DOI: 10.7860/JCDR/2016/23455.8875

Letter to Editor

Dermatology Section

Gigantic Hand: A Rare Case of Macrodystrophia Lipomatosa

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Keywords: Debulking surgery, Hypertrophied digit, Soft tissue enlargement

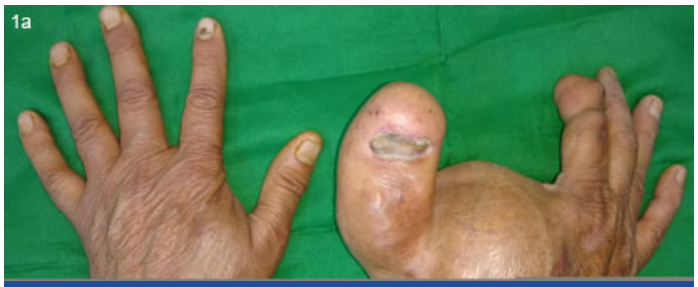
Dear Editor,

We read with great pleasure the recent paper wrote by Durairaj et al., published in your esteemed journal [1]. We hereby, share our experience of a similar case with different disease course and management thereby adding to reader's knowledge related to the topic.

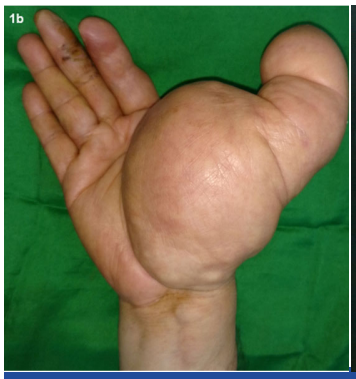
A 65-year-old male recently came to us for his very insidiously enlarging right hand. As per his history, the swelling was first noticed by him approximately fifteen years ago. Patient, a farmer by occupation had no problems working in field and hence chose not to seek any medical consultation till now. The swelling increased gradually to present size over a decade. During this period there were no neurological symptoms (related to nerve compression).

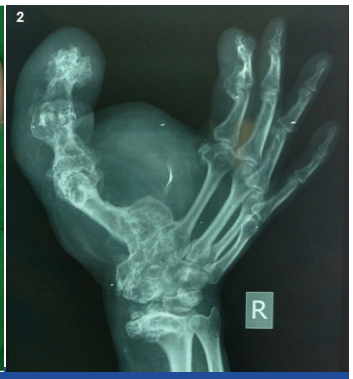
On examination, we found the right hand to be diffusely swollen, hypertrophied thumb and index finger leading to widening of first web space [Table/Fig-1a&b]. The swelling was soft in consistency, non-tender and there was no bruit. FNAC from the swelling showed lobules of mature adipocytes. On imaging, the radiograph showed hypertrophy of soft tissues and right thumb, with enlargement of right index finger, carpal bones and distal radius [Table/Fig-2]. MRI revealed focal hypertrophy of thumb and index finger due to soft tissue proliferation with areas of fatty infiltration and enlargement of median nerve. The characteristic clinical and imaging features, lead to the diagnosis of Macrodystrophia Lipomatosa (MDL). Patient was counselled regarding the insidious nature of disease and treatment options. He opted for observation despite having difficulty in his daily activities.

MDL is a rare non-hereditary congenital form of localized gigantism of fingers or toes, characterized by an increase in all mesenchymal elements, particularly fibro-adipose tissue [2]. In upper limb, the area of predilection is the segment supplied by the median nerve. The author's case bears similarity with that reported by Celebi et al., [1,3]. Both cases have a very young age of presentation, lower limb involvement and "progressive type" of disease. Contrarily, our case is very similar to another recently reported case by Maheswari et al., with upper limb involvement, older age of presentation and symptoms of over a decade [4]. Unfortunately, patient reported by Maheswari et al., had disease recurrence despite debulking surgeries done twice earlier. Hence, that time patient opted for limb amputation with artificial prosthesis placement [4]. As evident from above reported four cases, MDL can be either static or progressive, and the reasons for seeking consultation are mainly due to cosmetic issues, mechanical problems or development of neurovascular compression. Debulking surgery is indicated depending upon the extent and severity of the disease. But it often recurs in the affected extremity producing significant impact on patient morbidity [4,5] [Table/Fig-3] [1,3,4].



[Table/Fig-1a]: Diffuse swelling of the right hand, hypertrophied thumb and index finger leading to widening of first web space (Dorsal view).





[Table/Fig-1b]: Diffuse swelling of the right hand, hypertrophied thumb and index finger leading to widening of first web space (Ventral view).
[Table/Fig-2]: X-ray of the right hand.

Author	Age	Sex	Area of Involvement	Symptom Duration	Nerve Compression	Treatment Given	Follow-up
Durairaj et al., [1]	5 year	-	Left second toe	Difficulty in walking and wearing foot wear. Mass present since birth	Nil	Toe reduction surgery, partial amputation of destroyed bone with skin flap placement	No recurrence
Celebi et al., [3]	9 month	Male	Right second and third digit	Since birth	Nil	Excision of excess skin and adipose tissue and bone reconstruction surgery	No recurrence
Maheswari et al., [4]	20 year	Male	Involvement of entire left upper limb predominantly the radial aspect	Since childhood	Nil	Underwent limb reduction surgeries twice earlier. This time due to previous failed surgeries, he opted for left upper limb amputation above the elbow with prosthesis placement	Follow-up
Present case (Sahu et al.,)	65 year	Male	Right hand	15 years	Nil	Observation	On follow-up

[Table/Fig-3]: Recently reported cases of Macrodystrophia Lipomatosa (MDL), varied presentations, area involvement with treatment and follow-up [1,3,4].

This letter aims to make readers aware of varied spectrum of presentation with which this disease can present and it's the magnitude of enlargement, patient's preference and morbidity which plays a pivotal role in deciding the therapy.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Aug 09, 2016**
Date of Peer Review: **Aug 18, 2016**
Date of Acceptance: **Aug 18, 2016**
Date of Publishing: **Nov 01, 2016**