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Case Report

MACRODYSTROPHIA LIPOMATOSA

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SUMMARY: Macrodystrophia Lipomatosa is a congenital form of localized gigantism characterized by an increase in all mesenchymal elements particularly fibroadipose tissue. We describe two patients with Macrodystrophia Lipomatosa affecting the hand that exhibited characteristic radiological and MRI findings that distinguish the lesion from other conditions associated with localized gigantism.

CASE REPORT

Case 1: A 16-year old girl presented to the casualty department having grazed her second finger of her right hand few hours previously. The finger was tender with some swelling. History revealed that she was noted to have hypertrophy of her second and third fingers of her right hand during her first year. For her immediate injury she was treated with painkiller after excluding bone fractures. Photograph for her right hand showed hypertrophy of second and third fingers, Fig 1A. X-ray of the affected hand showed increase size of the bones and soft tissue of the second and third finger with multiple translucencies of the soft tissue. Fig 1B.

Case2: A 14 months girl was referred for X-Ray of her right hand because of macrodactaly of the second and third fingers. There was no family history of similar condition. She was noted to have this abnormality since birth. X-ray of her right hand showed severe macrodactaly Fig2A. MRI was done and showed massive accumulation of adipose tissue around the affected metacarpals, Fig2B.

DISCUSSION

Feriz first described Macrodystrophia Lipomatosa in 1925[1]. In 1967, Barsky[2] offered classification system for "true macrodactaly" dividing it into two forms. The first form is present at birth, and the size of the involved digit increases proportionally in relation to the rest of the body. The second, less common type can be complicated by fatty overgrowth in the palm, dorsum of the hand, and forearm. The degree of overgrowth increases faster than normal growth pattern. Our patients have the second type[2].

This condition is found in both the upper and lower extremities. Involvement of more than one digit is the most frequent finding; often adjacent digits are affected[3].

Most cases involve the middle and index fingers, corresponding to the territory supplied

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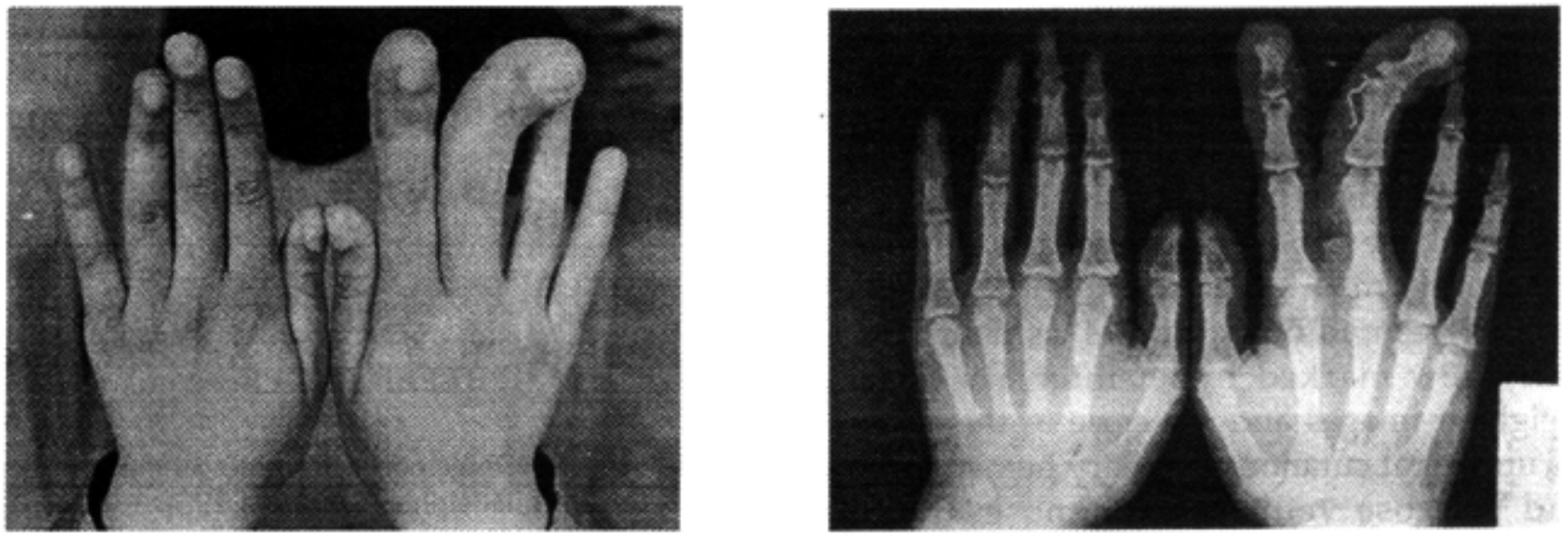

