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Macrodystrophia lipomatosa (Gigantism)-illustration of three cases

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Abstract
Macrodystrophia lipomatosa is localized hypertrophy of a limb or a segment of a limb leading to Gigantism and is a rare cause of congenital macrodactyly, with a disproportionate increase in fibroadipose tissue. It is more common in the foot than in the hand. We present three cases of macrodystrophia lipomatosa (MDL) of the lower limb followed by their surgical management. The first case is a 6 year old female child who presented with bulky pigmented right leg and thigh. We managed this case by growth modulation using a figure of 8 plate. The second case is a 1 year old female child with gigantism of right foot and right leg that was managed by debulking procedure and epiphysiodesis. The third case is of a 9 year old female child with the macrodactyly of the second and third toe on the right side. Epiphysiodesis was done for the second and third toe on the right side.

Keywords: Macrodystrophia lipomatosa, debulking, gigantism, figure of 8 plate, epiphysiodesis

Introduction
Macrodystrophia lipomatosa (MDL) is characterized by the progressive proliferation of all mesenchymal elements and is a rare cause of macrodactyly [1]. It is reportedly more common in the foot than in the hand [1]. Feris first described this condition in 1925. Macrodystrophia lipomatosa is a rare, congenital, nonhereditary form of localized gigantism [2]. It can present anytime from infancy to late adulthood but it is usually recognised at birth [3]. We are presenting a series of three cases of MDL and their outcome after surgery.

Case Presentation
The first case is a 6 year old female child who presented with limb length discrepancy and bulky pigmented right leg and thigh. There is a leg length discrepancy of 5.5 cm on the right side and both femur and tibia are longer. The girth of the thigh was 3 cm more on the affected side and the girth of the calf was 1.5 cm more on the affected side. Excessive hairs are present on the right leg. Multiple pigmented spots are present on the right leg. The right knee was in valgus of 20 degrees and there was anterior bowing of the right tibia. X-ray findings show an increase in neck shaft angle to 160 degrees and an increase in the length of both the femur and tibia. The clinical picture at the time of presentation is being shown in the following figures (Figure 1a, Figure 1b, Figure 1c). X ray images at the time of the first presentation is shown in the following figures (Figure 2a, Figure 2b, Figure 2c). We did growth modulation using a figure of 8 plate on the medial side of lower femoral and upper tibial epiphysis (Figure 3a, Figure 3b, Figure 3c) to correct the knee valgus. It was followed by figure of 8 plating done for the lateral aspect of lower femoral and upper tibial epiphysis after one and a half years for growth arrest after the knee valgus got corrected (Figure 4a, Figure 4b). There was figure of eight plates on both the medial and lateral side for one and a half year. It was followed by removal of medial plates after one and a half year as the knee started going into varus (Figure 6a). It was followed by the removal of lateral plate after one year. After three years thigh girth has reduced by 2 cm and the difference is 1 cm. The calf diameter has been reduced by 1 cm. Skin pigmentation has reduced (Figure 5a, Figure 5b, Figure 5c). A heel raise is no longer required on the normal side. Why the girth has decreased and pigmentation has improved cannot be explained but this has occurred after epiphysiodesis. The second case is a 1 year old female child with gigantism of right foot and right leg.

The patient presented with a large size of the right foot and right leg noticed by the parents at birth. On examination second, third and fourth toes are much larger. Foot is bulky because of excessive fibrofatty tissue on the dorsum and plantar aspect of the foot. X ray shows overgrown second, third and fourth metatarsals and overgrown second, third and fourth proximal phalanges. The right lower limb is longer by 2 cm (Figure 7a, Figure 7b). We removed bulky fibrofatty tissues through a transverse incision on the dorsum of the foot. Second, third and fourth metatarsals were exposed distally and epiphysis identified and curetted (Epiphysiodesis). Second, third and fourth proximal phalanx exposed proximally and epiphysis curetted (Epiphysiodesis). Wound was closed and a bilateral posterior splint was applied. After four months, using vertical incision, fibrofatty tissue on the plantar aspect of the foot was excised (Figure 9a, Figure 9b, Figure 9c, Figure 9d). It was followed by the removal of the head of the second and forth metatarsals after one year (Figure 10a).

The third case is a 9 year old female child with the macrodactyly of the second and third toe on the right side. Removal of the base of the proximal phalanx was done for the second and third toe on the right side.

