th ray of the left hand and removal of lipoma.	N/A	N/A	Left = unspecified improvement, Right = no improvement
5	Rohilla et al. [5]	2012	1	20	N/A	X-ray and MRI	Debulking and partial amputation of the overgrown digits (middle phalanges of the 2nd and 3rd digits) with preservation of distal phalanges.	N/A	N/A	N/A
6	Gao et al. [6]	2010	1	14	18 mo	X-ray and MRI	Debulking of the right upper limb, amputation of the right ring finger and the little finger	None	Satisfied	Significant improvement
7	Chiang et al. [7]	2010	1	28	N/A	X-ray, MRI, and nerve conduction study	4-years-old: amputation of the middle finger for cosmetic reasons. 25-years-old: median nerve release.	4 yo: numbness sensation over the palm. 25 yo: recurrent symptoms	N/A	N/A
8	Singla et al. [8]	2008	1	2	N/A	X-ray and MRI	Debulking and reconstruction of the fingers	N/A	N/A	N/A
9	Mahafza et al. [9]	2008	1	1	N/A	X-ray, MRI, and electrophysiologic exam	Debulking and release of syndactyly	N/A	N/A	N/A
10	D'Costa et al. [10]	2007	1	34	N/A	N/A	Amputation of the right thumb and debulking of the palm	N/A	N/A	N/A
11	Ho et al. [11]	2007	1	6	3 yr	Serial X-rays	6-years-old: amputation of the index and middle digits with shortening of the radial digital nerve to the ring finger and reconstruction of the ulnar digital nerve to the thumb with use of the dorsal radial sensory branch. 9-years-old: decline surgery	N/A	N/A	Fair improvement
12	Brodwater et al. [12]	2000	1	3	> 2 yr	Serial MRI	Exploration of the lesion and debulk for cosmetic reasons with open investigation of the median nerve. A carpal-tunnel release with neurolysis of the median nerve and radial and ulnar digital nerves of the middle finger with excision of the adjacent excess fatty tissue.	None	N/A	N/A
13	Meyer and Röricht [13]	1997	1	35	N/A	MRI, EMG, and nerve conduction studies	3-years-old: amputation of the fourth finger	N/A	N/A	N/A
14	Boren et al. [14]	1995	1	42	N/A	X-ray and MRI	Bone and soft tissue reduction in the thumb and carpal tunnel release	N/A	N/A	N/A
15	Pearn et al. [15]	1983	1	12	N/A	Not stated	Observation until puberty	Not applicable	Not applicable	Not applicable
16	Goldman and Kaye [16]	1977	2	5 N/A	7 yr N/A	Serial X-rays X-ray	Debulking Unknown surgery	N/A N/A	N/A N/A	N/A N/A

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Appendix 2. Continued

No.	Author(s)	Year	No. of cases	Age (yr)	Length of follow-up	Diagnostic tools	Intervention	Compli-cations	Outcome	
									Esthetic	Functional
17	Ranawat et al. [17]	1968	1	55	2 yr	X-ray, hemogram, urinalysis, night peripheral blood-smear studies for microfilaria bancrofti larvae, arteriogram	Lipoma excision, amputation of the index finger, debulking of the thumb	None	N/A	Significant improvement
Lower extremities										
1	Ceylan and Tuzuner [18]	2013	1	23	None	X-ray and MRI	Amputation of the second toe	N/A	N/A	Significant improvement
2	Sudesh et al. [19]	2012	6	3	At least 4 wk	X-ray and USG	Debulking and amputation of distal phalanx	None	N/A	Significant improvement
				3				Skin blackening		Significant improvement
				2				Undercorrection		No improvement
				4				Skin blackening		Significant improvement
				5				Skin blackening		Significant improvement
				4				None		Significant improvement
3	Upadhyay et al. [20]	2011	1	22	N/A	X-ray, USG, and CT scan	Childhood: surgery on the left foot. 22-years-old: debulking with orthotic supplementation, a trans metatarsal amputation	N/A	N/A	N/A
4	Guzoglu et al. [21]	2012	1	1 mo	14 mo	X-ray and MRI	7-month-old: debulking	N/A	N/A	Unspecified improvement
5	Kozanoglu et al. [22]	2008	1	48	N/A	X-ray, MRI, and electrophysiological analysis	Correction of the right great toe surgically	N/A	N/A	N/A
6	Ho et al. [11]	2007	1	4 mo	> 20 yr	Serial X-rays,MRI, and scanograms	6-month-old: amputation of the second and third. 9-month-old: debulking. 5-years-old: debulking of the central portion of the foot and liposuction. 12-years-old: amputation of the remaining left toes, left distal femoral epiphysiodesis. 13-years-old: syme amputation with left calf debulking; left buttock and thigh debulking few weeks later; proximal osteotomy to shorten the tibia by 4 cm.	N/A	Satisfied	Significant improvement
7	Tuzuner et al. [23]	2005	1	64	N/A	X-ray, CT scan, and angiography	Refused surgery	Not applicable	Not applicable	Not applicable
8	Oztürk et al. [24]	2004	1	40	2 yr	X-ray, USG, and CT scan	Partial debulking and reconstruction of the first right toe.	None	N/A	Significant improvement
9	Watt and Chung [25]	2004	1	1 mo	3 yr	Not stated	14-month-old: modest debulking of the dorsal and plantar aspects, and the amputation of the syndactylous second/third toe unit. 18-month-old: amputation and debulking procedures of the great toe, second and third metatarsals. The underlying soft tissue was excised and the flaps were rotated and advanced to close the midfoot cleft. 2-years-old: debulking of the plantar surface of the foot to improve the aesthetic results.	None	Satisfied	Significant improvement
10	Wang et al. [26]	1997	1	10	N/A	X-ray and MRI	Multiple: amputation of second distal proximal phalanx	N/A	N/A	N/A
11	Soler et al. [27]	1997	1	8 mo	27 yr	Serial X-rays and MRI	8 mth: amputation of the left foot, and the distal ends of the second and third digits by the disproportional growth of his digits. 27-years-old: surgical reconstruction of the left foot.	N/A	N/A	N/A

