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Bilateral Macrodystrophia Lipomatosa Hand with Fibrolipohamartoma of Median and Ulnar Nerve

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Clinical Image

Macrodystrophia lipomatosa (LMD) is a non-hereditary congenital unknown etiology disorder of localized gigantism characterized by abnormal proliferation of the fibroadipose tissue [1]. It can sometimes be associated with syndactyly, symphalangism, polydactyly, brachydactyly, or clinodactyly [2].

We exhibit a 26-year-old man referred to our institution to explore a swelling of both hands' middle and index fingers, present since birth and progressively worsening. He presented fingers painless hypertrophy on clinical examination extending to the palm (Figure 1A) with limited active and passive mobility of the metacarpophalangeal and interphalangeal joints. A standard radiograph showed bony hypertrophy of the finger phalanges with degenerative changes in the adjacent joints (Figure 1B). The ultrasound (US) revealed soft-tissue hypertrophy and interfascicular adipose tissue proliferation splaying the nerve fascicles giving an enlarged appearance (Figure 1C). Magnetic resonance imaging (MRI) confirmed hypertrophy of the median nerve with cable appearance and interfascicular fat (Figure 1D) extended to the thenar and digital branches and moderate ulnar nerve involvement (Figure 1E).

Macrodystrophia is present since birth with swelling of the affected digit or limb, and sometimes loss of function and pain due to neurovascular compression or degenerative changes [1]. Radiographs show soft tissue overgrowth with osseous hypertrophy with increased length, width, and cortical thickening [1] with or without bony outgrowths and degenerative changes in the adjacent joints. Ultrasound shows subcutaneous and inter- or intra-muscular fat hypertrophy and fibrolipohamartoma of the nerve [1]. MRI reveals interfascicular adipose tissue proliferation splaying the nerve fascicles giving a coaxial cable and a coronal spaghetti appearance and intermuscular fat hypertrophy in the muscles. Computed tomography is not indicated [1]. Bilateral involvement is rare [1]. The most common nerve territory distribution is the median nerve and the medial plantar nerve in the upper and lower limb, respectively [1], but this is the first description of joint affection of both median and ulnar nerves.

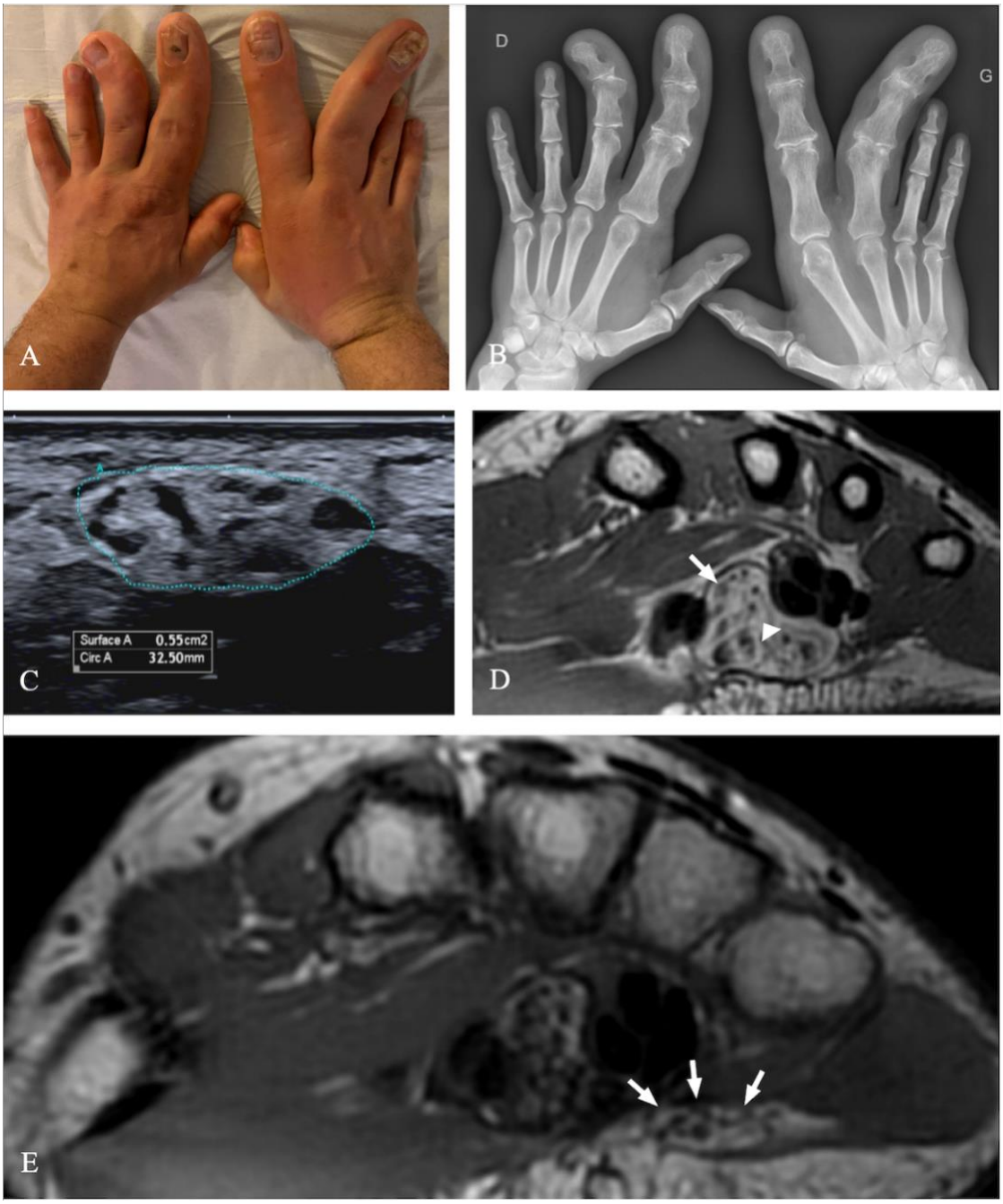

Figure 1: Bilateral macrodystrophia lipomatosa hand with fibrolipohamartoma of median and ulnar nerve; **(A):** Picture showing enlarged 2nd and 3rd fingers of the patient; **(B):** X-ray image showing hypertrophy of the soft tissues, enlarged metacarpals and phalanges of the 2nd and 3rd digits of both hands; **(C):** Ultrasound images showing an enlarged median nerve with prominent hypoechoic nerve fascicles and hyperechoic interfascicular fat; **(D):** T1-weighted MR image (TR/TE = 731/9.4 ms) in the axial plane reveals a cable appearance (arrows) with interfascicular fat (arrowhead); **(E):** T1-weighted MR image (TR/TE = 731/9.4 ms) in the axial plane shows the modular ulnar nerve involvement (cable appearance with interfascicular fat (arrows)).

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