Discussion

MDL is a congenital localized gigantism involving the digits or limbs. The lower limb is more frequently involved than the upper limb. The abnormal area is usually along a specific sclerotome. The exact etiology of MDL is unknown. Various hypotheses exist. It could be due to a variable form of neurofibromatosis, in-utero disturbance of growth limiting factor, disturbed fetal circulation [4]. It is usually associated with syndactyly, brachydactyly, polydactyly or clinodactyly [4]. A study done by Pebam Sudesh *et al*. shows the simple debulking procedure is sufficient for MDL [3]. Males and females are equally affected, however in our study all three patients are female child [3]. The problem that occurs in a patient with MDL is usually cosmetic or functional. Compression of neurovascular structures or secondary osteoarthritis can arise that can lead to hindrance in the function [3]. A study done by Andrew J Watts *et al*. advised multiple reconstructive procedures including debulking of dorsal and plantar surfaces of the foot and amputation [5]. A balance between functional and aesthetic outcome is required in cases of MDL [5]. Surgical treatment mainly relies on midtarsal and midcarpal amputations [5]. Below-knee amputation have been reported as successful and necessary procedures [6]. Study done by Tamara A Topoleski *et al*. shows proximal phalangeal epiphysiodesis as a good modality for the treatment of macrodactyly of the toe [7].

The various differential diagnosis of MDL includes neurofibromatosis type1, Maffuci syndrome, Ollier's syndrome, fibrolipomatous hamartoma, Klippel-Trenaunay-Weber syndrome. There is a positive family history in all of these differential diagnosis.

Depending upon the patient's symptoms, age, extent and severity of the disease, surgery is indicated. Removal of a ray or debulking and toes or finger amputation is the most appropriate procedure in a localized form of a disease that can lead to cosmetic improvement without causing any neurological problems [5]. Epiphysiodesis, debulking procedure and various osteotomies are indicated for more severe form of the disease. We did debulking and epiphysiodesis procedure in our second case and epiphysiodesis in our third case. Surgical procedures can lead to skin necrosis that can take time to heal. The disease can recur in the affected extremity. Macrodystrophia lipomatosa should be managed early in children to prevent disproportionate growth so that functional disability can be kept to the minimum.

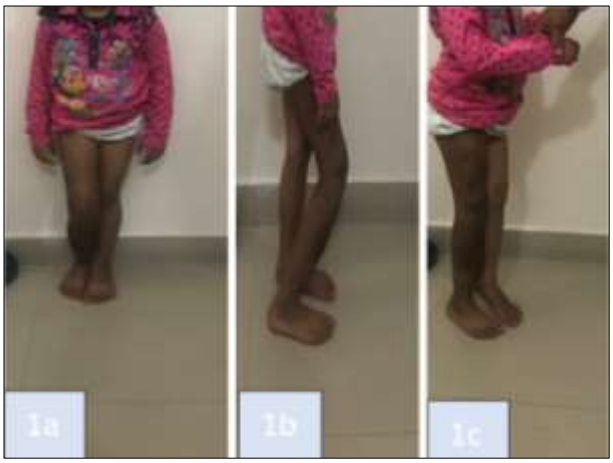

Fig 1: Clinical picture at the time of presentation is being shown in the following figures (Figure 1a, Figure 1b, Figure 1c)

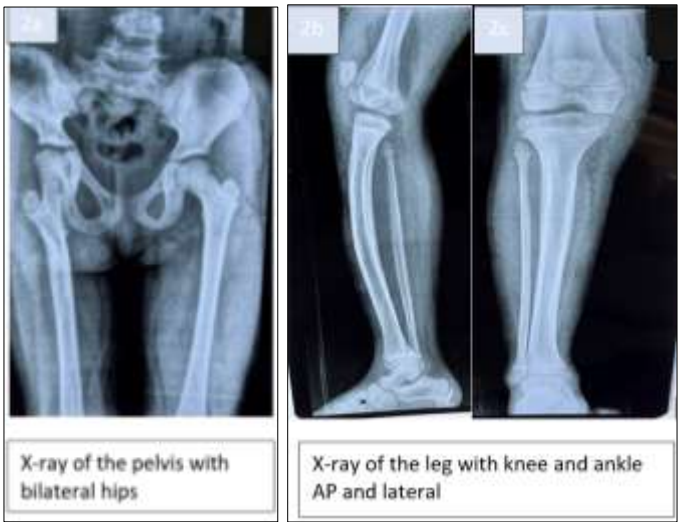

Fig 2: X-ray findings at the time of the first presentation. (Figure 2a, 2b and 2c)

Fig 3: Figure showing X-ray and Carm images showing medial epiphsiodesis using a figure of eight plate (Figure 3a, 3b and 3c)

