Localized Gigantism of Foot: Macrodysrophia: Lipomatosa

Mehmet Erdil[1], Hasan Huseyin Ceylan[1]

[1]*Department of Orthopaedics and Traumatology, Bezmialem Vakif University, Istanbul, Turkey*

A 19-month-old female child was brought by her parents with an abnormal appearance of her foot. Her parents reported her left foot to be larger than her right one since birth [Figure 1, 2]. Detailed clinical history revealed that the swellings were present since birth and had gradually increased to its present size. No positive family history was noted. No cutaneous skin lesions, edema or bruits were present on her foot and leg. A radiograph of the right foot revealed diffuse enlargement of the 2nd, 3rd and 4th metatarsals and phalanges of the left foot both in length and transverse diameter, with surrounding soft tissue enlargement and degenerative changes of the joints [Figure 3]. MRI examination showed abundant fatty infiltration of the soft tissues and cortical thickening of bones.

Histopathologic examination of the biopsied specimen found hypertrophic fibroadipose tissues. Based on clinical, radiological and histopathological findings, we diagnosed it as a case of localized macrodystrophia lipomatosa (MDL) of foot and amputation was conducted on all three toes. The family's complaints improved after surgical reconstruction and the child started walking and wearing her shoes easily after two months. Her foot had gained a normal appearance when compared to the preoperative foot.

Figure 1: Left leg bigger than right one

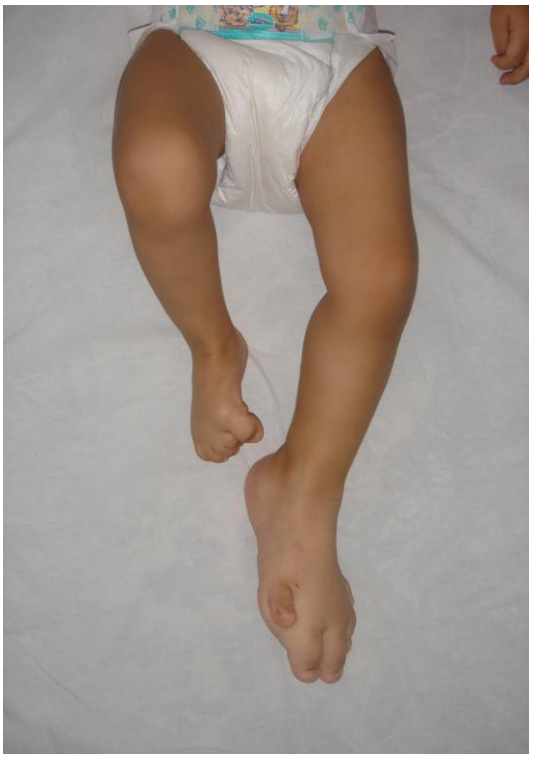
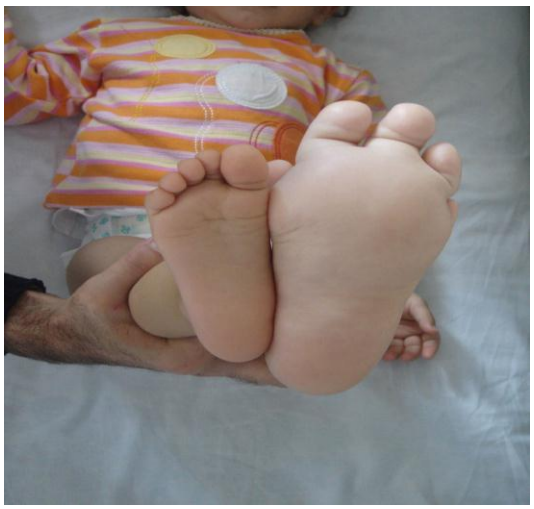


Figure 2: Left foot is hypertrophic in nature



MDL is an uncommon congenital type of localized gigantism that can involve the digits or an entire extremity and was first described by Feriz in 1925 [1-5]. It is characterized by progressive growth of all mesenchymal tissues as well as asymmetrical enlargement of fibroadipose tissue of fingers or toes. This condition rarely affects the whole extremity [2].

Radiological investigations, especially MRI, can help to make a definitive noninvasive diagnosis and to differentiate it from other causes of macrodactyly [3, 4]. The exact etiology of unilateral overgrowth of the limb due to MDL is not known; various hypotheses exist, including limb bud development anomalies during embryologic period of life [3].

This unusual entity is considered to be congenital and classified with other phakomatoses. Differential diagnosis includes several rare disorders such as Proteus syndrome, Bannayan-Riley-Ruvalcaba syndrome, familial lipomatosis,

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Correspondence to: Dr Hasan Huseyin Ceylan

Address: Department of Orthopaedics and Traumatology, Bezmialem Vakif University, Istanbul, Turkey

Email:drhhc@yahoo.com

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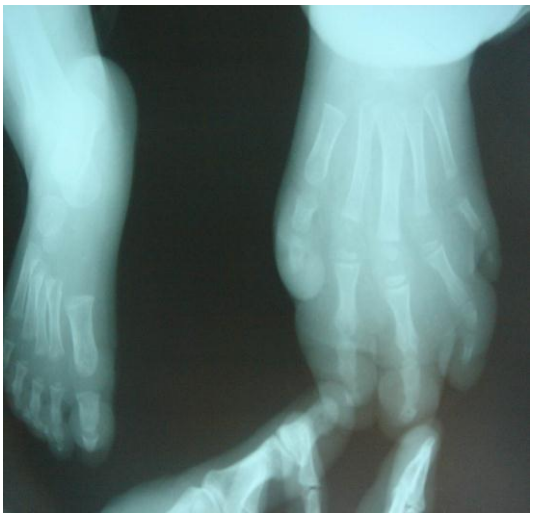
symmetrical lipomatosis, encephalocraniocutaneous lipomatosis, congenital aggressive lipomatosis, and lipoblastomatosis. Lipomas may also occur in Gardner syndrome and occasionally in neurofibromatosis. Fatty infiltration may be found in some cases of Klippel-Trenaunay syndrome [2, 4].

General clinical problems related with MDL are cosmetic and mechanical due to an enlarged extremity [1, 3, 4]. Surgical reconstruction of the affected extremity is the preferred way of treatment.

Figure 3: X-ray examination revealed that metatarsal and phalangeal bones of left foot are all hypertrophic along with the surrounding soft tissue.



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