Fig 1: A. Photograph of the affected hand shows enlargement of the second and third fingers of the right hand.
B. Bilateral hand X-ray shows hypertrophy of the soft tissue and the bone of the second and third fingers.

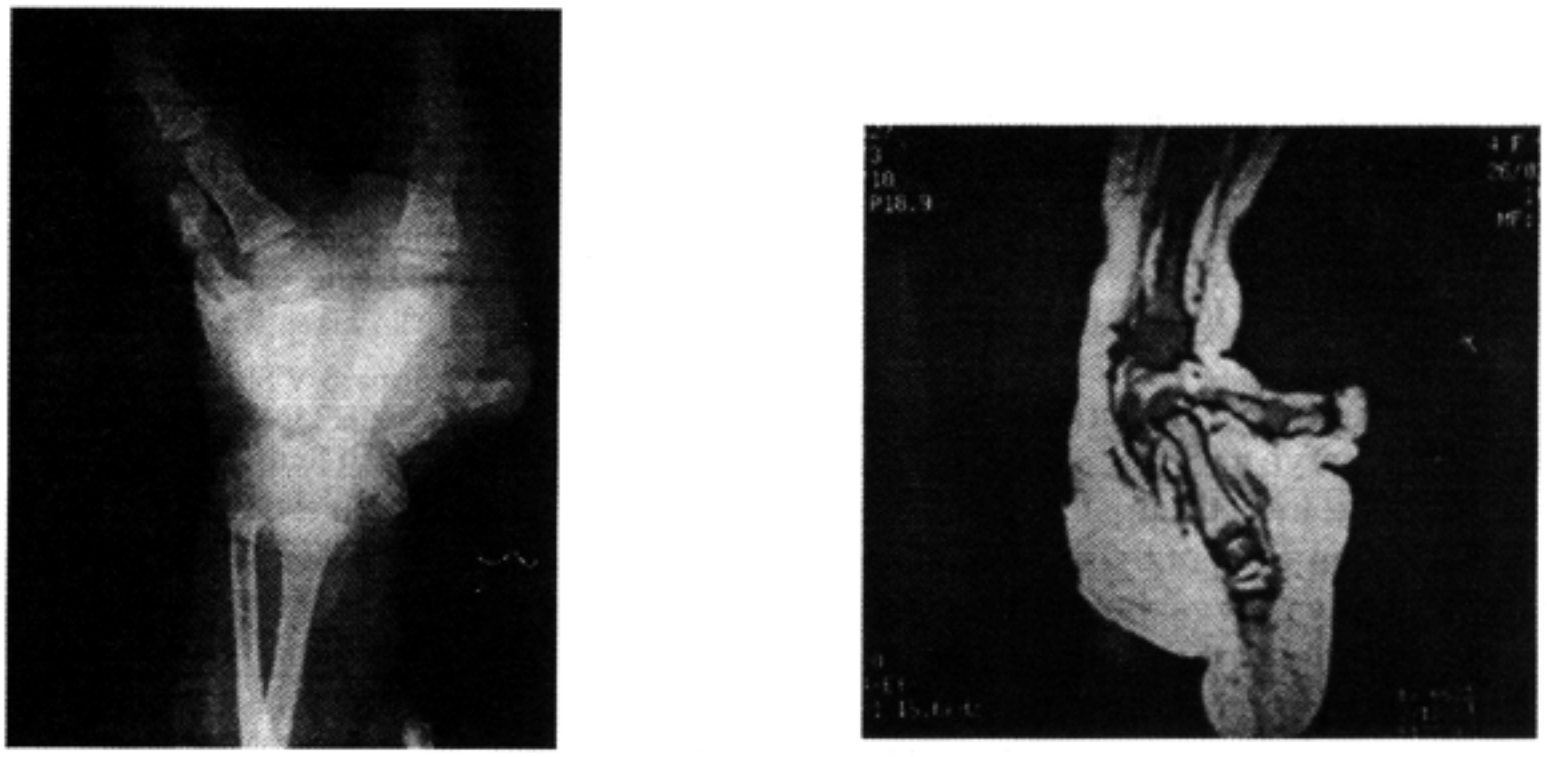

Fig 2: A. X-ray of the hands shows severe macrodactaly of the second and third fingers.
B. T1W MR images of the affected hand shows massive accumulation of adipose tissue around the involved metacarpal bones.

by the branches of the median nerve. Macrodystrophia Lipomatosa is almost always unilateral. Syndactaly and polydactaly are rare association[4].

The differential diagnosis includes neurofibromatosis, Klippel-Trenaunay-Weber syndrome, lymphangiomatosis, hemangiomatosis, and fibrolipomatus hamartoma of the nerve. Both neurofibromatosis and Macrodystrophia Lipomatosa can demonstrate limb deformity and enlargement. The later does not share the neurocautanous or familial prevalence of neurofibromatosis. Digital overgrowth ceases at puberty with Macrodystrophia Lipomatosa, and cosmoses are the primary reason for surgery.

RADIOLOGICAL APPEARANCE

Typical radiological findings of Macrodystrophia Lipomatosa are overgrowth of all tissues of digits. The phalanges are elongated and enlarged in the transverse diameter. Overgrowth of tissue may extend to the sole

or the palm of the hand leading to widened intermetacarpal spaces. The X-ray reveals soft tissue and bony overgrowth with translucencies in the soft tissue due to an increase in adipose tissue[5].