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Appendix 2. Continued

No.	Author(s)	Year	No. of cases	Age (yr)	Length of follow-up	Diagnostic tools	Intervention	Compli-cations	Outcome	
									Esthetic	Functional
12	Viola et al. [28]	1991	1	3 mo	10 yr	X-rays, serial laboratory examination, renal USG, IV urograms, arteriography of the lower limbs and kidneys, and 13 bone age analysis	First year: thinning and shortening osteotomies of the 1st and 2nd proximal phalanges and corresponding metatarsals, debulking of the entire medial foot, and a 30 varus osteotomy of the distal tibia. Second year: distal femoral varus osteotomy, debulking of the groin to the medial malleolus, and a distal tibialvarus shortening osteotomy with excision of the lateral third of the Achilles tendon and release of the lateral ankle ligaments. 2-years 7-month-old: amputation of the first and second toes. 3-years 8-month old: syme amputation. 5-years-old: distal femoral and proximal tibial epiphyseal closing wedge osteotomies. 9-years-old: a varus subtrochanteric osteotomy surgery and liposuction of the proximal medial right lower limb (395 g mature adipose tissue).	None	N/A	Significant improvement
13	Bansal and Harmit [29]	1989	1	6	3 yr	X-ray	A mid-thigh amputation	None	N/A	Fair improvement
14	Curry et al. [30]	1988	1	52	None	X-ray and CT scan	Amputation of the distal portion of the second toe	N/A	N/A	N/A
15	Baruchin et al. [31]	1988	1	2	1 yr	X-ray	Debulking and removal of the hypertrophied toes, with ephysiodesis of the metatarsals	None	N/A	Significant improvement
16	Moran et al. [32]	1984	1	18	N/A	X-ray	1-year-old: amputation of the terminal phalanx of the third toe and osteotomy of the proximal phalanx of the second toe.	N/A	N/A	N/A
17	Goldman and Kaye [16]	1977	4	N/A N/A N/A 44	N/A N/A N/A N/A	X-ray X-ray X-ray X-ray	Unknown surgery Unknown surgery Unknown surgery Observation	N/A N/A N/A Not applicable	N/A N/A N/A Not applicable	N/A N/A N/A Not applicable

N/A, not available; MRI, magnetic resonance imaging; EMG, electromyography; USG, ultrasonography; CT, computed tomography.

Appendix 3. Digital presentation of Macrodystrophia lipomatosa in upper extremity