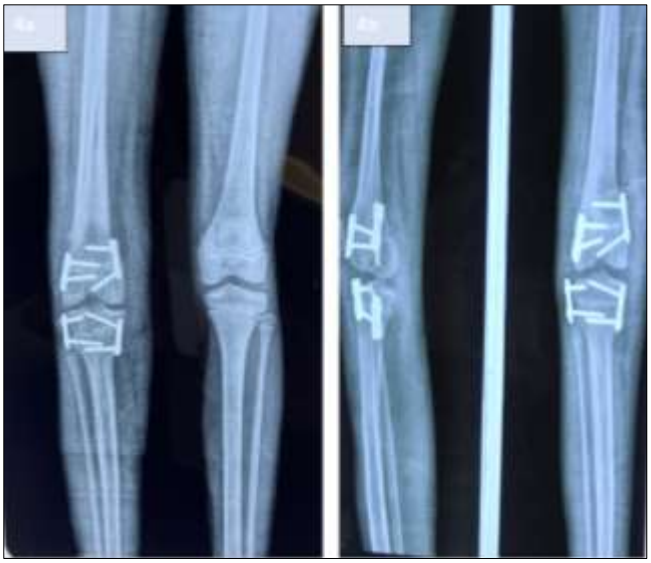

Fig 4: Showing X-ray shows figure of 8 plating done for lateral aspect of lower femoral and upper tibial epiphysis after one and a half year when the valgus got corrected

Fig 5: Showing clinical picture after 3 years

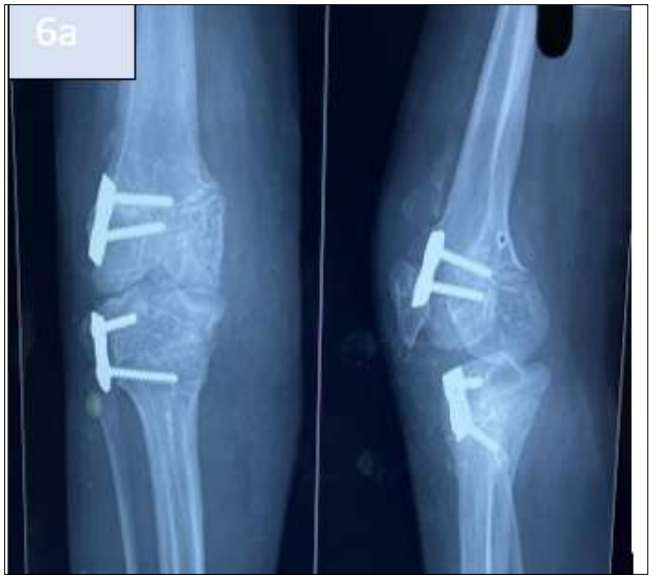

Fig 6: Showing X-ray picture after removal of medial plate to correct varus deformity

