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Clinical-Medical Image

Unusual Cause of Macrodactyly

El Houss Salma*, Zebbakh Hajar , Lrhorfi Najlae , EL Haddad Siham , Chat Latifa, Allali Nazik

Department of Radiology Mother and Child, The children's Hospital, Mohammed5 University, Rabat, Morocco

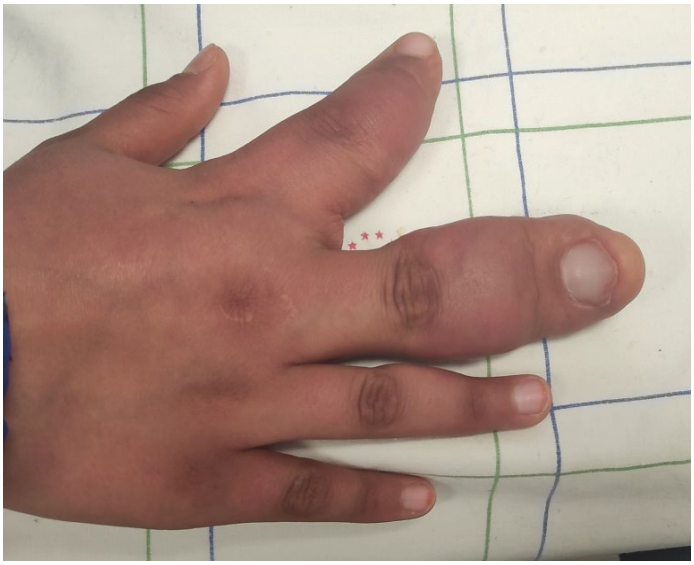

Figure 1: Clinical picture of macrodactyly.

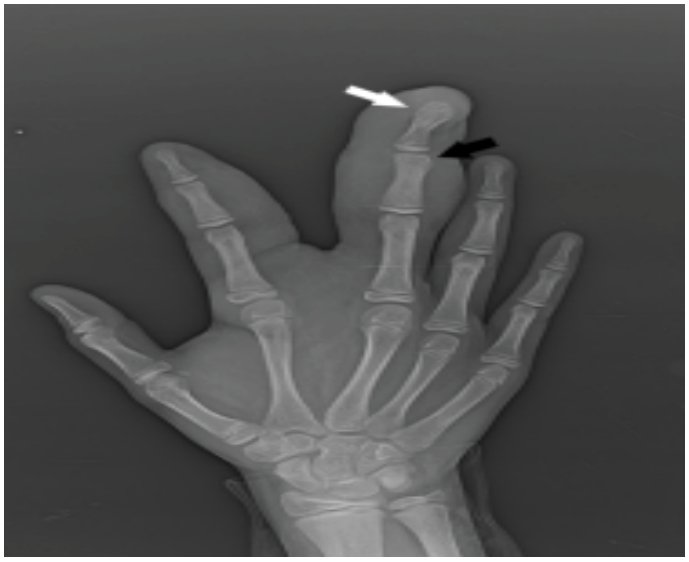

Figure 2: Standard radiograph of the right hand, showing an increase in the volume of the second and third fingers with densification of the soft tissues of the 2nd, 3rd and palmar fingers. Hypertrophy and individualization of a bony outgrowth of the middle phalanx of the 3rd finger (black arrow) with a flared and enlarged appearance of the distal end of the distal phalanx of the 3rd finger giving a mushroom-like appearance (white arrow).

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***Corresponding author:** El Houss Salma, Department of Radiology Mother and Child, The children's Hospital, Mohammed5 University, Rabat, Morocco, Tel: + 00212642294009; E-mail: salma.elhous94@gmail.com*