	Authors	1	2	3	4	5	Addition	Side
1	Yesilada et al. [1]			Upper extremity				R
2	Albright et al. [33]			Upper extremity				B
3	Kwon et al. [2]	+	+	–	–	–	Thenar	L
4	Cöloğlu [3]	–	–	–	+	–	–	L
5	van der Meer et al. [4]	–	–	–	+	+	–	LB
		–	–	+	+	–	–	RB
6	Rohilla et al. [5]	+	+	+	–	–	Shoulder	L
7	Gao et al. [6]	–	–	–	+	–	Upper extremities	R
8	Chiang et al. [7]	–	–	+	–	–	–	L
9	Singla et al. [8]	–	–	–	+	+	–	L
		+	+	–	–	–	–	R
10	Dillman and Strouse [34]	–	+	+	–	–	–	L
11	Mahafza et al. [9]	–	+	+	+	–	–	R
12	D'Costa et al. [10]	+	+	–	–	–	–	R
13	Ho et al. [11]	–	+	+	–	–	–	L
14	Turkington and Grey [35]	+	–	–	–	–	–	R
15	Sone et al. [36]			Hand and forearm				N/A
				Hand and forearm				N/A
16	Brodwater et al. [12]	–	–	+	–	–	–	L
17	Di Ianni et al. [37]	+	+	+	–	–	Forearm	R
18	Meyer and Röricht [13]	–	–	–	+	–	–	N/A
19	Wang et al. [26]			Forearm				R
20	D'Costa et al. [38]			Hand				R
21	Loro et al. [39]			Hand				L
22	Boren et al. [14]	+	–	–	–	–	–	R
23	Hildebrandt et al. [40]			Wrist and Hand				R
24	Jain et al. [41]			Upper limb and shoulder				R
25	Blacksin et al. [42]	–	+	–	–	–	Upper arm	R
26	Pearn et al. [15]	–	+	–	–	–	Forearm	L
27	Laval-Jeantet et al. [43]	+	+	–	–	–	–	L
28	Goldman and Kaye [16]	–	+	+	–	–	–	N/A
		–	+	–	–	–	–	N/A
29	Yaghmai et al. [44]			Hand			–	L
				Hand			–	L
				Hand			–	R
30	Ranawat et al. [17]	+	+	+	–	–	Forearm	L
31	Prasetyono TOH, Hanafi E, Astriana W[a]	+	+	–	–	–	Thenar	R
		11	16	10	7	2		15R, 14L, 2B, 5N/A

R, right; B, bilateral; L, left; LB, left bilateral; RB, right bilateral; N/A, not available.
[a]This study.

Appendix 4. Digital presentation of macrodystrophia lipomatosa in lower extremity

	Authors	1	2	3	4	5	Addition	Side
1	Ceylan and Tuzuner [18]	–	+	–	–	–	–	R
2	Sudesh et al. [19]	–	+	–	–	–	–	N/A
		–	+	–	–	–	–	N/A
		–	+	–	–	–	–	N/A
		–	–	+	–	–	–	N/A
		–	+	–	–	–	–	N/A
		–	–	+	–	–	–	N/A
3	Koplay et al. [45]	+	–	–	–	–	–	L
4	Upadhyay et al. [20]	–	+	+	+	–	–	L
5	Guzoglu et al. [21]	–	+	+	+	–	Dorsal	R
6	Khan et al. [46]	+	+	+	–	–	–	R
		–	+	–	–	–	–	R
		–	+	+	–	–	–	L
		–	+	+	–	–	–	L
7	Kozanoglu et al. [22]	+	–	–	–	–	Ankle, cruris	R
8	Pandey [47]			Foot				R
9	Fritz and Swischuk [48]	+	+	+	+	–	–	R
10	Ho et al. [11]	–	+	+	–	–	–	L
11	Tuzuner et al. [23]	+	+	+	–	–	–	R
12	Oztürk et al. [24]	+	–	–	–	–	–	R
13	Watt and Chung [25]	+	+	+	–	–	–	R
14	Ly and Beall [49]			Forefoot				L
15	Verma and Yadu [50]	–	+	–	–	–	–	R
16	Aydos et al. [51]			Foot				B
17	Wang et al. [26]	–	+	–	–	–	–	R
				Plantar and dorsal				R
18	Soler et al. [27]	–	+	+	–	–	–	L
19	Viola et al. [28]	+	+	–	–	–	Lower limb	R
20	Bansal and Harmit [29]			Lower limb				L
21	Curry et al. [30]	–	+	+	–	–	–	L
22	Baruchin et al. [31]	+	+	+	–	–	–	L
23	Moran et al. [32]	–	+	+	+	–	–	L
		–	+	–	–	–	–	R
24	Goldman and Kaye [16]	–	+	+	+	–	–	N/A
		–	+	+	–	–	–	N/A
		–	+	–	–	–	–	N/A
		–	–	–	+	–	–	N/A
25	Yaghmai et al. [44]			Foot				R
				Foot				B
26	Prasetyono TOH, Hanafi E, Astriana W[a)]	–	–	–	+	+	Calcaneal, Achilles	R
		–	+	+	+	–	–	L
		9	27	18	8	1		17R,12L,2B,10N/A

R, right; B, bilateral; N/A, not available; L, left.
[a)]This study.

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