Although distal phalanges are broadened or expanded, the trabicular architecture, however, remains normal. MR imaging allows distinction between all other entities, as neurofibromas show marked hyper-intensity on T2-weighted images and are close to the nerves. Its unilateral cutanous capillary hemangiomas and varicose veins can diagnose klippel-Trenaunay-Weber syndrome.

Diffuse swelling and pitting oedema, characteristic findings of lymphangiomatosis, were not seen in the patients with Macrodystrophia Lipomatosa[6,7].

In hemangimatosis long TR/TE sequences would show a striated and septated configuration of high-signal-intensity channels that correlate with the vascular channels and fibrous strands, which are found in hemangiomas.

When confined to the hand, fibrolipomatous hamartoma of the nerve and Macrodystrophia Lipomatosa may be indistinguishable on the bases of MRI findings. Both of these lesions can be found in the median nerve distribution, and both will demonstrate fibrous thinking of the nerve on MR imaging, but in fibrolipomatous hamartoma the fat deposition is within the nerve sheath, whereas Macrodystrophia Lipomatosa demonstrates fat deposition within the whole limb[6].

MR imaging is uniquely suited to investigate Macrodystrophia Lipomatosa. Accurate soft tissue characterization will confine the differential diagnosis to only few possibilities redundancy of fatty tissue and fibrous thickening of a nerve, in the presence of macrodactaly, should lead to a diagnosis of Macrodystrophia Lipomatosa.

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