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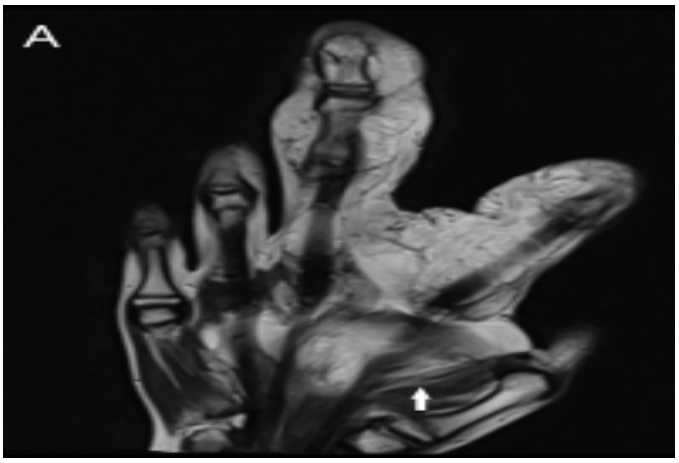

Figure 3: Magnetic resonance imaging of the right hand, T1-weighted sequence in coronal section showing hypertrophy of the palmar subcutaneous fat, as well as that of the 2nd and 3rd fingers, associated with adipose infiltration of the branches of the median nerve and of the muscles of the hand, which contain areas in T1 hypersignal (white arrow).

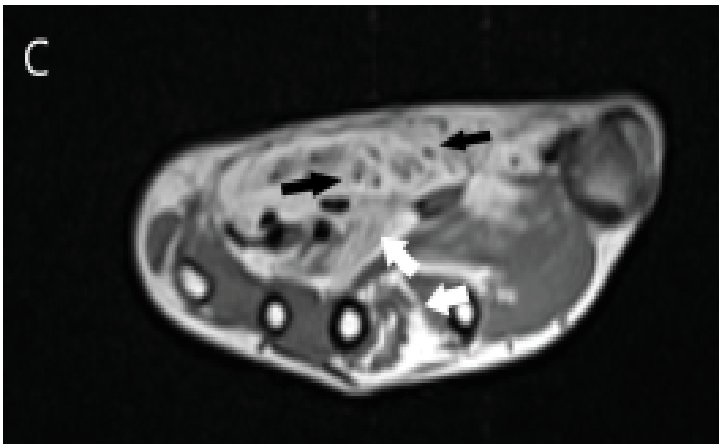

Figure 4: Magnetic resonance imaging of the right hand, T1-weighted sequence in axial section fatty infiltration of muscles (white arrows) and nerve bundles of the median nerve (black arrows) which are also hypertrophied.

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Marcrodystrophia lipomatosa is a rare congenital anomaly characterized by an abnormal growth of the mesenchymal tissues of one or more digits (usually the 2nd or 3rd digit) of the same limb (hand or foot); the damage can sometimes be bilateral or involve a whole limb [1]

This gigantism is secondary to an abnormal proliferation of the adipose tissue of the mesenchymal elements involving the subcutaneous tissue, the muscle, the nerve sheath, the bone marrow and the periosteum.

It is a non-hereditary pathology whose physiopathology is still poorly elucidated, The sex ratio of this disease is one, two subtypes have been described: the static type where the growth of the hypertrophied finger progresses at the same rate as that of the other fingers and the progressive type where the growth of the hypertrophied finger progresses more rapidly than that of the other fingers [2].

Standard radiography (Figure 1) can detect soft tissue and bone hypertrophy, the presence of radiolucent areas due to the presence of adipose tissue. On the CT scan (Figure 2), the excessive growth of bone and the proliferation of adipose tissue in the muscle fibers of the area concerned can be used as a guide to the diagnosis. Magnetic resonance imaging (Figure 3) is pathohgnomonic and can easily demonstrate fatty infiltration (T1 hypersignal with fat saturation on DP-STIR sequence) of subcutaneous tissue, muscle and nerve branches, Linear hypointense fibrous bands can be noted in this abnormal fat corresponding to the nerve bundles, bone hypertrophy and cortical thickening in the affected body part may lead to exostoses as bony outgrowths of the involved bone [1].

In addition to the interest of MRI in the positive diagnosis (Figure 4), it also allows the exclusion of other differential diagnoses such as: Neurofibromatosis type I, lymphangiomatosis, Klippel-Trenaunay disease [3], Hemangiomatosis and Proteus syndrome.

The treatment is surgical and is performed for aesthetic purposes while preserving neurological function.

Keywords: Neonatal macrodactyly; Bone hypertrophy; MRI

References

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