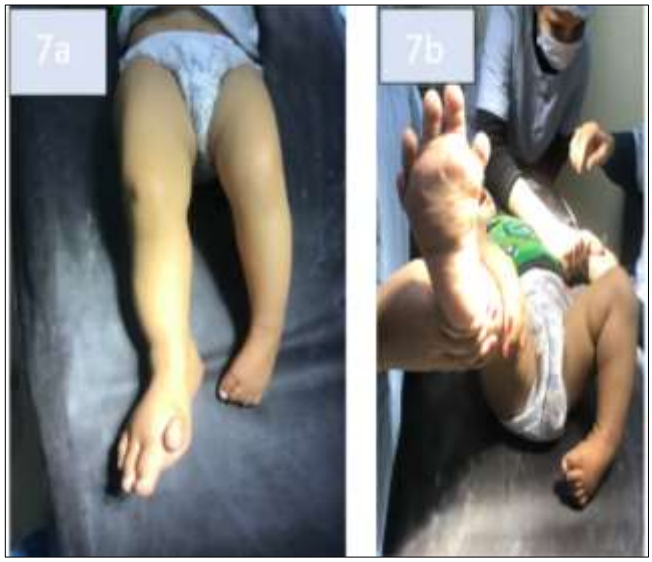

Fig 7: Showing clinical picture at the time of presentation

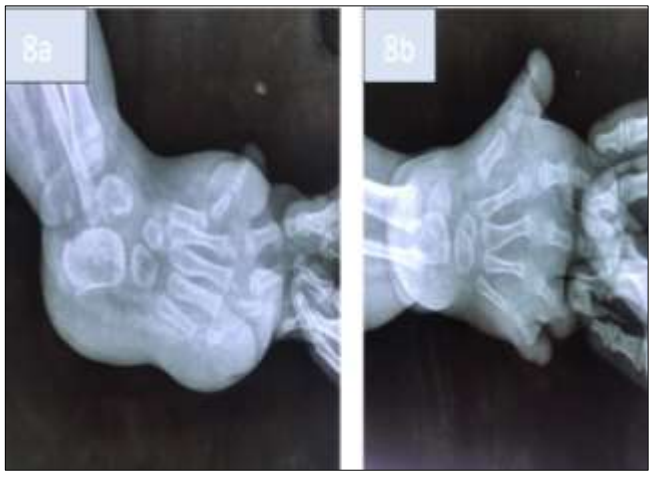

Fig 8: Shows X-ray images at the time of presentation

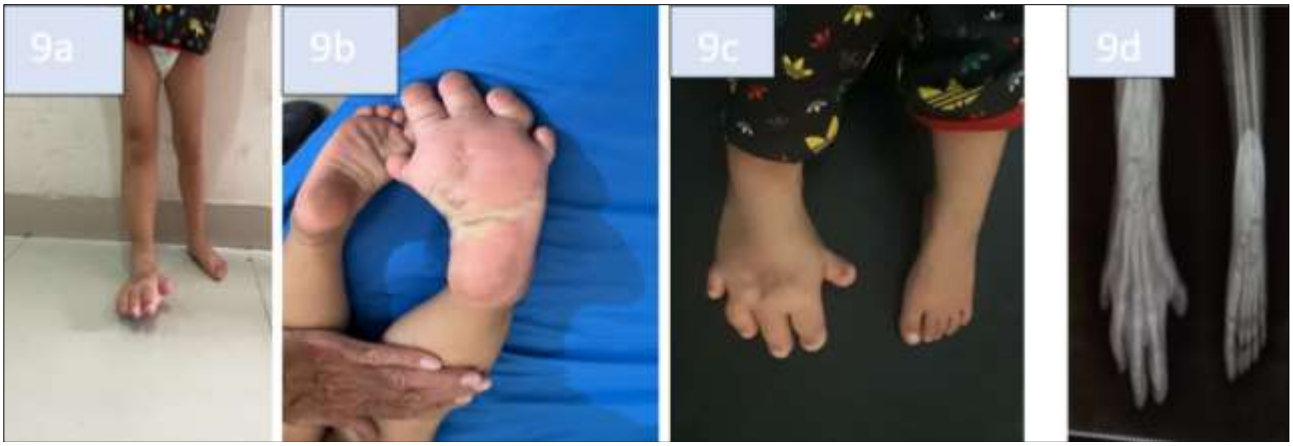

Fig 9: Shows Clinical images and x-ray findings after one and a half year

Fig 10: Shows clinical picture after final procedure

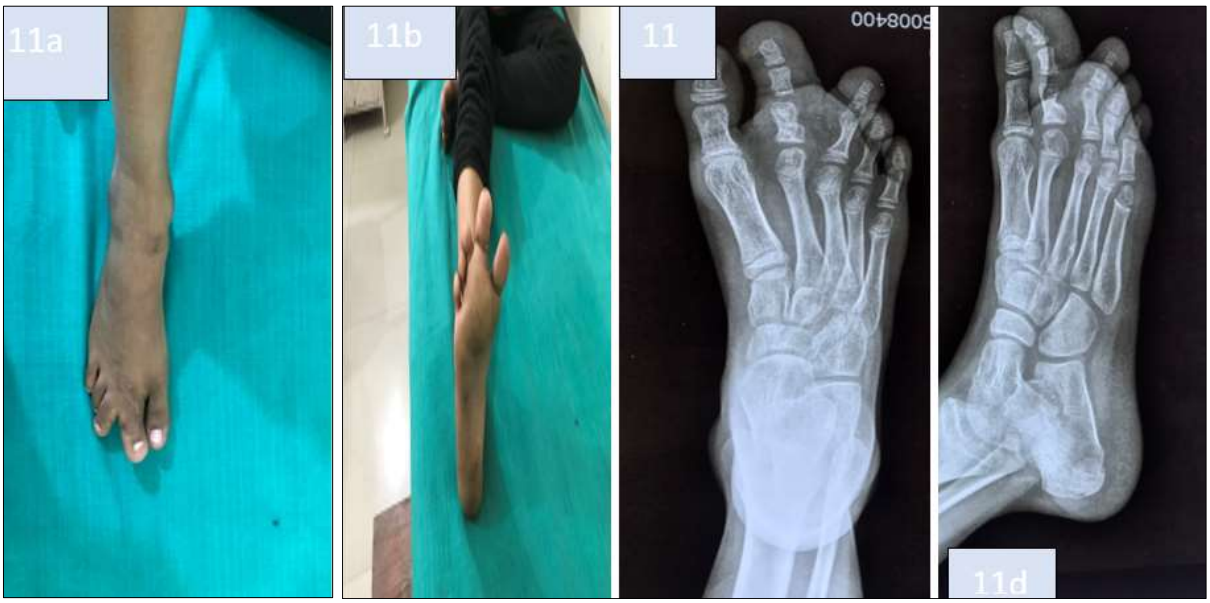

Fig 11: Shows Clinical picture and x-ray

Conclusion

Early management and frequent follow-up in a child with MDL is the key to contain the size of the limb or a part of it and thereby minimizing the chances of disproportionate and uneven growth of the limb that may eventually end up in an amputation. Multiple procedures are often required. Diagnosis is accomplished on the basis of clinical and radiological evaluation. The management is mainly surgical. Often multiple surgical procedures are required.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the Journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil.

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